Clinical Scenarios

Essential case series

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Edited by

Roland Gamini Sirisinghe

Win Min Thein

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Writing this book took longer than we expected however we believe more rewarding than we have ever imagined. None of this would be possible without the support of family and friends of Taylor's School of Medicine. We would like to extend our special thanks to students who are patiently waiting.

Contributors

Preface

This first volume of the ‘Clinical Scenarios – Essential Case Series’ is a compilation of case discussions by a group of experienced staff, who have many years of experience in teaching undergraduate medical students, and also examining medical undergraduates in their examinations at various stages of clinical learning.

Medical textbooks in the clinical fields are aplenty. These include a fair number of books based on clinical cases. Even under these conditions we are confident that adding a further title to this already substantial collection of available books is worthwhile. This confidence is derived from two attributes of this book, which we think distinguish it from many others available to medical students.

Firstly, the discussion of the cases is intentionally kept as close to a bedside teaching session as possible. Thus the discussions that follow each of the cases attempts to bring out the thought process that is required when dealing with the patient rather than being a mere academic discourse on the respective diseases. The tone of the discussion is also kept at a more conversational level than the formal textbook level, of course only to a level permitted by the written language. However, keeping in mind that factual information is necessary knowledge for students and medical practitioners, essential information is provided. But the book should not be looked at as a provider of comprehensive factual information on these cases. With the authors being experience examiners at undergraduate clinical examinations, the discussions are also formulated to indicate the probable lines of questioning at clinical examinations. The cases, therefore, are presented somewhat differently from a classical ‘case study’ format of presentation. It is our hope that these case discussions that are more from a student point of view will be appreciated and found to be of much benefit by the students.

Secondly, this book, unlike almost all other similar titles, is not specialty specific. The level of knowledge expected of a fresh medical graduate is different in different specialties. There are also dissimilarities in the aspects of patient care empahsised at undergraduate level in different specialties. In some specialties the expected competency may be primarily diagnosis while in others immediate management is essential. The cases in different specialties in this

book reflect, to some extent, this difference. This is an advantage of this book, which is often not apparent in specialty-specific book – certainly textbooks and even case-based texts.

This book is suitable for medical students at all stages of their clinical training, whether at entry level or preparing for their final examinations. The book is best not used as a resource of factual information; as previously mentioned only information relevant to the discussion is available in this book. It is best used to learn the thought process a student should aim to develop during clinical training. The book includes a few representative cases from each discipline. Therefore, it is not intended to be a comprehensive compilation of all common diseases one encounters in each specialty. But the student may extrapolate the line of thought from one disease entity to another, particularly in the same specialty. This itself could be considered as part of clinical training.

The book used questions that follow each case as a means of illustrating the questions a student must raise in his own mind while seeing a patient. The answers to the questions are provided in the discussion. The student, while using this book, must not merely learn the answer to a particular question; it is equally important to learn what questions the student must raise in his own mind.

We would consider our objective achieved, if students, with the use of this book, become more adapt in developing a thinking mind as all clinicians should have.

Roland Sirisinghe

Win Min Thein

Contributing Authors

Dr. Abdul Kareem Meera Mohaideen
Associate Professor of Radiology
School of Medicine
Taylor's University
Malaysia

Dr. Anitha Ponnupillai
Associate Professor of Obstetrics & Gynaeology,
School of Medicine
Taylor's University
Malaysia

Dr Benjamin Samraj Prakash Earnest
Senior Lecturer in Internal Medicine
School of Medicine
Taylor's University
Malaysia

Dr. Eugene Leong,
Associate Professor, and Consultant Obstetrician & Gynecologist,
School of Medicine
Taylors University
Malaysia.

Dr Ihab Ali
Associate Professor of ORL and Head &Neck Surgery
School of Medicine
Taylor's University
Malaysia

Dr Jo Ann
Senior Lecturer in Community Medicine
School of Medicine
Taylor's University
Malaysia

Dr Khine Pwint Phyu
Senior Lecturer in Obstetrics & Gynaecology
School of Medicine
Taylor's University
Malaysia

Dr Lim Yin Sear
Senior Lecturer, Paediatric Haematologist & Oncologist.
School of Medicine
Taylor's University
Malaysia

Dr Loh Keng Yin
Associate Professor of Family Medicine
School of Medicine
Taylor's University
Malaysia

Dr Prabal Bhargava
Associate Professor of Ophthalmology
School of Medicine
Taylor's University
Malaysia

Dr Punithavathy Shanmuganathan
Senior Lecturer in Family Medicine
School of Medicine
Taylor's University
Malaysia

Dr Su-Yin Lim
Senior Lecturer in Internal Medicine (Neurology)
School of Medicine
Taylor's University
Malaysia

Dr Tint Lwin
Senior Lecturer in Emergency Medicine
School of Medicine
Taylor's University
Malaysia

Dr Win Min Thein
Associate Professor of Orthopaedics
School of Medicine
Taylor's University
Malaysia

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Case 1

A 44-year-old woman presents with fever, painful right ankle and abdominal discomfort for the past 2 weeks. She was diagnosed with rheumatic mitral valve stenosis 5 years back. On examination, there are red painless macules on both palms and clubbing of fingers. Pulse is regular and blood pressure is 110/76 mmHg. The right ankle is tender along the joint line with joint effusion. Cardiovascular examination reveals a pan-systolic murmur in the mitral area and a mid-diastolic murmur. Echocardiography reveals a new-onset mitral regurgitation in addition to mitral stenosis with vegetations on the anterior mitral leaflet.

Questions

1. State your provisional diagnosis? Give reasons.
2. How will you confirm your diagnosis?
3. How will you investigate this patient?
4. How will you manage this patient?

Discussion

1. **State your provisional diagnosis? Give reasons.**

   This patient has rheumatic mitral valve stenosis and now presents with a febrile illness. The conditions to be considered under the situation are infective endocarditis and rheumatic fever (reactivation). Even though a painful large joint involvement might suggest rheumatic fever, it could also occur due to septic arthritis from an infective embolus in infective endocarditis. The skin rash of infective endocarditis is seen on the fingers (Osler's nodes) and palms (Janeway lesions), while that of rheumatic fever is a ring lesion (erythema marginatum) with central clearing seen on the limbs and torso. Clubbing, new-onset mitral regurgitation and vegetations on the mitral valve indicate infective endocarditis.

2. **How will you confirm your diagnosis?**

   Diagnosis of infective endocarditis requires fulfilling modified Duke's criteria. There are 2 major (convincing echocardiographic evidence and culture of an organism in at least 2 samples) and 5 minor criteria (fever, predisposing factor, suspicious echocardiographic lesions, culture of an organism in one sample and immunological/embolic phenomena). To make a definitive diagnosis of infective endocarditis 2 major or 1 major and 3 minor or 5 minor criteria should be fulfilled. This patient has convincing echocardiographic evidence (1 major criterion), fever, predisposing factor and embolic/immunologic phenomena (3 minor criteria) fulfilled, making a definitive diagnosis of infective endocarditis.

3. **How will you investigate this patient?**

   Even though a definitive diagnosis could be made by using the available physical findings and investigations according to Duke's criteria, further investigations would be required to manage this patient and to identify complications of infective endocarditis.

Culture of the organism from blood samples obtained 30 minutes apart from different sites (before administration of antibiotics) helps identify suitable antibiotics to treat the patient (by de-escalation from empirical antibiotic to the most sensitive one) apart from being a major criterion for diagnosis.

Full blood count and peripheral smears could help identify microangiopathic haemolytic anaemia. Urinalysis and renal functions tests could identify renal involvement (crescentic glomerulonephritis). ECG helps identify AV block which might indicate presence of paravalvular abscess. Chest x-rays, cardiac biomarkers (BNP/NTproBNP) and echocardiography would help identify presence of heart failure which usually occurs when valves are destroyed and require surgical intervention.
Other complications like visceral abscesses, disseminated intravascular coagulation, septic arthritis may be investigated with ultrasonogram, coagulation screen and joint aspiration respectively.

Serum electrolytes, ESR/CRP, Rheumatoid factor and complement levels may be useful too. CT scan and MRI scan are useful for identifying valve damage and for neuroimaging in patients with cerebral emboli.

4. How will you manage this patient?

Wherever possible patients with endocarditis should be treated by an “endocarditis team” which would consist of cardiologists, cardiothoracic surgeons, infectious disease physicians, neurologists, neurosurgeons, and microbiologists.

Initial management should concentrate on airway, breathing and circulation. Particularly, look out for patients with cardiac failure who would have severely damaged valves requiring early surgery (50% of patients with IE would require surgery). Haemodynamic stability should be the goal and managed with appropriate haemodynamic monitoring and inotropic drugs.

It is essential to obtain blood cultures prior to the initiation of antimicrobial therapy, as even one dose often eliminates bacteraemia, producing a false negative culture report. The most common organism causing infective endocarditis on native valves is Streptococcus viridans for which a beta-lactam antibiotic or vancomycin is given for 4-6 weeks.

Case 2

A 60-year-old man presented with recurrent respiratory and urinary infections over the past few months. Now he presents with fever, weight loss and swelling in the neck for 3 weeks. On examination, he is pale and there are multiple bilateral enlarged cervical lymph nodes which are noted to be discrete, rubbery and non-tender. There is no organomegaly or palpable mass on abdominal examination. Peripheral blood smear shows lymphocytosis which is confirmed to be due to monoclonal B cells on immunophenotyping. Haemoglobin is 90 g/L and direct Coomb's test is positive.

Questions

1. Give 3 differential diagnosis.
2. What is your diagnosis? Give reasons.
3. How will you assess prognosis in this patient?
4. How will you manage this patient?

Discussion

1. **Give 3 differential diagnosis.**

    a. Chronic lymphocytic leukaemia
    b. Hodgkin's disease
    c. Non-Hodgkin's lymphoma

2. **What is your diagnosis? Give reasons.**

    Chronic lymphocytic leukaemia

    Even though all the above conditions can present with cervical lymphadenopathy with similar characteristics, fever and weight loss, lymphomas often cause a myelophthisic anaemia with leukoerythroblastic picture with bone marrow involvement. This occurs due to extramedullary haematopoiesis releasing myeloid precursors into the circulation with characteristic tear drop cells (dacryocytosis). This is often associated with splenomegaly and lymphopenia in lymphomas. Autoimmune haemolytic anaemia due to warm antibodies may be seen in all 3 but more so in chronic lymphocytic leukemia (CLL). Itching and alcohol induced pain are characteristic of Hodgkin's lymphoma (HL). Extranodal involvement (splenic and hepatic) and involvement of deeper lymph node groups (paraaortic, mesenteric & mediastinal) are more common in non-Hodgkin's lymphoma (NHL). All three conditions are most commonly due to monoclonal proliferation of B lymphocytes and NHL is more common among the three. However, CLL is more likely to cause peripheral lymphocytosis. Hypogammaglobulinemia accounts for the recurrent infections in CLL.

3. **How will you assess prognosis in this patient?**

    Prognosis primarily depends on the stage. Patients with lymphadenopathy [8 years], organomegaly (splenomegaly and hepatomegaly) [5 years] and marrow infiltration (indicated by anaemia [2 years] and thrombocytopenia [1 year]) have a poorer prognosis compared patients with lymphocytosis alone [13 years median survival].

Also, patients who express ZAP70 and CD38 and those with unmutated variable region of the immunoglobulin gene have a poor prognosis.

4. **How will you manage this patient?**
    a. Consider chemotherapy if the patient is symptomatic. CLL is an indolent malignancy and treatment is required only if there is evidence of bone marrow failure. Only a third of patients have a rapid progression. The rest may progress slowly, not progress at all or even regress spontaneously. Patients should be reassured about the indolent nature of the disease.
    b. Fludarabine + rituximab ± cyclophosphamide is 1st line therapy. Ibrutinib, chlorambucil, bendamustine, ofatumumab and stem-cell transplantation may have a role.
    c. Managing specific complications:
        i. Steroids may be used for autoimmune haemolytic anaemia.
        ii. Radiotherapy for lymphadenopathy and splenomegaly.
        iii. Splenectomy may help improve low blood counts due to autoimmune destruction or hypersplenism and can provide relief in massive splenomegaly.

Case 3

A 50-year-old man presents with polyuria and polydipsia for the last 4 weeks. He leads a sedentary lifestyle. On examination, he has a BMI of 30 and abdominal girth of 110 cm. He has corneal arcus, xanthelasma and oral candidiasis. Pulses are equal and 76 beats per minute. Blood pressure is 110/74 mmHg. Cardiovascular examination and examination of the lower limbs are normal. A random blood glucose is 11 mmol/L.

Questions

1. What is your diagnosis? Give reasons. How will you confirm your diagnosis?
2. How will you manage this patient?
3. How will you assess glycaemic control on follow-up?
4. What are the complications associated with poor control of this condition?

Discussion

1. **What is your diagnosis? Give reasons. How will you confirm your diagnosis?**

   Type 2 diabetes mellitus (DM)

   This patient has risk factors for diabetes mellitus viz., age, sedentary lifestyle and central obesity.

   The random blood glucose is 11 mmol/L which indicates DM. This should be confirmed with another test for DM. One of the following tests can be used to establish a firm diagnosis of diabetes:

   a. Fasting plasma glucose (FPG) >6.9 mmol/L (>125 mg/dL)
   b. Random plasma glucose >11.1 mmol/L (=200 mg/dL) with diabetes symptoms such as polyuria, polydipsia, fatigue, or weight loss
   c. 2-hour post-load glucose >11.1 mmol/L (=200 mg/dL) on a 75 g oral glucose tolerance test
   d. HbA1c >48 mmol/mol (6.5%)

   All of these require confirmation with a second test, which may be the same test or a different test.

2. **How will you manage this patient?**

   a. Appropriate diet
      i. More vegetables
      ii. Low in fats and carbohydrates
   b. Lifestyle changes
      i. Increase physical activity
      ii. At least walk for 30 min. on most days
   c. Medications
      i. Insulin and Oral hypoglycaemic agents
         1. Biguanides - metformin

2. Sulfonylurea - glibenclamide, glipizide
3. Thiazolidinediones - pioglitazone, rosiglitazone
4. Alpha-1-glycosidase inhibitor - acarbose, oglibose
5. Meglitinides - repaglinide, mitiglinide, nateglinide
6. GLP-1 agonist - exenatide, liraglutide, lixisenatide, semaglutide
7. SGLT-2 inhibitor - canagliflozin, dapagliflozin, empagliflozin, ertugliflozin
8. DPP-4 inhibitor - sitagliptin, saxagliptin, vildagliptin, linagliptin
9. PTP1B inhibitor - flavonoids, phenols, terpenes, sterols (>300 naturally occurring substances, experimental)
10. Insulin (Human & analogue)
    a. Regular short acting insulin
    b. Intermediate acting insulin NPH
    c. Rapid-acting analogue - aspart, lispro, glulisine
    d. Long-acting analogue - detemir, glargine, degludec

d. Treat other cardiovascular risk factors
    i. Patients with type 2 diabetes have a very high risk of concurrent hypertension (80% to 90%), lipid disorders (70% to 80%), and overweight or obesity (60% to 70%)

3. How will you assess glycaemic control on follow-up?

Using HbA1c (glycosylated haemoglobin), values <6.5% indicate good control

4. What are the complications associated with poor control of this condition?

Acute complications

Diabetic ketoacidosis

Non-ketotic hyperosmolar state

Hypoglycaemia (Somogyi phenomenon)

- Chronic complications
  - Microvascular complications
    - Retinopathy
    - Neuropathy
    - Nephropathy
  - Macrovascular complications
    - Coronary artery disease
    - Stroke/transient ischaemic attack
    - Peripheral artery disease
- Miscellaneous
  - Periarthritis
  - Dermopathy (panniculitis)
  - Poor wound healing
  - Predisposition to infections
  - Foot ulcers and gangrene
  - Cataract
  - Rubeosis iridis (neovascular glaucoma)

Case 4

A 30-year-old man who received a tattoo 8 months ago developed fever and jaundice since then. On examination his liver is just palpable below the right costal margin. Results of his liver function tests and viral markers are given below.

Bilirubin (Total) 38 mmol/L [Normal 1-20 mmol/L]

Bilirubin (Direct) 18 mmol/L [Normal <5 mmol/L]

Alanine aminotransferase 398 U/L [Normal 7-56 U/L]

Aspartate aminotransferase 245 U/L [Normal 10-40 U/L]

Alkaline phosphatase 56 U/L [Normal 20-140 U/L]

Gamma glutamyl transpeptidase 36 U/L [Normal 10-50 U/L]

Albumin 2.9 g/dL [Normal 3.5-5.5 d/dL]

INR 1.26

HBsAg +ve

Anti-HBs -ve

Anti HBc IgM -ve

Anti HBc IgG +ve

HBeAg +ve

Anti HBe -ve

HBV DNA 650000 IU/ml

Serology for HAV/HEV/HIV are negative

Questions

1. What is your diagnosis? Give reasons.
2. How will you assess the degree of necro-inflammation and fibrosis in this patient?
3. How will you manage this patient?

Discussion

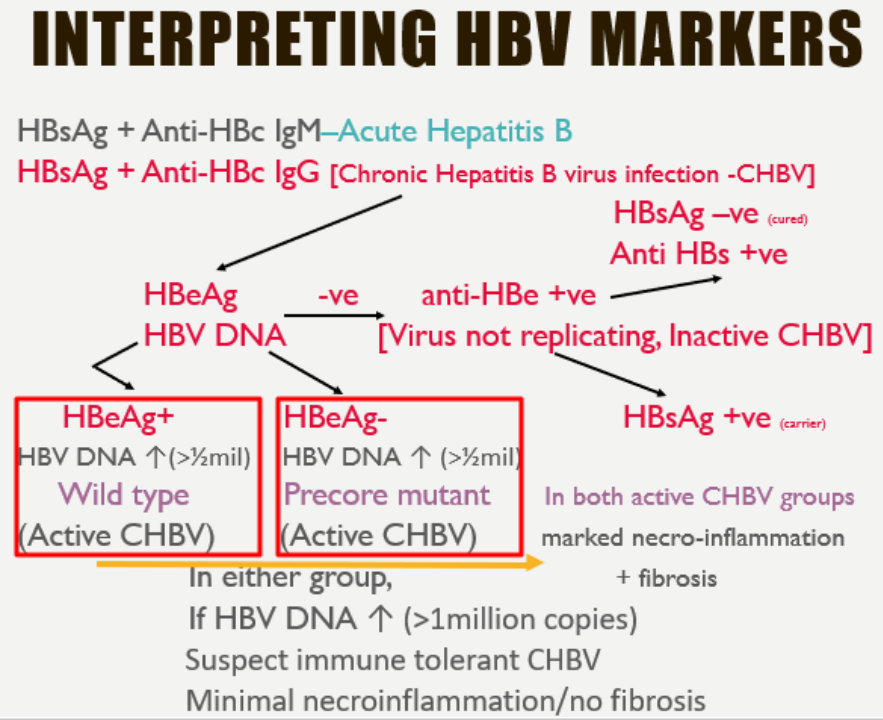
1. **What is your diagnosis? Give reasons.**

The liver function test shows a hepatocellular pattern with low albumin indicating chronicity of the liver disease. HBsAg is +ve indicating presence of hepatitis B virus (HBV) infection. Anti HBc IgG is +ve indicating chronic HBV infection. Since HBeAg is +ve and HBV DNA is raised, this is a chronic immune active HBV infection. Since the HBV DNA load is <1 million and the transaminases (being markers of ongoing necro-inflammation) are raised, this is not immune tolerant chronic HBV infection.

The three classes of chronic HBV infection are given below

a. Immune tolerant chronic HB: HBeAg +ve, HBV DNA >1 million IU/ml, liver biopsy -minimal necro-inflammation and no fibrosis
b. Immune active chronic HB: HBeAg +ve or -ve, HBV DNA >2000 IU/ml (HBeAg -ve) or >20000 IU/ml (HBeAg +ve), moderate to severe necro-inflammation, with/without fibrosis
c. Immune inactive chronic HB: HBeAg -ve, HBV DNA <2000 IU/ml, no necro-inflammation, variable fibrosis

An algorithm for interpreting markers of HBV infection is given below



2. How will you assess the degree of necro-inflammation and fibrosis in this patient?

Ideally, a liver biopsy should be done to identify the degree of necro-inflammation and fibrosis using the modified histologic activity index (HAI, Knodell Ishak score) in this patient. But in the presence of prolonged prothrombin time (INR >1.1), alternatives may be considered. Transaminases are reasonable markers of necro-inflammation and many other alternatives are available for non-invasive assessment of fibrosis.

Scores based on serological markers are available and include the following.

a. Fib-4 score (based on age, ALT, AST & platelet count)

FIB-4 was developed to correlate with Ishak levels of fibrosis (by biopsy) with 3 levels:

i. 0-2 (mild fibrosis)

ii. 3-4 (moderate fibrosis)

iii. 5-6 (severe fibrosis/cirrhosis)

b. FibroTest score. The estimation is made by measuring 6 standard serum biomarkers (gamma-glutamyl transferase, total bilirubin, alpha-2-macroglobulin, apolipoprotein A1, haptoglobin, and alanine aminotransferase)

FibroTest Score	Stage	Interpretation
0.00-0.21*	F0	No fibrosis
0.21-0.27*	F0-F1	No fibrosis
0.27-0.31*	F1	Minimal fibrosis
0.31-0.48*	F1-F2	Minimal fibrosis
0.48-0.58*	F2	Moderate fibrosis
0.58-0.72*	F3	Advanced fibrosis
0.72-0.74*	F3-F4	Advanced fibrosis
0.74-1.00	F4	Severe fibrosis (Cirrhosis)

Alternatively imaging studies can be used to assess liver fibrosis and include the following.

Transient elastography or shear wave elastography [also called Fibroscan]
(Similar to USG –TE provides a reliable method of detecting cirrhosis and excluding significant fibrosis, particularly when the results are supported by clinical and laboratory data.)
Scarring has 4 stages: • F0 means no scarring • F1 is mild fibrosis • F2 is moderate fibrosis • F3 is severe fibrosis • F4 is cirrhosis or advanced fibrosis

MR elastography
The main advantage over ultrasound elastographic techniques is that a larger portion of the liver is sampled, reducing sampling bias.

3. How will you manage this patient?

The treatment goal in chronic hepatitis B is to eliminate the virus (HBsAg +ve → HBsAg -ve), suppress serum HBV DNA to undetectable levels and achieve seroconversion (Anti HBs -ve → Anti HBs +ve). Treatment may sometimes achieve seroconversion from HBeAg +ve to HBeAg -ve and anti-HBe +ve in chronic HBV infection.

Entecavir, tenofovir disoproxil or tenofovir alafenamide, or peginterferon alfa 2a (not combination) are the recommended first-line drugs for chronic immune-active hepatitis B infection. However, achievement of all treatment goals is rare but disease progression and complications are prevented equally with any of the drugs. Treatment may be stopped on achievement of either of the treatment goals. Antiviral drug therapy is not recommended in acute, chronic immune-tolerant or chronic inactive hepatitis B.

Case 5

A 60-year-old man presents with a 4-day history of increasing breathlessness associated with wheezing and a productive cough with yellowish sputum. He denies any haemoptysis, chest pain or fever. Normally, he can walk comfortably for about 300 meters on a flat surface before having to stop to rest. Over the past few days, he has felt breathless with routine housework and has had to stop to catch his breath after walking 20 to 30 meters.

Over the past few years, he has had several episodes of 'chesty coughs' associated with breathlessness, wheezing and sputum production without fever. These episodes typically lasted 5 to 7 days and were self-limiting, therefore he had not sought medical help for them. He decided to seek help this time as he feels that the current episode 'is the worst one yet'. His past medical history is notable for hypertension. He currently takes amlodipine 5 mg daily. He has smoked 10 to 15 cigarettes a day since the age of 20.

On examination, he demonstrates pursed lip breathing and use of accessory respiratory muscles, and there is a widespread wheeze. His blood pressure is 142/90 mmHg, pulse is 88 (regular), respiratory rate 28 breaths per minute, pulse oxygen saturation 88% on air and temperature 36.6°C.

His ECG shows sinus rhythm and his cardiac biomarkers are normal. Laboratory investigations reveal a normal haemoglobin level, white cell count, renal function and brain natriuretic peptide (BNP).

His chest X-ray reveals hyperinflated lung fields. The cardiac contours appear normal and there are no signs of consolidation or effusion.

Questions

1. What is your provisional diagnosis and how can you confirm it in this patient?
2. How would you assess the severity of this condition?
3. What are the main treatment modalities in the acute management of this condition?
4. What are the main principles of long-term management in this condition?

Discussion

1. What is your provisional diagnosis and how can you confirm it in this patient?

The patient's history of recurrent episodes of cough with sputum, breathlessness and wheezing should make you consider an underlying diagnosis of chronic obstructive pulmonary disease (COPD), asthma, heart failure or bronchiectasis. The older age of onset of symptoms in this patient makes asthma less likely, while the lack of a history of copious purulent sputum and recurrent bacterial infections make bronchiectasis unlikely. The long history of smoking supports the diagnosis of COPD.

Patients with COPD typically present with a longstanding history of chronic cough, sputum and exertional dyspnoea. Others, like this patient, may present with intermittent episodes of increased cough, sputum and dyspnoea suggestive of acute exacerbations of COPD (AECOPD).

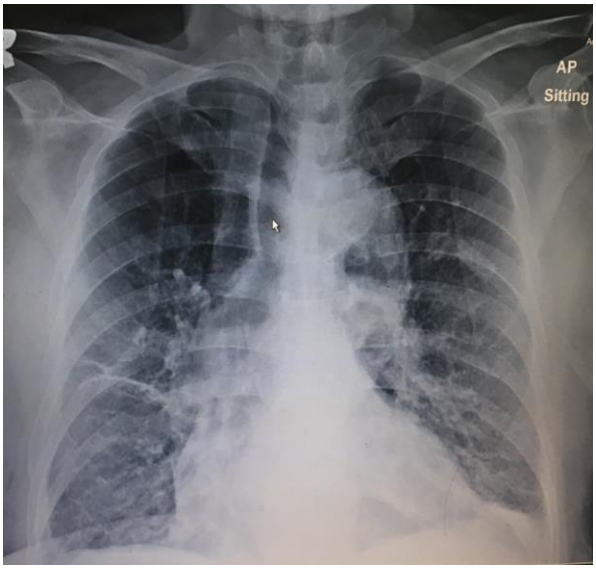
The diagnosis of COPD is confirmed on spirometry which should show evidence of airflow obstruction, typically demonstrating a FEV1/FVC ratio of 0.70 or less on post-bronchodilator spirometry. However, spirometry should not be performed during an acute exacerbation as it will not accurately reflect the patient's usual disease severity. Spirometry for COPD is usually performed when the patient is stable and recovered from the episode of exacerbation.

While a chest X-ray has a low sensitivity for detecting signs of COPD, it is useful for the evaluation of other possible causes of breathlessness such as pneumonia, heart failure or pneumothorax. The chest X-ray findings in this patient did not suggest any of these differentials. In advanced COPD, chest X-ray features may include hyperinflated lung fields extending beyond six anterior ribs and flattening of the diaphragmatic contours (Figure 1). Emphysematous bullae may be present. If cor pulmonale is present, enlargement of the pulmonary arteries at the hila and enlargement

of the cardiac silhouette may be seen. Infective changes may be seen on the chest X-ray during an infective exacerbation.

Therefore, the diagnosis of AECOPD is a clinical diagnosis based on the history, examination findings, and the exclusion of alternative diagnoses that can cause similar symptoms.

Figure 1. This is an anterior-posterior (AP) chest X-ray of a patient who presented with an acute exacerbation of COPD. The X-ray is rotated, giving rise to the appearance of 'tracheal deviation'. There are no mediastinal masses. The lung fields are hyperinflated and there is flattening of the diaphragm. There are increased bronchovascular markings in the mid to lower zones bilaterally. There is linear atelectasis in the right mid-zone. The heart size is exaggerated due to the AP projection.



2. How would you assess the severity of this condition?

Assessing severity in AECOPD

The severity of AECOPD is assessed clinically and guides the management approach, discussed in the next section. During any exacerbation, typical physical examination findings include tachypnoea, tachycardia, and diffuse wheezing. Signs of severe

respiratory insufficiency include the use of accessory muscles of respiration, an inability to lie flat, inability to speak in complete sentences, diaphoresis, agitation, reduced breath sounds and thoracoabdominal asynchrony. Impending respiratory arrest is manifested by an inability to maintain respiratory effort, paradoxical breathing, cyanosis, haemodynamic instability or reduced consciousness.

Arterial blood gas sampling is indicated in all patients presenting with severe respiratory insufficiency. There is no role for spirometry or peak expiratory flow measurement in AECOPD.

Assessing the severity of underlying COPD

The severity of the patient's underlying COPD should be assessed when he is stable and recovered from his exacerbation. One of the most widely used staging systems for COPD severity is the Global Initiative for Chronic Obstructive Lung Disease (GOLD) scoring which takes into consideration patients' symptoms using the modified Medical Research Council dyspnoea scale and COPD assessment test (CAT) and exacerbation frequency (Table 1), as well as spirometry findings (Table 2). Combined, this information can help direct therapeutic management.

Table 1. GOLD classification by symptoms and exacerbation frequency

GOLD classification	Exacerbation frequency, hospital admissions	CAT and mMRC scores	Severity
A	0 or 1, no hospital admissions	CAT < 10, mMRC 0 or 1	Minimally symptomatic, low risk of exacerbations
B	0 or 1, no hospital admissions	CAT ≥ 10, mMRC ≥ 2	More symptomatic, low risk of exacerbations
C	≥2 exacerbations per year/ one requiring hospital admission	CAT < 10, mMRC 0 or 1	Minimally symptomatic, high risk of exacerbations

D	≥2 exacerbations per year/ one requiring hospital admission	CAT ≥ 10, mMRC ≥ 2	More symptomatic, high risk of exacerbations

Table 2. GOLD classification by spirometry findings

GOLD classification	Severity	% of predicted FEV1
GOLD 1	Mild	> 80%
GOLD 2	Moderate	50 – 80%
GOLD 3	Severe	30 – 50%
GOLD 4	Very severe	< 30%

3. What are the main treatment modalities in the acute management of this condition?

The main treatment modalities in AECOPD in the hospital setting are summarized as follows:

- Bronchodilators and corticosteroids

  Inhaled short-acting beta adrenergic agonists such as albuterol are the mainstay of treatment for AECOPD, usually combined with an anti-muscarinic agent such as ipratropium and administered via a nebulizer. For most cases of AECOPD requiring hospitalization, a course of systemic corticosteroids is usually administered.

- Exacerbation trigger and antibiotic therapy

  AECOPD is commonly triggered by a respiratory tract infection (viral or bacterial) but can also be idiopathic or due to air pollution. Alternative causes of acute dyspnoea in COPD patients should also be considered, such as decompensated heart failure, acute myocardial infarction or pulmonary embolism.

An increase in sputum volume and purulence alongside worsened dyspnoea are features that are more indicative of a bacterial infection, hence antibiotics may be beneficial in these patients.

- Respiratory support

  Supplemental oxygen therapy should aim to achieve an oxygen saturation of between 88% and 92% (equivalent to a PaO_2 of 60 to 70 mmHg) in AECOPD patients. Excess oxygen risks worsening hypercapnoea in those with chronically elevated $PaCO_2$ due to blunting of their ventilatory drive.

  Non-invasive positive pressure ventilation (NIPPV) is indicated patients who develop acute respiratory acidosis (type 2 respiratory failure). NIPPV reduces the need for intubation, the length of stay in hospital, and mortality.

4. What are the main principles of long-term management in this condition?

- Symptomatic control with bronchodilators

  Although airway obstruction is not fully reversible in COPD, the mainstay of pharmacological therapy in COPD are bronchodilators. Treatment regimens vary according to disease severity. For patients with mild symptoms and low risk of exacerbation (GOLD A), a short-acting beta agonist (SABA) and/or short-acting muscarinic antagonist (SAMA) to be used when required are usually sufficient.

  For patients with moderate to severe symptoms but low risk of exacerbation (GOLD B), a regular long-acting beta agonist (LABA) or muscarinic antagonist (LAMA) should be added to the above regimen.

  Patients with a high risk of exacerbation (GOLD C and D) should have a regular LAMA along with a SABA to be used as required. If severe breathlessness is present, a combined glucocorticoid-LABA inhaler may be added in.

- Long-term O_2 therapy for hypoxaemia

Long-term, continuous oxygen therapy is recommended in COPD patients with chronic hypoxaemia ($PaO_2 \leq 55$ mmHg or pulse oxygen saturation ≤ 88%), targeting oxygen saturation to 88 to 92%, to improve survival and quality of life.

- Others:
  - Smoking cessation
  - Pulmonary rehabilitation
  - Prevention of respiratory infections with vaccinations against pneumococcus, influenza and pertussis

References

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Case 6

A 65-year-old right-handed woman is brought to the emergency department with new-onset weakness and difficulty speaking upon waking from sleep this morning. She has no significant past medical history of note.

On examination, there is weakness of her right arm and leg. There is noticeable drooping of the right corner of her mouth and loss of the right nasolabial fold. She appears to have a gaze preference towards the left. Her speech is effortful, full of grammatical errors, and she has difficulty repeating phrases but is able to follow commands. Deep tendon reflexes are brisk on the right side.

Questions

1. State your provisional diagnosis based on the clinical history.
2. Based on the examination findings, localize the neurological lesion and explain your reasoning.
3. What investigation should be performed first and why?
4. If the investigation given above is reported as normal, how would you proceed to manage this patient in the immediate period?

Discussion

1. State your provisional diagnosis based on the clinical history.

The occurrence of a sudden onset focal neurological deficit is strongly suggestive of an acute cerebrovascular accident (CVA) such as an ischaemic or haemorrhagic stroke. A focal neurological deficit comprises a set of signs or symptoms that can be localized to a specific anatomical area within the central nervous system.

Intracerebral strokes typically result in a loss of function depending on the area involved, causing weakness, numbness, loss of coordination, visual defects or cognitive deficits. Symptoms occur abruptly and are usually maximal within minutes of onset. While ischaemic strokes occur more frequently than haemorrhagic strokes, there are no reliable indicators to distinguish between the two based on the patient's history and presentation alone. Associated symptoms such as headache, seizures or disturbance of consciousness are non-specific and may be a feature of both ischaemic and haemorrhagic strokes.

2. Based on the examination findings, localize the neurological lesion and explain your reasoning.

The patient has a right hemiplegia, expressive dysphasia and left gaze preference. This localizes to the left frontal lobe of the brain.

A more detailed explanation of the relevant functional neuroanatomy is as follows:

The corticospinal tract descends ipsilaterally from the primary motor cortex and travels medially through the brainstem before crossing over at the level of the medulla and entering the contralateral spinal cord. Hence, a lesion affecting the left corticospinal tract anywhere above the medulla will give rise to right-sided hemibody weakness.

The upper motor neuron (UMN) component of the facial nerve descends ipsilaterally from the primary motor cortex as part of the corticobulbar tract, travels through the internal capsule, and connects with the facial nucleus in the contralateral pontine tegmentum. The facial nucleus and the fascicle of the facial nerve make up the lower motor neuron (LMN) component. A lesion of the facial nerve at the nucleus or fascicle will cause a classic 'LMN pattern' comprising of weakness of the ipsilateral upper and lower portions of the face. In contrast, a lesion of the UMN component of the facial nerve (corticobulbar tract) will cause weakness of

only the lower portion of the contralateral face while sparing the forehead and eyelid, giving the classic 'UMN pattern' as seen in the patient. This distinction is seen because the upper portion of the face is innervated by the bilateral corticobulbar tracts.

The patient has Broca's aphasia, a form of non-fluent aphasia. There is loss of grammar (agrammatical speech) and speech can appear effortful. Typically, repetition is also impaired but comprehension is intact. This localizes to Broca's area located in the inferior frontal lobe of the dominant hemisphere. For the vast majority of right-handed individuals, the left cerebral hemisphere is dominant.

Gaze preference, also called gaze deviation, is an involuntary deviation of the eyes horizontally due to a lesion affecting the frontal eye field (FEF) of the frontal cortex which normally communicates with the brainstem to help generate horizontal saccadic eye movements. A lesion of the left FEF will cause involuntary deviation of the eyes towards the left. Because the brainstem nuclei and their efferent pathways remain intact, there is no ophthalmoparesis and the Doll's eye reflex is preserved. Gaze preference must be differentiated from a horizontal gaze palsy caused by a lesion of the sixth cranial nerve nucleus. It is also sometimes misinterpreted as visual neglect caused by a non-dominant parietal lobe lesion. A concomitant homonymous hemianopia may be present in patients with larger lesions extending to the optic radiation, and this can make it difficult to detect gaze preference.

In this patient, you would expect to find a lesion affecting the left frontal cortex. This region is perfused by the left middle cerebral artery, specifically the anterior-superior M1 division of the middle cerebral artery (MCA) [drawing].

3. What investigation should be performed first and why?

Plain CT imaging of the brain is usually the first diagnostic investigation of choice in all patients with suspected stroke. Ischaemic brain tissue appears darker (more hypodense) than its surrounding tissues, whereas blood from an acute haemorrhage appears bright (hyperdense) on plain CT imaging, which distinguishes ischaemic strokes from haemorrhagic strokes (Figure 1).

Occasionally, an acute intraluminal thrombus may be visible on plain CT imaging as a focal hyperdensity within a large vessel, the earliest visible sign of infarction. This is most often observed in the MCA, referred to as the 'hyperdense MCA sign'.

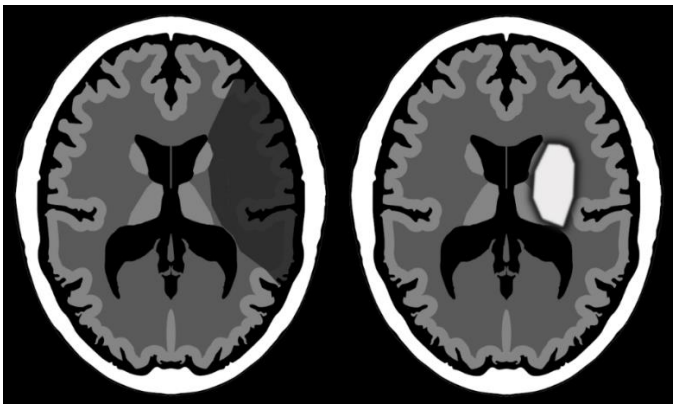

Figure 1. Diagrammatic representation of the CT appearance in ischaemic stroke (left) and acute haemorrhagic stroke (right)

4. **If the investigation given above is reported as normal, how would you proceed to manage this patient in the immediate period?**

Early ischaemic strokes, especially those smaller in volume or located in the posterior fossa, are not often easily visible on initial plain CT imaging of the brain. Thus, the CT scan may occasionally appear normal at the time of presentation. However, plain CT imaging is highly sensitive for detecting and excluding acute haemorrhage, which is an important differential. In most cases, a normal CT brain scan along with a history that is strongly suggestive of CVA are sufficient to support the diagnosis of acute ischaemic stroke, as in the case of this patient. Based on the clinical picture so far, this patient has a diagnosis of a left MCA stroke and should be managed according to local stroke management protocols.

Reperfusion therapy using intravenous tissue plasminogen activator (rtPA) or mechanical thrombectomy is the mainstay of hyperacute stroke treatment. The aim of reperfusion therapy is to salvage the ischaemic penumbra, which is at risk of becoming infarcted, surrounding the infarcted core, thus limiting the neurological deficit and reducing mortality and morbidity. Current guidelines recommend that rtPA is administered within 3 to 4.5 hours of stroke onset in eligible patients, while for mechanical thrombectomy the time window for intervention is 6 to 8 hours. For patients with suspected large vessel occlusion such as the case of this lady with

MCA stroke, CT angiography can help identify a possible target for mechanical thrombectomy.

Unfortunately, the patient in this case awoke from sleep with symptoms of a stroke ('wake-up stroke'), thus posing a therapeutic challenge as the exact time of onset of her stroke is unknown. When patients present with an unknown time of onset (but within 24 hours of onset), it is possible to identify those who may still benefit from reperfusion therapy using advanced imaging techniques. The presence of 'mismatch' between MRI diffusion and FLAIR (fluid-attenuated inversion recovery) imaging suggests that their stroke would have likely occurred within the 3-to-4.5-hour window and hence, the patient can be considered for reperfusion therapy.

All ischaemic stroke patients should have secondary prevention measures initiated as soon as it is feasible, comprising of antithrombotic therapy (typically an antiplatelet agent such as aspirin or clopidogrel) and control of modifiable atherosclerotic risk factors such as hypertension, diabetes mellitus, dyslipidaemia and smoking. It is also important to identify the possible stroke mechanism, particularly atrial fibrillation and carotid artery stenosis, in which prompt intervention will prevent stroke recurrence. All stroke patients should ideally be managed in a dedicated stroke unit for acute care and rehabilitation. Stroke-related complications such as infection, deep venous thrombosis, pressure ulcers and nutrition-related issues should be anticipated and prevented through appropriate interventions.

The key principles of stroke management are summarised below:

- Reperfusion therapy to minimise residual disability
- Secondary prevention with antithrombotic therapy and control of risk factors
- Prevention and treatment of stroke-related complications
- Facilitate recovery and improve neurological function through rehabilitation

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Case 7

A 40-year-old woman presents with worsening tingling and numbness, followed by weakness of her distal upper and lower limbs over the past 5 days. Symptoms started in her feet, progressing to her legs, then to upper limbs. For the past 24 hours she has been experiencing double vision in all directions of gaze, facial weakness, dysarthria and difficulty swallowing. She also complains of urinary difficulties. She is now unable to walk unaided.

About 2 weeks ago she suffered from 'a bout of flu' and had to stay in bed for 3 days, eventually recovering.

Examination reveals absent deep tendon reflexes and flexor plantar responses bilaterally. Limb tone is flaccid. In her lower limbs, there is weakness of bilateral ankle dorsiflexion and plantarflexion, and bilateral knee flexion and extension (MRC 3). In her upper limbs, there is weakness of her distal flexors and extensors (MRC 4). In addition, there is loss of sensation to pinprick below her knees and elbows, while vibration and joint position sense are lost in her fingers and toes. Cranial nerve examination reveals paresis of the third and sixth cranial nerves bilaterally, facial diplegia, and tongue weakness.

Auscultation of her chest is unremarkable and her oxygen saturation on pulse oximetry is 97% (on air), however, she has a weak cough. The rest of her systemic examination is unremarkable.

A non-contrast MRI scan of her brain and whole spine is normal. Routine nerve conduction studies performed on the day of admission showed normal conduction velocities. Her cerebrospinal fluid was acellular but had an elevated protein of 700 mg/L.

Questions

1. Where is the neurological lesion and what are the differential diagnoses?
2. What are the clinical features in the scenario that support your diagnosis?
3. What are the main pitfalls in the diagnosis and management of this condition?

Discussion

1. Where is the neurological lesion and what are the differential diagnoses?

The patient presents with tetraparesis and multiple cranial neuropathies. This immediately raises the suspicion of a brainstem lesion. However, the lack of other features to suggest a central nervous system disorder (increased limb tone, hyper-reflexia and extensor plantar response), points towards a disorder of the peripheral nervous system. The absence of deep tendon reflexes and the pattern of sensory loss in a 'glove and stocking' distribution are suggestive of a peripheral neuropathy. Weak cough in this patient suggests involvement of respiratory muscles, particularly the expiratory muscles.

The relatively rapid evolution of the patient's peripheral neuropathy raises the possibility of Guillain-Barré syndrome (GBS), vasculitic neuropathy, or a toxic neuropathy. It is uncommon for peripheral neuropathies to cause respiratory insufficiency except for GBS. Vasculitic neuropathies tend to be painful and there are often other signs of a systemic vasculitic disorder. There is nothing in the patient's history or examination findings to suggest exposure to drugs or toxins such as organophosphates which could cause a neuropathy. Another cause of acute flaccid paralysis with cranial nerve involvement to consider is botulism, however the presence of prominent sensory signs and symptoms in this patient and lack of exposure to botulinum toxin would make this disorder of the neuromuscular junction unlikely.

2. What are the clinical features in the scenario that support your diagnosis?

This patient most likely has GBS. The diagnosis of GBS is made clinically based on the history, examination findings and supportive investigative findings. The most common form of GBS is acute demyelinating inflammatory polyneuropathy (AIDP) where the pathology is primarily demyelination affecting not only the motor and sensory peripheral nerves, but also the cranial nerves, nerve roots, dorsal root ganglia, and autonomic ganglia. Also known as classic GBS, this presents as a rapidly progressive, symmetrical sensorimotor polyardiculoneuropathy. Distal paraesthesia,

often with numbness and sometimes pain, are usually early features. This is followed soon after by ascending flaccid limb weakness which may involve the respiratory muscles and cranial nerves. AIDP is a monophasic illness, with symptom progression observed up to 4 weeks from onset.

Like the patient in this scenario who had a bout of 'flu', there is often a history of antecedent infection which supports the theory of molecular mimicry in the pathogenesis of this condition. Several organisms have been implicated, including *Campylobacter jejuni*, Epstein-Barr virus and Cytomegalovirus, although in most cases the offending organism is never actually identified.

The following investigative findings support the diagnosis of GBS:

- Neuroimaging

  Neuroimaging is most often performed to exclude an alternative diagnosis, where indicated. MR imaging of the brain and spinal cord is normal in GBS, however, there may be visible enhancement of the nerve roots with administration of IV contrast.

- Neurophysiological testing

  The characteristic abnormalities on nerve conduction studies depend on the timing of the test. Findings include prolonged F wave latencies in the earliest stages, followed by prolonged distal latencies, conduction blocks and temporal dispersion. Reduced conduction velocities characteristic of demyelination appear later, around the third or fourth week. Reduced compound muscle action potential (CMAP) amplitudes are indicative of axonal loss and carry a poorer prognosis, as seen with some variants of GBS. As with in the case above, the timing of nerve conduction studies and parameters measured can affect the diagnostic yield, and neurophysiological testing should be repeated in this patient in about 3 weeks' time.

- Cerebrospinal fluid

The presence of elevated cerebrospinal fluid protein (above 550 mg/L) with a normal white blood cell count (albuminocytologic dissociation) is characteristic, as seen in this case.

- Autoantibody testing

Autoantibodies to gangliosides, myelin components and other neuronal structures are measurable in some patients with GBS and are associated with specific variants as described in the next section.

3. What are the main pitfalls in the diagnosis and management of this condition?

- Clinical heterogeneity of Guillain-Barré syndrome

While the most common form of GBS is AIDP, there is an increasing number of variants or subtypes with differing clinical presentations being recognized, which can make the diagnosis challenging. Some examples of these variants are shown in table 1.

Table 1. Variants of Guillain- Barré syndrome and their key characteristic findings

Variant or subtype	Characteristic findings
Miller-Fisher syndrome	Ophthalmoplegia, ataxia and areflexia. Associated with anti-GQ1b antibodies.
Acute motor axonal neuropathy (AMAN)	Weakness only, with preserved sensation and tendon reflexes. Axonal involvement of motor nerves on neurophysiological testing. May be associated with *C. jejuni* infection. Associated with Anti-GM1, GD1a, GD1b antibodies.
Acute motor and sensory axonal neuropathy (AMSAN)	As with AMAN, but more severe and with sensory involvement. Marked axonal loss on neurophysiological testing.

Bickerstaff encephalitis	Altered sensorium, ophthalmoplegia, oropharyngeal weakness, and ophthalmoplegia. MRI brain may be normal or show high T2 signal in brainstem. May be associated with anti-GQ1b antibodies.
Pharyngeal-cervical-brachial variant	Predominantly neck, upper limb, pharyngeal and facial weakness.
Paraparetic variant	Lower limb weakness with sparing of the upper limbs.
Pandysautonomia	Postural hypotension, gastroparesis, urinary dysfunction, erectile dysfunction. Tendon reflexes may be preserved. Normal nerve conduction studies.

- Respiratory failure and autonomic dysfunction

Evaluation and monitoring of the patient with GBS should include respiratory function assessment at regular intervals using clinical examination and bedside spirometry. Specific features of respiratory failure due to neuromuscular weakness are described in Table 2 below.

Table 2. Respiratory muscle weakness and associated clinical features.

Site of neuromuscular weakness	Clinical features
Bulbar muscles	Aspiration, drooling, dysphonia, post-prandial coughing, dysarthria
Diaphragm/inspiratory muscles	Orthopnoea, difficulty speaking in complete sentences, morning headaches/sleepiness, use of accessory muscles, shallow breathing.
Expiratory muscles	Weak cough

Elective intubation should be considered in those with impending respiratory failure as dictated by the presence of significant bulbar dysfunction with difficulty clearing

secretions, deteriorating forced vital capacity (FVC) on bedside spirometry, poor blood gas parameters or signs of respiratory distress.

Autonomic dysfunction commonly occurs in GBS, which may manifest as malignant hypertension, hypotension, cardiac arrhythmias, increased sweating, and bladder or bowel dysfunction. Stringent monitoring including cardiac monitoring is recommended.

- Indications for treatment and response to treatment

Immune-based therapies comprising of intravenous immunoglobulin (IVIg) or plasma exchange reduce recovery time and improve prognosis in GBS. Because these treatments carry significant risks of adverse effects, treatment is normally reserved for patients who are unable to ambulate or have rapidly progressive weakness. Although both treatments are equally effective, IVIg is more widely available and easier to administer. Occasionally patients may demonstrate fluctuations and even deterioration after a brief period of improvement following immunotherapy. Most patients recover to full ambulation within 1 year. However, incomplete recovery of weakness is common in patients with axonal involvement. Contrary to most immune-mediated diseases, corticosteroids do not have clinical benefit in GBS.

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Case 8

A 45-year-old lady visits her family doctor with the problem of recurrent upper abdominal discomfort which has lasted more than 6 months. She often purchases some over the counter medicine which brings relief, but the discomfort has recurred in the last 4 weeks. There is no vomiting, but she feels lethargic. She has never been investigated before for this problem and there is no significant past medical history.

Presenting problem: Dyspepsia.

- Dyspepsia is one of the most common medical problems in the primary care setting. Up to 20-30% of people in the community visit their family doctor for dyspepsia.
- Dyspepsia is defined as recurrent, non-specific upper abdomen discomfort. Patients may describe it as fullness, bloating, pain, indigestion, burning etc.
- There are many underlying aetiologies leading to dyspeptic symptoms such as peptic ulcer disease, gastro-oesophageal reflux disease, hepatobiliary disease and gastrointestinal tract malignancy.

<u>Approach to the problem</u>:

1. Identify RED-FLAGS in dyspepsia through history, physical examination and investigations.
2. Identify patients who require upper gastrointestinal (GI) endoscopy/referral.
3. Manage associated problems such as *Helicobacter pylori*, ulcer disease or associated risk factors.
4. Screen for GI malignancy.

Questions

1. What are the important aspects of history that should be elicited in this patient?
2. What are the important red flags that need to be elicited in the medical history?

3. What is the significance of red flags?

4. What are the important physical examinations that need to be performed in this patient?

5. What are the important investigations that need to be performed in the assessment of this patient?

Discussion

1. **What are the important aspects of history that should be elicited in this patient?**

History to be obtained:

A. Dyspeptic symptom

- Onset: How did it start? Sudden acute, or insidious over time?
- Duration: How long has it been there? What is the duration of each episode?
- Aggravating / relieving factor:

  Discomfort aggravated by food: possibility of gastric ulcer.

  Discomfort aggravated by fatty food: possibility of biliary colic.

  Discomfort aggravated by bending over or lying down, relieved by sitting up: gastro-esophageal reflux disease (GERD)

  Discomfort relieved by antacids: gastritis , GERD
- Character/nature of the discomfort:
  - Burning pain: gastritis, peptic ulcer disease, GERD
  - Colicky pain: biliary colic
  - Sharp burning pain: pancreatitis
  - Sour belches (acid brash): GERD
- Radiating pain
  - To back: pancreatitis
  - To right hypochondrium: hepatobiliary disease
  - To the chest and throat: GERD

B. Associated upper GIT symptoms

- Nausea, protracted vomiting
- Haematemesis: peptic ulcer, malignancy
- Heartburn, regurgitation, sour taste: GERD
- Dysphagia: oesophageal cancer, stricture
- Early satiety : stomach cancer, pancreatic cancer
- Abdominal distension: hepatobiliary disease

- Jaundice, dark coloured urine: hepatobiliary disease

C. Associated lower GIT symptoms

- Malaena (blood from upper GI tract)
- Diarrhoea / constipation: colorectal cancer, inflammatory bowel disease
- Steatorrhoea: pale coloured stools
- Fresh PR bleed

D. Associated constitutional symptoms

- Loss of appetite, loss of weight: constitutional symptoms of malignancy
- Fever: constitutional symptoms of malignancy
- Symptoms of anaemia

E. Past history

- Past history of peptic ulcer disease, GERD or biliary disease & treatment
- Previous history of upper GI endoscopy and any significant findings.
- Previous history of *H. pylori* status.

F. Medical illness

- Hypertension, diabetes mellitus
- Hepatobiliary disease
- Chronic joint pain (especially ingestion of non-steroidal anti-inflammatory drugs - NSAIDs)
- Hypo/hyperthyroidism (may present with GI upset)

G. Family history

- Family history of peptic ulcer and GI malignancy

H. Drug history

- Chronic NSAIDS usage (NSAIDs for joint pain, or aspirin for ischemic heart disease and stroke)
- Steroids
- Traditional medicine

I. Social / Personal / Occupational history

- Smoking: peptic ulcer disease, cancer
- Alcohol intake: liver disease, pancreatitis
- Stress: peptic ulcer disease.

2. What are the important red flags that need to be elicited in the history taking?

3. What is the significance of red flags?

The following are RED-FLAGS in history:

- Haematemesis and malaena.
- Protracted vomiting.
- Loss of weight and loss of appetite.
- Early satiety or fullness.
- Jaundice and anaemia
- New onset of dyspepsia above the age of 50

Note: Significant past history of gastric ulcer but unknown treatment status must be treated with caution! There is a risk of malignant transformation in previous non-healed gastric ulcer.

4. What are the important physical examinations that need to be performed in this patient?

Physical Examination: The major aim is to examine for clinical features suggestive of underlying GI pathology.

- General examination: vital signs; blood pressure, pulse, temperature.
  (Important to see that the patient is not in shock, which can happen in bleeding peptic ulcer)
- Examine for pallor: indicates chronic blood loss (especially in peptic ulcer disease or malignancy)
- Examine for signs of chronic anaemia: koilonychia, angular stomatitis, atrophic glossitis
- Examine for jaundice and associated stigmata of chronic liver disease: for hepatobiliary cause.
- Examine nutritional status: wasting or malnutrition in malignancy.
- Examine for cervical lymph nodes: GI malignancy (Virchow's node in stomach or pancreatic cancer)
- Examine abdomen: hepatosplenomegaly or any palpable abdominal mass caused by tumour or intrabdominal lymphadenopathy indicates GI malignancy.
- Perform a digital per-rectal examination: looking for malaena or fresh blood.

The following are RED-FLAGS in physical examination

- Anaemia
- Muscle wasting
- Palpable lymphnodes (eg; Virchow's nodes)
- Palpable abdominal mass
- Skin changes: Acanthosis nigricans
- PR examination reveals maelena

5. What are the important investigations that need to be performed in the assessment of this patient?

Investigations: The major aims are:

- Ascertain haemoglobin status.
- Investigate according to suspected cause.

1. Basic haematological assessment including haemoglobin and full blood count to confirm anaemia and its severity.

2. If suspected hepatobiliary disease, liver function test is indicated. Further referral for ultrasound of the hepatobiliary system is indicated.

3. Upper GI endoscopy remains as the gold standard test to exclude gastroduodenal ulcers, GERD, and upper gastrointestinal cancers and *H. pylori* infection.

Indications for oesophagogastroduodenoscopy:

- Dyspepsia onset above 50 years of age.
- Unexplained weight loss.
- Protracted vomiting / dysphagia.
- Unexplained early satiety.
- Palpable abdominal mass, Virchow's nodes or acanthosis nigricans.
- Presence of anaemia, malaena or haematemesis.
- Recurrent pain in a previous confirmed peptic ulcer / *H pylori* positive patient.
- Positive stool occult blood.
- Symptoms persist or recur after six to eight weeks of empirical (symptomatic) therapy.
- Strong family history of malignancy.

Special note:

What is functional dyspepsia?

1. Functional dyspepsia only applies if underlying GI pathology has been excluded. This is usually in the young population and the patient should not present with any red-flags. Functional dyspepsia is also known as non-ulcer dyspepsia and may be the diagnosis in up to 60% of patients presenting with dyspepsia.

2. In the community, many patients will subject themselves to taking antacids, which is the treatment for benign gastritis. Many patients with recurrent symptoms of dyspepsia treated as gastritis (without investigations) have peptic ulcer disease or biliary stone.

Take home message:

- Dyspepsia is a common problem encountered in the community.
- Recognising RED-FLAGs in dyspepsia is most crucial.
- GI malignancy must be excluded in recurrent dyspepsia especially above age of 50.
- Screening of *Helicobacter pylori* in dyspeptic patients is important.
- Patients with RED-FLAGs require further evaluation with upper GI endoscopy

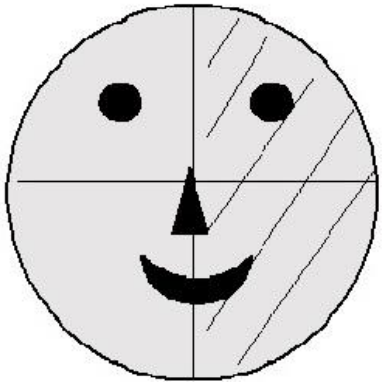
----------End----------

Case 9

A 35-year-old man visits his family doctor with the problem of sudden onset of facial weakness of one day duration. He experiences left sided sagging of the mouth and is unable to close his left eye which is tearing. The right side of his face is not affected. His trunk and limbs are not affected.

Presenting problem: facial nerve palsy, lower motor neuron type.

The clinical presentation is illustrated in Figure 1:



- One sided facial muscle weakness.
- Unable to close the affected eyelid.
- Loss of frontal creases.
- Sagging of angle of mouth
- Loss of nasolabial fold

The associated underlying pathology is unilateral, and is on the same side as that of facial weakness.

Figure 1: Classical features of lower motor neuron facial nerve palsy.

Questions

1. What are the important aspects of history that should be obtained?
2. What are the important aspects of physical examination that should be performed?
3. What are the important investigations for this patient?
4. What is Bell's Palsy?

Discussion

Approach to the patient:

1. **What are the important aspects of history that should be obtained?**

- Onset: acute or subacute?
- Is this first episode or recurrent?
- Any known predisposing factors or underlying medical disorder such as diabetes mellitus or any infection.
- Constitutional symptoms: weight loss, poor appetite if it is associated with underlying chronic infection or malignancy.
- Facial pain or discomfort.
- Tearing: any changes.
- Loss of taste especially anterior 2/3 of the tongue.
- Hearing loss: underling 8 cranial nerve disease (e.g. acoustic neuroma)
- Ear discharge: chronic otitis media (suppurative unsafe type)
- Peri-auricular skin changes: inflammation with vesicles (e.g. herpes zoster)
- Nasal symptoms: blocked nose or recurrent discharge.
- Epistaxis: underlying nasopharyngeal malignancy.
- Trauma: previous history of head and neck injury.
- Previous history of facial surgery.
- Family history of facial palsy.

2. **What are the important aspects of physical examination that should be performed?**

- Inspection: look for the gross abnormalities seen on the face – such as those in Figure 1.
- Examine the external auricular canal for any infection and vesicles.
- Examine the oral cavity and throat to exclude any gross pathology.
- Examine around the mastoid process.
- Examine for parotid swelling.

- Examine for cervical lymphadenopathy.
- Perform neurological examination of the facial nerve: look at frontal area describe the loss of frontal creases and absence of frontalis muscle movement, inability to close the affected eye, loss of nasolabial fold, weakness of the orbicularis oris muscles and sagging angle of the mouth on affected side.
- Compare the left and right side of the face and note any differences.
- May need complete otolaryngological assessment in suspected head and neck malignancy.

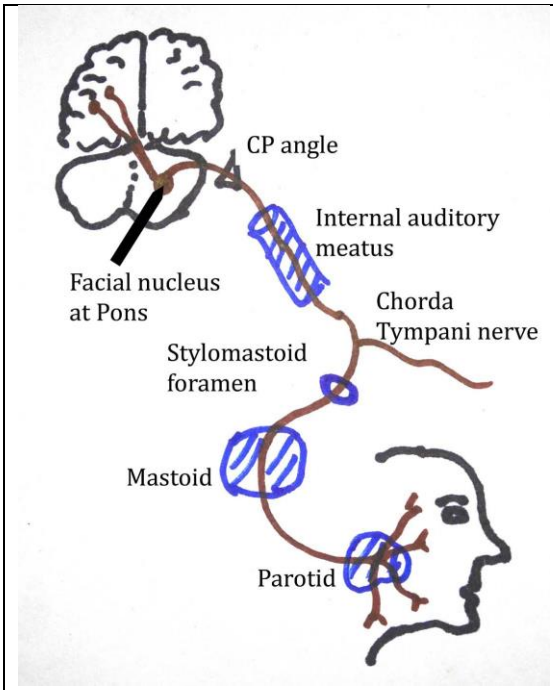
3. What are the important investigations for this patient?

- Pure tone audiometry (PTA) is indicated to exclude underlying acoustic neuroma (tumour of cranial nerve VIII) which is often silent. PTA will reveal unilateral sensory neural deafness.
- Other investigations are according to suspected causes. (Figure 2)
- Fasting blood glucose to exclude diabetes mellitus.
- Causes of facial nerve palsy are closely related to diseases along the anatomical pathway from intracranial to extracranial. (Figure 2)

In order to describe the underlying causes of lower motor neuron facial nerve palsy, it is important to understand applied anatomy of facial nerve pathway.

Figure 2: Applied Anatomy of facial nerve pathway

Diagramatic description of facial nerve pathway	Anatomical pathway of facial tract	Common pathology associated
	• Facial nerve nucleus is situated at pons.	Pontine disease: tumour or hemorrhage
	• Facial nerve travels through	CP angle tumour: acoustic neuroma

	cerebellopontine region. This is the intracranial cisternal portion.	
	• It enters internal auditory meatus. This is the canalicular (meatal) portion.	Middle ear disease: Chronic suppurative otitis media with cholesteatoma
	• Facial nerve exits the skull via stylomastoid foramen.	Mastoid disease: mastoiditis
	• Extra cranial portion end in the parotid gland region supplying the facial muscles.	Parotid tumour
	• Entire tract	Infection e.g.: Herpes zoster, Herpes simplex, Demyelinating diseases, Nerve transection due to trauma

4. What is Bell's Palsy?

Bell's palsy is an acute idiopathic facial nerve palsy resulting in weakness of the facial muscles. It is named after Sir Charles Bell (1774–1842), who first described this condition. It is one of the most common causes of lower motor neuron facial nerve palsy seen in the community. It

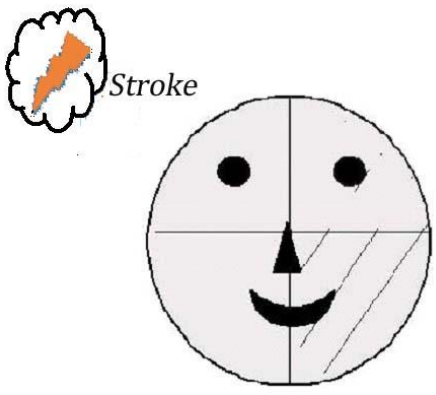
can affect any age group, but peak incidence is around middle age; 40-50 years of age. The symptoms usually last one to two weeks but some patients may have residual weakness up to three months or more. Known risk factors include diabetes mellitus and previous episode of Bell's palsy. The aetiology is unknown, but a study has shown a possible association with herpes simplex infection. Acyclovir with short course prednisolone is found to shorten the course of illness.

Important Clinical Note:

1. It is not correct to diagnose patient with lower motor neuron lesion as Bell's palsy without examining for possible causes, even though Bell's palsy is commonly seen in the community. Known underlying causes affecting the facial nerve along its pathway must be treated. (Figure 2)

2. Supranuclear lesions (i.e lesions above the 7 cranial nerve nucleus) will cause a contralateral upper motor neuron pattern facial weakness. Such lesions may arise from lesions of the facial area of the motor cortex or anywhere along the corticobulbar tract such as the internal capsule. In upper motor neuron facial nerve palsy, only the contralateral angle of the mouth is affected (Figure 3). The forehead and eye lids of both sides are unaffected. Patient can close his eyes. This is due to double innervation of the frontal region and orbicularis oculi.

Figure 3: Classical features of upper motor neuron palsy associated with motor stroke

	• Contra-lateral lower quadrant angle facial muscle weakness. • Contralateral sagging of angle of the mouth • Contralateral loss of nasolabial fold • Both eyes unaffected • Frontal muscles not affected

----------End----------

Case 10

22-year-old Ms R, previously healthy, G2P1 lady at 8-week period of amenorrhoea presents to Emergency Department with lower abdominal pain associated with per vaginal bleeding for 2 days. Per vaginal bleeding is minimal. Pain is in suprapubic area and right iliac fossa, sharp in nature, and radiating to right shoulder tip. Pain score is 8/10. She has associated nausea but denies fever, passage of grape like vesicles or any fleshy clots. On examination, she has mild pallor. Her oral temperature is 37.2 degrees Celsius, blood pressure 110/73 mmHg and pulse rate 102/minute. Abdomen is soft, but mild tenderness is noted at the right iliac fossa; no rebound tenderness and no mass felt.

Questions

1. What are the possible differential diagnoses? Give one most likely provisional diagnosis.
2. How will you support/ confirm the diagnosis? State the expected findings that support/confirm the differential diagnoses you consider.
3. How will you manage this case?

Discussion

1. **What are the possible differential diagnoses? Give one most likely provisional diagnosis.**

In formulating the differential diagnoses and the provisional diagnosis in this patient, the following are the major points you should consider -

- Female at reproductive age group
- Amenorrhoea
- Per vaginal bleeding
- Lower abdominal pain radiating to shoulder tip
- Mild pallor
- Tachycardia
- Tenderness in lower abdomen

Based on the history and examination findings, you should consider

1. Ectopic pregnancy
2. Miscarriage
3. Molar pregnancy
4. Other causes of per vaginal bleeding such as any cervical pathologies, coagulation disorders
5. Other acute abdomen such as acute appendicitis, ovarian cysts accidents etc

However, the patient does not give a history of passing any product of conception (expected in miscarriage) or grape like vesicle passing (expected in molar pregnancy). There is no mass (no uterus larger than date) as seen in molar pregnancy. She has no fever (as in some causes of acute abdomen such as appendicitis). Since Ms R is said to be previously healthy lady, it can be assumed that there is no history suggestive of bleeding diathesis, and there has been no abnormal Pap smear report.

Therefore, the provisional diagnosis is considered to be ectopic pregnancy.

2. **How will you support/ confirm the diagnosis? State the expected findings that support/confirm the differential diagnoses you consider.**

Once you arrived the diagnosis, the next step is to consider how you will support/ confirm the diagnosis and expected findings. The following physical examination and investigations will help you in this task.

1.Vital signs

Expected findings: Signs of shock, pallor

2. Abdominal examination

Expected findings: Abdomen tense, tenderness in lower abdomen (ectopic pregnancy/ miscarriage), uterus larger than date (molar pregnancy)

3. Pelvic examination (Speculum and bimanual examination)

Expected findings: Cervical excitation positive (ectopic pregnancy), cervical os open and product of conception might be seen (miscarriage), uterus larger than dates and grape like vesicles might be seen at cervical os (molar pregnancy)

4. Urine pregnancy test (UPT)

Expected findings: UPT negative - exclude pregnancy/ UPT positive - confirm pregnancy

5. Ultrasound pelvis is the most basic investigation;

Expected findings: empty uterus, extra uterine gestational sac and free fluid (ectopic pregnancy)

6. Full blood count

Expected findings: Anaemia due to bleeding (ectopic pregnancy and miscarriage), raised total WBC count due to possible infection eg. septic miscarriage

In addition, the following are required for the continued care of the patient.

7. Blood grouping, Rhesus status: in case patient needs blood transfusion. If patient is rhesus negative, we need to consider anti D immunoglobulin to prevent rhesus isoimmunisation

8. Serum b-hCG: If patient's vital signs are stable, clinical assessment and radiological assessment are inconclusive of location of pregnancy, patient needs to be admitted to hospital and serum b-hCG monitored every 48 hours.

Serum hCG: repeated 48h later:

- the rate of rise is important
- a rise of ≥66% suggests an intrauterine pregnancy
- a suboptimal rise is suspicious of an ectopic pregnancy.

Further findings on a subsequent assessment of Ms R are given below.

Abdominal examination:

Soft, mild tenderness at right iliac fossa, uterus is not palpable per abdomen

Pelvic examination:

Cervical os is closed, cervical excitation test is positive, uterus is 8 weeks size, minimal blood is noted

Ultrasound pelvis:

The uterus is normal and empty with an endometrial thickness of 4.6mm. The left ovaries are normal while there is a right suspected corpus luteal cyst. There is also a right adnexal mass present measuring about 1.7x1.1 cm. The abdomen has minimal free fluid in the peritoneal cavity

Full blood count

Parameter	Results	Normal value
White Blood Cell	8.80 x10^9/ L	3.5-12.0 x10^9/ L
Haemoglobin	9.4 g/dL	12-16 g/dL
Haematocrit	37.2 %,	37-47%
Platelet	329 x 10^9/ L	150-400 x 10^9/ L

Blood group: O positive

Serum Beta-hCG: 1900 IU/L (Normal <25 IU/L)

3. **How will you manage this case?**

At this point, you should consider right tubal ectopic pregnancy as your definitive diagnosis. Next question will be management of Ms R.

The steps in the immediate management would be

1. Monitor vital signs
2. Assess airway, breathing, and circulation
3. Inform operation theatre, blood bank, and anaesthetist
4. Take the consent for emergency diagnostic laparoscopy and proceed to the procedure

Laparoscopy is performed and below are the findings:

Right tube and ovary identified, noted only blood clots at the fimbrial end and removed clots, no active bleeding noted and tube appears normal. No ectopic pregnancy is identified.

Left tube and ovary identified, both are normal

Liver surface normal

Uterus normal

Minimal hemoperitoneum

No adhesion

Clots and suction products sent for histopathological examination.

In view of negative scope, you need to call your senior to confirm and proceed with suction and curettage (S&C) in case the diagnosis is nots not an ectopic pregnancy but it is a case of miscarriage.

Blood clots removed from right fallopian tube and S&C samples need to be send for histopathological examinations

Now, you need to observe the patient postoperatively and repeat serum Beta-hCG after 48 hours because you are still not sure the location of pregnancy. (Uterus was empty and negative findings on the scope).

Beta-hCG after 48 hours: 550 IU/L (It came down from 1900 IU/L).

Since Ms R is well postoperatively with accepatable haemoglobin level and beta-hCG is in a downward trend, you can discharge the patient and follow up in two weeks.

On follow up, you need to review histopathology result and Beta-hCG level.

Here is Ms R's result in two weeks' follow up. She is now asymptomatic

Beta-hCG: 126 IU/L
Histopathology: Consistent with ectopic pregnancy tissue (Sample from the Right fimbriae-end blood clot)

With these results, you can confirm that it was a right fimbriae pregnancy. During laparoscopy, normal appearance of the tube is likely due to early ectopic pregnancy (Beta-hCG is only 1900 IU/L on initial presentation).

Since Beta-hCG is coming down now and she is asymptomatic, you may discharge her from your gynaecological care but see her in next pregnancy as early as possible, because she has a higher chance of having an ectopic pregnancy again

1. Reference: Collins S, Arulkumaran S, Hayes K, Jackson S, Impey L. Oxford Handbook of Obstetrics and Gynaecology, 3rd ed. Oxford University Press; 2013.

Case 11

You are the houseman on labour ward. In room no. 5, a 36-year-old Para 4 had a normal delivery 35 minutes ago. She is now actively bleeding, and the placenta is still in situ. Her BMI is 36 kg/m^2. Baby boy Apgar score 8/10 and weighs 3.5 kg. You are summoned by the staff nurse as she is concerned about the amount of bleeding.

Questions

1. How would you assess the blood loss?
2. What is your diagnosis?
3. How would you manage her?
4. What are the risk factors for PPH?
5. How would you prevent PPH?

Discussion

1. How would you assess the blood loss?

Identification of the severity of bleeding: Visual estimation of blood loss is inaccurate. Accurate methods like blood collection drape for vaginal delivery (quantification of blood loss immediately after the child's birth, keeping in mind that most of the fluid collected before delivery of the placenta is amniotic fluid and urine), counting the number of saturated pads, weighing of packs and sponges used to absorb blood (1 ml of blood weigh approximately 1 gm) are used to assess estimated blood loss. The clinical signs and symptoms for hypovolemia should be included in the assessment of haemorrhage.
Assessment of vital signs: Blood pressure, pulse rate, respiratory rate and SpO2

Based on blood collection drapes, the estimated blood loss (EBL) is about 1500 ml. She looks pale and her BP is 95/60 mmHg, pulse rate is 110/min, SpO2 99% in room air and RR 16 breaths/min. Her uterus is above the umbilicus with retained placenta with ongoing active vaginal bleeding.

2. What is your diagnosis?

Major primary postpartum haemorrhage (PPH) with retained placenta.

3. How would you manage her?

Initiate major PPH protocol
Communication, resuscitation, monitoring, investigation, and management must go hand-in-hand with multidisciplinary team care.

Communication:

- Call for help, activate emergency buzzer/PPH Code room 5
- Communicate with the patient and her birthing partner - this is important, and clear information of what is happening should be given from the outset.
- Summon multidisciplinary team involving senior members of staff. The consultant obstetrician and consultant anaesthetist should be alerted, and the

blood transfusion laboratory should be informed. One member of the team should be assigned the task of recording events, fluids, drugs, blood and components transfused, and vital signs.

Resuscitation:

- Initiate resuscitation measures- coordinate/delegate following tasks

Assess ABC

- Airway: Check patency, assess conscious level
- Breathing: Administer high concentration of O2 (10-15 L/min) via facemask
- Circulation: Evaluate circulation and institute following measures as necessary
    - Gain IV access, 2 peripheral cannulae, 14 G
    - Commence fluid resuscitation; crystalloid (2 L)/colloid (1.5 L) up to 3.5 L
    - Transfuse blood as soon as possible
    - Transfuse blood components – Packed red blood cells, fresh frozen plasma (FFP), cryoprecipitate, platelet concentrates
    - Correct disseminated intravascular coagulation defect (DIVC)
- Position the patient flat
- Keep patient warm

Monitoring and Investigation:

Perform immediate venipuncture (20 ml) for:

- Cross-match (4 units minimum)
- Full blood count
- Coagulation screen, including fibrinogen
- Renal and liver function for baseline

Continuously monitor pulse, blood pressure and respiratory rate (using oximeter, electrocardiogram and automated blood pressure recording)

Insert Foley catheter to monitor urine output with hourly urometer.

Management:

Examination of patient: Uterus is above umbilicus and retained placenta with ongoing active bleeding

- Transfer urgently to operation theatre (OT)
- Reassess in OT
- Consider O-ve blood transfusion if group-specific blood is not yet available.
- Inform blood bank/haematology (consider transfusion of Platelets / FFP / Cryoprecipitate: on advice of haematologist)
- Anaesthetist to maintain haemodynamic stability and determine appropriate anaesthesia.
- Perform manual removal of placenta (MRP) under general anaesthesia (GA) and aseptic precautions, by inserting the dominant hand into the cavity following the cord. Identify the plane between placenta and uterine wall, separate and remove in toto while placing the other hand on abdomen to prevent uterine inversion.
- Start on prophylactic antibiotics to prevent infection.
- Institute mechanical and pharmacological measures to stimulate uterine contractions to prevent uterine atony due to GA and prolonged third stage.
- Commence rubbing up the fundus and bimanual uterine compression to reduce bleeding.
- Ensure that the bladder is empty (Foley catheter in place) to enhance uterine contraction.
- The following pharmacological interventions are commenced depending on hospital protocol, contraindication, availability and response –
- Administer oxytocin 5 IU by slow intravenous injection (may have to repeat dose).
- Ergometrine 0.5 mg by slow intravenous or intramuscular injection (contraindicated in women with hypertension or heart disease).
- Oxytocin infusion (40 IU in 500 ml isotonic crystalloids at 125 ml/hour) maintained over 4 hours unless fluid restriction is necessary.
- Carboprost (15-methyl prostaglandin F2α) 0.25 mg by intramuscular injection repeated at intervals of not less than 15 minutes to a maximum of eight doses (use with caution in women with asthma)

- Misoprostol 800 micrograms rectally/sublingually.
- If mechanical and pharmacological measures fail to control the haemorrhage, surgical interventions should be initiated sooner than later (Refer Case 12).

Postoperative instructions:

- Monitor at obstetric high dependency unit or Intensive Care Unit (ICU)
- Record parameters on a modified early obstetric warning score (MEOWS) chart
- MEOWS: 0-1hr every 15mins, 1-2 hr every 30mins, hourly for 6 hours
- Hourly urinary output (should be >30 mls)
- Risk assessment for venous thromboembolism
- LMWH if platelet count is normal and no active bleeding (DIVC is corrected).

Documentation:

- Accurate documentation of a delivery with PPH is essential. The team member recording events on the structured proforma, the scribe, is crucial in the management of PPH.
- PPH should be notified through a clinical incident reporting or risk management system.

Debriefing:

- An opportunity to discuss the events surrounding the obstetric haemorrhage should be offered to the woman (possibly with her birthing partner/s) at a mutually convenient time.

4. What are the risk factors for PPH?

- Risk factors for PPH may present antenatally or intrapartum or even in the immediate postpartum period. The causes of PPH are related to abnormalities of one or more of four basic processes – ‘the four Ts’: tone, trauma, tissue and thrombin. The most common cause of PPH is uterine atony.

Uterine tone

- Multiple pregnancy
- Previous PPH
- Fetal macrosomia
- Failure to progress in second stage
- Prolonged third stage of labour
- General anaesthesia

Trauma

- Episiotomy
- Perineal laceration

Tissue

- Retained placenta
- Placenta accreta

Thrombin

- Pre-eclampsia

5. How would you prevent PPH?

1. Recognition of risk factors for PPH and delivery at tertiary care hospital.

   Advise delivery in a hospital with a blood bank on site.

2. Treating antenatal anaemia

   Antenatal anaemia should be investigated and treated appropriately as this may reduce the morbidity associated with PPH.

3. Reducing blood loss at delivery

Prophylactic uterotonics should be routinely offered in the management of the third stage of labour in all women as they reduce the risk of PPH.

- For women without risk factors for PPH delivering vaginally, oxytocin (10 IU IM) is the agent of choice for prophylaxis in the third stage of labour.
- For women delivering by caesarean section, oxytocin (5 IU slow IV) should be used.

- Ergometrine–oxytocin may be used in the absence of hypertension in women at increased risk of haemorrhage as it reduces the risk of minor PPH (500–1000 ml).
- Consider the use of intravenous tranexamic acid (0.5–1.0 g), in addition to oxytocin, at caesarean section to reduce blood loss in women at increased risk of PPH.

Case 12

A 30-year-old P3 has had a major postpartum haemorrhage (PPH) following a normal delivery. Mechanical and pharmacological measures have failed to control the bleeding. Examination has confirmed that there is no retained placental tissue in the uterine cavity and absence of trauma to genital tract.

Questions

1. What are the surgical options to arrest the bleeding?
2. What is your diagnosis?
3. What are the causes of secondary PPH?
4. How would you manage her?

Discussion

1**. What are the surgical options to arrest the bleeding?**

- Intrauterine balloon tamponade is an appropriate first-line 'surgical' intervention where uterine atony is the only or main cause of haemorrhage.
- Conservative surgical interventions may be attempted as second line, depending on available expertise.
    - Haemostatic brace or uterine compression sutures with absorbable sutures (vicryl or catgut).
    - B-Lynch - requires hysterotomy for its insertion and is particularly suitable when the uterus has already been opened for a caesarean section.
    - Modified B-Lynch/Hayman is double vertical compression sutures which do not require hysterotomy.
- Stepwise uterine devascularisation and internal iliac artery ligation
    - Stepwise uterine devascularisation involves successive ligation of (i) one uterine artery, (ii) both uterine arteries, (iii) low uterine arteries, (iv) one ovarian artery and (v) both ovarian arteries.
- Selective arterial occlusion or embolisation by interventional radiologist
- Hysterectomy
    - The decision for hysterectomy should be made by an experienced consultant clinician, preferably after discussion with a second experienced clinician.
    - Subtotal hysterectomy is the operation of choice.

Her bleeding was controlled with uterine balloon tamponade and was discharged home after 3 days with oral haematinics. She presents 5 days later with complaints of heavy bleeding PV and absence of breast milk.

2. What is your diagnosis?

Secondary PPH which is defined as abnormal or excessive bleeding from the birth canal between 24 hours and 12 weeks postnatally.

3. What are the causes of secondary PPH?

Endometritis, retained placental tissue, subinvolution of the placental implantation site, and gestational trophoblastic neoplasia.

4. How would you manage her?

Assess her haemodynamic status, blood loss and patient's concern.

Investigations include high vaginal swab to rule out endometritis, pelvic ultrasound scan to exclude retained placental tissue and color flow Doppler to diagnose pseudoaneurysms and arteriovenous malformations as a cause of secondary PPH.

Treatment:

Combination of clindamycin and gentamicin is an appropriate antibiotic regimen for the treatment of postpartum endometritis (Cochrane review).

Surgical evacuation of retained placental tissue should be undertaken or supervised by an experienced clinician under direct ultrasound guidance because of increased risk of uterine perforation and Asherman's syndrome.

Uterotonics, such as misoprostol and ergometrine, have been recommended in the management of secondary PPH, although evidence to support their use is limited.

Transcatheter arterial embolisation and balloon tamponade can be performed in cases of secondary PPH with ongoing bleeding.

5. What is the cause of absence of breast milk?

Probably Sheehan's syndrome due to pituitary apoplexy, avascular necrosis related to PPH.

Reference:

Mavrides E, Allard S, Chandraharan E, Collins P, Green L, Hunt BJ, Riris S, Thomson AJ on behalf of the Royal College of Obstetricians and Gynaecologists. Prevention and management of postpartum haemorrhage. BJOG 2016;124:e106–e149.

Case 13

Madam WW, a 36-year-old primigravida at EDD + 2 days period of gestation just had a failed vacuum delivery in the second stage of labour. She has been married for 2 years.
You are called to see her by the nurse.

Questions

1. What will you do ? Outline in 10 (Ten) steps.
2. Name FIVE (5) indications for vacuum delivery.
3. Why may a vacuum delivery fail?
4. What is the BEST REMEDY to her situation now ?

Discussion

1. **What will you do? Outline in 10 steps.**

- Call for help. Red Alert.
- Inform Consultant.
- Gain intravenous access with large bore intravenous cannula x 2
- Inform Anaesthetist
- Inform Paediatrician
- Empty the bladder with indwelling catheter.
- Check for fetal heart. Commence CTG Recording.
- Perform abdominal and vaginal examination.
- Send blood for group cross match (GXM)
- Inquire when was the time of the last meal
- Have consent form ready – for Emergency LSCS
- Inform husband / family.
- Keep patient fasting – "Nil By Mouth".

2. **Name FIVE indications for vacuum delivery.**

MATERNAL – Pregnancy induced hypertension, Poor maternal effort, maternal exhaustion, maternal heart disease, prolonged second stage of labour, maternal myasthenia gravis.
FETAL – Foetal distress, meconium seen at os in the 2nd. stage of labour (depends on circumstances)

3. **Why may a vacuum delivery fail?**

- Unrealised cephalo-pelvic disproportion (CPD)
- Big baby
- Os not fully dilated
- Wrong placement of vacuum cup onto fetal head
- Wrong technique of exerting force on the vacuum mechanism
- Bladder not emptied

- Station is still high
- Deflexed fetal head
- Excessive caput and moulding
- Unrecognised concomitant cord prolapse
- Unrecognised compound presentation
- Unrecognised brow presentation or others
- Undiagnosed locked twin - rare.

4. **What is the BEST REMEDY to her situation now?**

The safest – provided vacuum has been properly done and baby fails to descend at all – is an Emergency Caesarean Section by an experienced Senior Doctor in a tertiary centre with Paediatric and Aneasthetic support with direct Consultant involvement.

Madam WW proceeded to an Emergency Caesarean Section. During the LSCS, after delivery of the baby, Madam WW kept on bleeding and bleeding.

5. **Give your comments on possible causes of continued haemorrhage after delivery of the baby.**

Causes of continuing haemorrhage – uterine atony, lateral angle uterine tear, unresolved cervical tear, unresolved vaginal tear, forgotten to suture episiotomy originally done for the vacuum delivery, disseminated intravascular coagulation (DIVC), retained placenta, placenta accreta or increta, pre-existing clotting abnormalities and arterio-venous malformation of uterus.

6. **What steps will you take to save Madam WW?**

Non- Surgical: Correct volume depletion by giving packed red cells. Correct any DIVC with fresh frozen plasma (FFP), cryoprecipitate and platelet concentrates with guidance from the Haematologist or Blood Bank Consultant. Consider Factor 7. Or the US Army coagulation powder (in extreme cases). Good anaesthetic care with adequate oxygenation, adequate analgesia, blood products, blood pressure optimization, inotropic support (if required), correction of DIVC, good venous access.

Surgical: Call consultant. ‘Call for Help’. Surgical procedures in this situation include a combination of bilateral internal iliac artery ligation, repair uterine angle tear, securing placental bed in uterus of bleeders, packing, B – Lynch sutures, haemostatic Bakri Ballon tamponade.

If all else fails – hysterectomy may be necessary. Care must be taken to avoid damage to the ureters and bladder. Double secure haemostasis in view of DIVC (if DIVC is either established or impending). Surgical drains can give early warning of continued bleeding in the postoperative period. But blocked drains can give a misleading message.

Others : Close observation of all other bodily functions is required – renal, cardiac, lungs and maintenance of good oxygenation. Multi-disciplinary management with post-operation HDU / ICU care would be ideal. Blood Bank support needs to be sought. Some may require relaparatomy.

7. **When will a hysterectomy be required**?

Hysterectomy is required if all other methods fail and maternal life is at stake. It should be undertaken before irreversible shock occurs provided Consultant is already in attendance in the operation theatre with Anaesthetist. It is best carried out in a tertiary centre with full blood bank support. In an emergency setting – husband’s consent is best obtained. If husband is unreachable or not present – two consultants’ signature is required – as it is a lifesaving procedure.

8. **How could post-partum haemorrhage be prevented?**

Steps to minimize post-partum haemorrhage and its consequences may be instituted at different levels. (these are listed out systematically).

A. Pre-pregnancy: Correct anaemia. Investigate cause. Treat the cause.
B. Patient risk stratification: KKM Red Book has good stratification system. Identify anaemic patients, previous PPH, grand-multiparity, special conditions – on warfarin, aboriginal, vegetarians, Jehovah Witnesses, herbs, others.
C. During pregnancy: Regular Hb checks, vitamins, deworm, healthy balanced diet

D. During delivery: Expert care, tertiary centre, group, screen and hold (GSH), intravenous line access, full blood count (FBC) predelivery, active management of 3rd. Stage of labour, syntocinon, syntometrine (in the absence of contraindications).
E. After delivery: Adequate nutrition, check placenta for completeness, suture relevant tears, careful examination (not to miss tears), expert care. Full blood count.
F. Labour Room Level: Well trained staff at all levels, labour room drills, equipment ready, equipment updated, 1 to 1 nursing, blood bank support, good laboratory support.
G. Hospital Level: Maintenance of 24-hour Blood Bank services, blood donor services.
H. State Level: Increase employment, Increase well-being of people, safe water, safe Roads
I. National Level: Increase health allocation, increase specialist training, increase number of well-trained doctors, nurses, midwife trained nurses.

Overall : Give Health Priority.

References

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6. Vacuum Extraction failure is associated with a large head circumference. Doron Kabiri et al. J Matern Fetal Neonatal Med 2019 Oct; 32(20): 3325-3330 (epub).

Case 14

Madam B, a 49-year-old Para 5 lady has had no menstrual periods for the past 6 years. She has mild diabetes mellitus (DM). She is a non-smoker.

Three months ago she had a period that lasted 6 days requiring 3 sanitary pads per day for the first 5 days of the period. She was quite surprised and shocked. She went to see a general doctor who told her it is "AUB". There was some mild dysmenorrhea.

She was prescribed progestogen tablets to complete from Day 1 to Day 21 and to repeat for the next 4 cycles. The tablets worked. Her bleeding regained its regularity with normal menstrual flow.

Two weeks ago at her gynaecology follow-up, a pap smear was performed. An ultrasound of the pelvis showed an anteverted uterus that measured 8.3cm x 7.9cm x 6.5cm. She was told that her cervix was bulky but smooth with no fungating mass. Her HBA_{1C} was 5.5%.

Questions

1. Outline her clinical problems.
2. Rebut the statement - her DM is well controlled.
3. What further history is required?
4. State the physical examination that is important.
5. Outline FIVE important investigations, in order of importance, with justifications.
6. Her Pap smear shows squamous cell carcinoma. Outline subsequent management.
7. Colposcopic directed punch biopsy shows CIN 1 changes. Comment.
8. What are the indications for cone biopsy in this patient ?
9. Cone biopsy was performed in this patient. The histopathology report shows: acute on chronic cervicitis with areas of ulceration. What is your comment ?
10. State Five POSSIBLE diagnoses for above.

Discussion

1. Outline her clinical problems.

Early menopause

Postmenopausal bleeding

Mild diabetes mellitus – HBA_1c is 5.5% which suggests good control.

Alleged AUB (bbnormal uterine bleeding) - but does not fit criteria of AUB.

Uterus is enlarged - menopausal uterus should be smaller

Cervix is bulky

Progestogen tablets given cyclically without a proper diagnosis or pap smear results or histopathological confirmation of normality.

2. Rebut the statement - her DM is well controlled.

Her HBA_1c suggests that her 3-month blood glucose is within acceptable range. However, this has to be supported by serial blood sugars, clinical evidence of no end organ damage like - normal fundoscopy, no glove and stocking paraesthesia, no microalbuminuria, normal renal function and no evidence of small arterial disease i.e. evidence of ischaemic heart disease or skin changes attributable to diabetes mellitus.

3. What further history is required?

Full menstrual history, full obstetric and gynaecological history - full medical, surgical, medication, allergy, family history, systemic review and any history of inter menstrual bleeding or postcoital bleeding. Loss of weight. Loss of appetite. Cough. Bone pains. Pap smear history.

4. State the physical examination that is important.

- Full general examination including height, weight, lymph nodes, blood pressure and pulse rate
- Examination of the thyroid gland
- Examination of cardiovascular and respiratory systems
- Neurological examination including fundoscopy

- Abdominal examination including inguinal lymph nodes, full pelvic, rectal and rectovaginal examination with speculum examination

5. Outline important investigations, in order of importance, with reasons.

- Full blood count with differential counts - to ensure no anaemia
- Renal profile - to check for good renal function
- Liver Function Tests - to ensure liver function is good
- Random Blood Sugar - to check her blood glucose status
- Urine Full Examination and Microscopic Examination - to rule out Urine infection or proteinuria or state of glycosuria
- Serum Ca 125 - if elevated might be a cause for concern but it can be elevated in some benign or malignant conditions.
- Ultrasound pelvis - endometrial thickness if high (more than 4mm in postmenopausal women) - suspect endometrial abnormality/hyperplasia/mitotic lesions possible
- Others - Electrocardiography, Chest radiography, CT scan of abdomen, pelvis and thorax.

6. Her Pap smear shows squamous cell carcinoma. Outline subsequent management.

Inform consultant. A biopsy of the cervix is needed for histopathological confirmation. Pap smear is a cytological result that is subject to intra-observer and inter-observer variation. Cervical cancer staging with cystoscopy in the operation theatre by a trained Gynae-Oncologist is needed. Further management depends on FIGO stage of cancer. GENERALLY, if patient is fit and she is FIGO Stage 2a and below - radical surgery is possible with pelvic lymph node dissection. If FIGO Stage 2b and above – she will need radiation or chemoradiotherapy with individualized care.

7. Colposcopic directed punch biopsy shows CIN 1 changes. Comment.

Need to check if correct specimens are reported on. There is cytological (squamous cell carcinoma) and histopathological discrepancy (CIN 1). A cone biopsy or large loop excision of the transformation zone (LLETZ) may be required to obtain more tissue for confirmation. This should be discussed with consultant, patient and family.

8. What are the indications for cone biopsy in this patient?

Obligatory cone biopsy as there is a discrepancy between pap smear and colposcopic directed cervical punch biopsy.

9. Cone biopsy was performed in this patient. The histopathology report shows: acute on chronic cervicitis with areas of ulceration. What is your comment?

Pap smear results (squamous cell carcinoma) review at Tumour Board and Cytopathologists together with entire history, findings and all tests including the cone biopsy.

10. List Five POSSIBLE diagnoses in this lady.

1. 49 Para 5.
2. Diabetes mellitus.
3. Premature menopause.
4. Post-menopausal bleeding.
5. Cytological diagnosis of squamous cell carcinoma of the cervix but histopathology does not support it.
6. Uterus bulky; cone biopsy will be essential (not optional).

References :

1. A Review of Cervical Cancer : Incidence and Disparities. Ama Buskwofie et al. J Natl Med Assoc. 2020 Apr;112(2) : 229-232 (epub)
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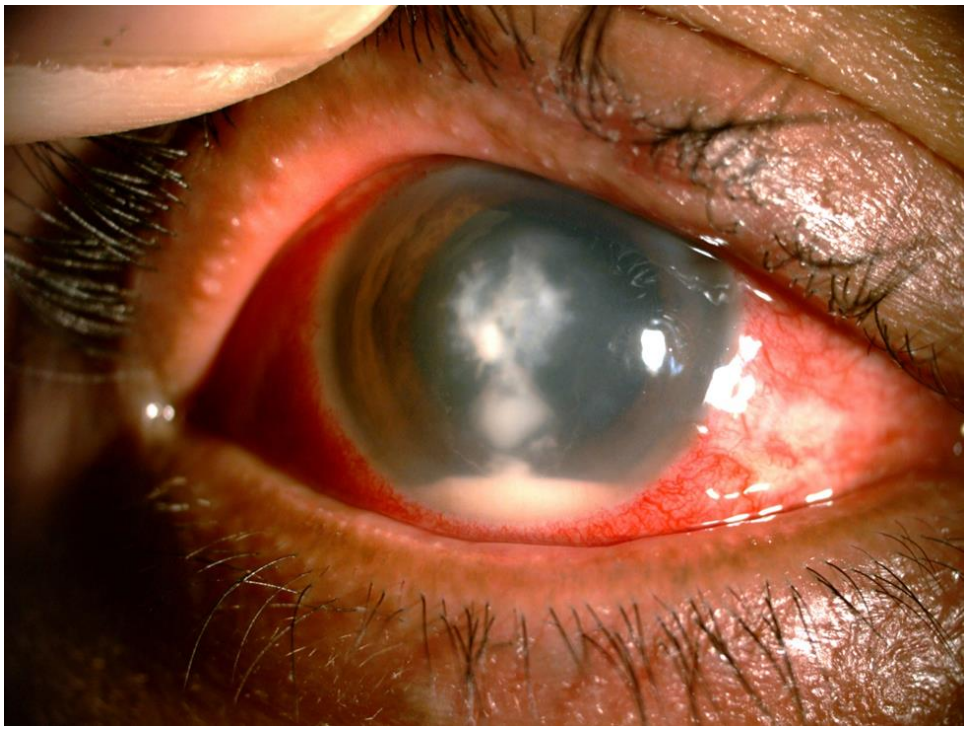
4. Current Treatment for Cervical Cancer : An Update. Sombeer Sharma et al. Anticancer Agents Med Chem. 2020;20(15):1768-1779
5. Quality and Liability issues with the Papanicolaou smear : the problem of definition of errors and false negative smears. DD Davey. Arch Pathol Lab Med 1997 Mar;121(3):267-9

Case 15

Mr. TH, a 55-year-old rubber tapper, presents with redness, watering and blurring of vision in his right eye for the past 10 days. He accidentally injured his eye while working in the plantation. On examination there is a greyish white, dry elevated lesion with feathery margins on the cornea and a hypopyon.

Slit lamp photograph of the affected eye is presented below:



Questions

1. State the possible differential diagnosis
2. What other information may be helpful in making a provisional diagnosis? Explain how the information is helpful
3. State the provisional diagnosis with justification
4. Name the investigations that will help confirm the provisional diagnosis
5. What are the important principles in the medical management of this condition?

Discussion

1. **State the possible differential diagnoses**

Important considerations in this scenario include:

- Clinical presentation - an “Acute Red Eye”
- A “rubber tapper”- with trauma to the eye (possibly by a vegetative matter)
- Characteristics of the corneal lesion and the presence of a hypopyon

The important causes of acute red eye that must be considered in the differential diagnosis include:

- Acute conjunctivitis
- Acute anterior uveitis
- Corneal ulcer/ keratitis.
- Acute congestive glaucoma
- Endophthalmitis
- Episcleritis/ scleritis

2. **What other information may be helpful in making a provisional diagnosis? Explain how the information is helpful.**

   The lesion on the cornea and the presenting complaints as well as the history of trauma to the eye strongly suggest inflammatory/ infective affection of the cornea (corneal ulcer/ keratitis).

To rule out other possible conditions it is prudent to ask for associated symptoms like nausea, vomiting, headache (acute congestive glaucoma), presence and nature of discharge/ matting of lashes (acute conjunctivitis), marked photophobia (anterior uveitis) and associated systemic conditions – diabetes, tuberculosis, autoimmune disorders etc. (anterior uveitis/ scleritis) as

well as past ocular history of surgery/trauma (endophthalmitis) or previous attacks/ colored halos (acute congestive glaucoma).

With the presence of a corneal lesion, it is equally prudent to identify risk factors like use of contact lenses (especially extended wear type), contaminated ocular medications or contact lens solutions, indiscrete use of topical steroids, tear deficiencies, structural alterations in the lids and diseases of the ocular adnexa.

Another important aspect in a patient presenting with a “Red Eye” is to clinically determine the type of congestion in the conjunctiva that is responsible for the red eye appearance.

Features that help to differentiate between conjunctival congestion and ciliary congestion are listed in the table below.

Feature	Superficial (Conjunctival congestion)	Deep (Ciliary congestion)
Site	More marked in the fornices and fade towards the limbus	More marked around the limbus and fade towards the fornix
Colour	Bright red	Purple or dull red
Arrangement of vessels	Superficial and branching	Deep and radiating from limbus
Plane of the vessels	In the conjunctiva	Below the conjunctiva
On moving conjunctiva	Congested vessels also move	Congested vessels do not move
On mechanically squeezing out the blood vessels (direction of blood flow)	Vessels fill rapidly from fornix towards limbus (centripetal)	Vessels fill slowly from limbus towards fornices (centrifugal)
Blanching, i.e., on putting one drop of 1 in 10000 adrenaline	Vessels immediately blanch	Limited blanching

Vessels involved	Posterior conjunctival	Anterior ciliary
Common causes	Acute conjunctivitis	Acute iridocyclitis, Keratitis

3. State the provisional diagnosis with justification

Provisional Diagnosis: fungal corneal ulcer/ fungal keratitis

Justification:

- History of trauma by a vegetative matter in this patient
- Delayed presentation
- Less symptoms; more signs
- Red eye
- Lesion on the cornea - A fungal corneal ulcer is typically seen as a grayish- yellow, dry looking, elevated lesion affecting the paracentral cornea with feathery margins. Satellite lesions may be present. Often associated with collection of pus in the anterior chamber (hypopyon).

4. Name the investigations that will help confirm the provisional diagnosis

To confirm the diagnosis, perform:

- Fluorescein staining (to confirm the presence of an epithelial defect)
- Corneal scraping for culture and sensitivity – culture on Sabouraud Dextrose Agar (to confirm fungal aetiology)
- Confocal microscopy

5. What are the important principles in the medical management of this condition?

Finally, you should understand the important principles in the medical management of this condition.

These include:

- Protection/ cleanliness of the eye.

- Dark glasses.
- Topical appropriate antibiotic/ antifungal therapy.
- Topical cycloplegics (atropine).
- Topical intra ocular pressure lowering agents.
- Topical lubricants/ artificial tears.

Continuous Learning Issues:

- Evidence based information on common pathogens responsible for infective keratitis (local and global variations)
- Evidence based information on protocols for medical management of infective keratitis (local and global variations)

Case 16

Mrs. AB, a 65-year-old housewife presents with sudden diminution of vision. Early morning, she noticed that she was unable to see clearly with her right eye. Externally the eye appears normal and there are no other ocular symptoms. She has had diabetes mellitus for the past 22 years.

Questions

1. What are the possible differential diagnoses to consider?
2. What further history, physical examination and investigations would help arrive at a diagnosis?
3. State the provisional diagnosis with justification
4. What are the important principles in the management of this condition?

Discussion

1. **What are the possible differential diagnoses to consider?**

Important considerations in this scenario include:

- Clinical presentation - a “sudden painless loss of vision”
- No other accompanying symptoms
- Normal external appearance of the affected eye

The important causes of sudden painless diminution of vision include:

- Retinal detachment
- Vitreous hemorrhage
- Central retinal artery occlusion.
- Central retinal vein thrombosis.

2. **What further history, physical examination and investigations would help arrive at a diagnosis**

To rule out important conditions it is prudent to ask for:

- Preceding attack of transient loss of vision (amaurosis fugax) – suggestive of vascular accidents.
- Associated symptoms like floaters/ flashes – predisposing to retinal tear / retinal detachment.
- High/ progressive myopia (Pathological myopia) – predisposing to retinal tear / retinal detachment.
- Associated systemic conditions – hypertension, hypercholesterolemia.
- Duration and type of diabetes; glycaemic control.
- Other risk factors for central retinal artery occlusion like endocarditis, atrial myxoma, inflammatory diseases of the blood vessels, and predisposition to forming blood clots.

- Systemic coagulation disorders and blood dyscrasia such as leukemia and thrombocytopenia, use of anticoagulation and antiplatelet agents- increase the risk of vitreous hemorrhage.

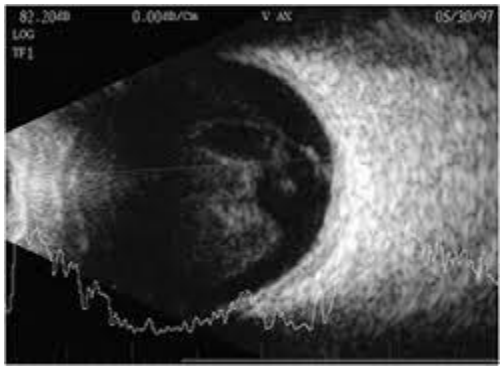
The next important step is to perform relevant examination/ investigations to reach a provisional diagnosis and determine the cause.

Points to consider:

The following investigations will help in arriving at a diagnosis.

- Distant direct ophthalmoscopy (To see the red reflex)
- Direct/ indirect ophthalmoscopy
- B-mode ultrasonography
- Fundus fluorescein angiography

On examination there is absence of the red reflex in the right eye. B-Scan ultrasonography picture is given below:



Provisional diagnosis: vitreous hemorrhage. (as a sequel of proliferative diabetic retinopathy)
Justification:

- History of long-standing diabetes in this patient
- Sudden painless loss of vision
- Absence of red reflex
- B- scan showing multiple echogenic shadows within the vitreous cavity.

To rule out accompanying complications, perform:

- Tonometry
- Gonioscopy
- Complete ocular examination (To note for iris neovascularization etc.)

4. What are the important principles in the management of this condition?

Finally, you should understand the important principles in the management of this condition.

These include:

- Wait for the hemorrhage to be absorbed for a minimum duration of 3-6 months.
- Monitor and control intra-ocular pressure.
- Pars plana vitrectomy.
- Panretinal LASER photocoagulation.
- Anti- vascular endothelial growth factor (VEGF) agents
- Combination treatments
- Prophylactic LASER photocoagulation for the fellow eye.
- Good glycaemic control
- Watch for other systemic effects of long-standing diabetes.

Continuous Learning Issues:

- Evidence based information on common causes of vitreous hemorrhage.
- Evidence based information on protocols for management of vitreous hemorrhage especially combined treatment strategies.
- Evidence based information on the role of alternative medicine in the management of vitreous hemorrhage and other complications of long-standing diabetes.

Case 17

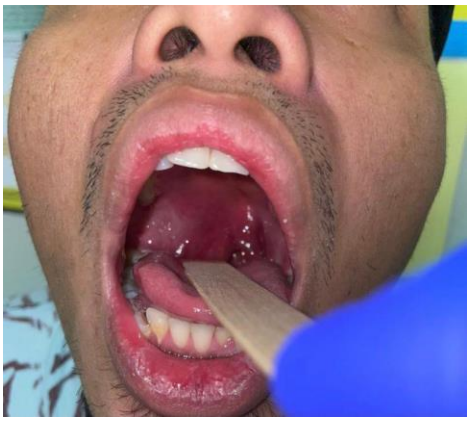
A 22-Year-old male presents with a 5- day history of pain on swallowing, difficulty in taking orally and fever. More recently he is unable to open his mouth fully. He was seen by his general practitioner and given oral antibiotics. But his symptoms continued to worsen.

On Examination:

Temperature: 39^{o}C, Pulse: bounding, 90 beats/minute

He has trismus, right tonsil is grade III with a membrane over it, and right soft palate is bulging.

The image below shows the appearance of the pharynx.



Questions

1. What is a peritonsillar abscess (Quinsy)?
2. What is the differential diagnosis of quinsy?
3. What are the complications of peritonsillar abscess?
4. How do you manage peritonsillar abscess?

Discussion

With the above information, we should consider all conditions that present with a membrane over the tonsil as possible diagnoses, such as:

1. Diphtheria
2. Acute follicular tonsillitis
3. Infectious mononucleosis
4. Vincent's angina
5. Agranulocytosis
6. Leukemia

However, considering the other features of each cause mentioned above, none of them causes trismus nor a bulging soft palate. So, our reflection should deviate to another group of pathologies which has both enlarged tonsils and soft palate bulging as clinical features, and also consider whether the finding in this patient is an enlarged tonsil or a medialised tonsil.

If we assume it is medialisation of the tonsil, parapharyngeal abscess is a condition which presents with medialisation of the tonsil and trismus, and may follow acute follicular tonsillitis, but with no soft tissue bulging. Now, we are left with only one diagnosis which is peritonsillar abscess which presents with trismus, fever, odynophagia, bulging soft palate with or without acute tonsillitis.

1. **What is a peritonsillar abscess (Quinsy)?**

Peritonsillar abscess is a suppuration related to the pharynx where the pus accumulates between the capsule of the tonsil and the lateral pharyngeal wall (peritonsillar space), usually follows acute tonsillitis where Crypta Magna becomes obstructed, infected. It is recently described as an infection of minor salivary glands called Weber's glands in the supra tonsillar fossa, more frequent in males.

Quinsy usually presents with:

1. High grade fever

2. Headache
3. Trismus (difficulty opening the mouth) due to contraction of medial pterygoid and masseter muscles.
4. Dysphagia and odynophagia (patient cannot even swallow his own saliva)
5. Halitosis
6. Otalgia
7. Level II cervical lymphadenitis (jugulo digastric lymph node)
8. Marked hyperaemia and oedema of the tonsillar and palatal regions; tonsil itself may be almost or completely obscured.
9. Usually follows acute tonsillitis but tonsil may be inflamed, enlarged or normal, even can happen in patients with remnants.
10. Bulging of soft palate ipsilaterally; there may be a yellowish spot on the soft palate swelling (impending rupture point)
11. Excessive salivation, coated tongue, and torticollis (sternocleidomastoid muscle spasm) specially in children

If untreated the abscess usually bursts

2. What is the differential diagnosis of quinsy?

1. Parapharyngeal abscess
2. Retropharyngeal abscess
3. Tonsillar tumors

3. What are the complications of peritonsillar abscess?

- Parapharyngeal abscess (commonest complication)
- Laryngeal oedema (life-threatening)
- Cavernous sinus thrombosis
- Hemorrhage
- Rupture of the carotid artery
    - Mediastinitis has been reported

4. How do you manage peritonsillar abscess?

- Investigations: Swab culture & sensitivity
- Medication in the early stage
    1. Antipyretics
    2. Analgesics
    3. Broad spectrum antibiotic, oral or parenteral (causative organism mostly *Streptococcus hemolyticus*)
- Drainage - if conservative medical management fails and symptoms worsen

How and where to drain?

1. Be sure that the appropriate instruments are available
    - Good functioning suction machine and a suction tip
    - Quinsy knife (or scalpel covered with adhesive plaster except the tip)
    - Quinsy forceps
2. Patient sits upright during the procedure.
3. Performed mostly under local anesthesia except children and some selective cases
4. Admit the patient for monitoring
5. Commence intravenous broad spectrum antibiotics

Points of drainage

1. Impending rupture point (yellow spot)
2. Meeting point between 2 lines (one passing along base of uvula and other along last upper molar tooth
3. 5 mm lateral to meeting point of 2 lines (base of uvula and another one passing along the attachment of anterior pillar to the base of tongue)

Post-drainage management

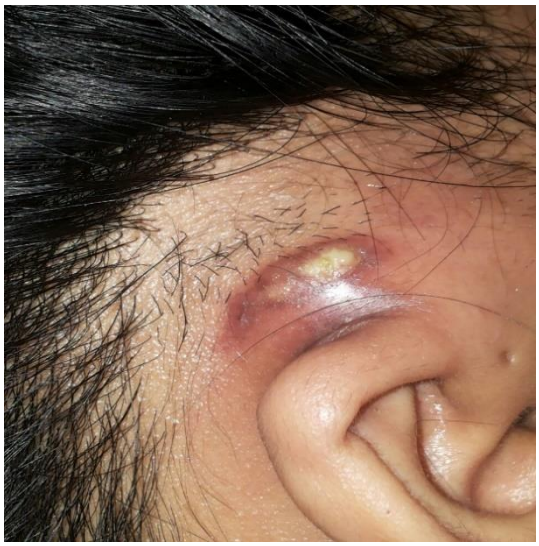
1. Examine the patient next day and ensure that the abscess has been evacuated totally and the abscess cavity is clean

2. Keep the patient in hospital till all clinical manifestations resolve (no trismus, no fever, no soft palate bulging or hugely reduced and no fever)
3. Continue the antibiotic for at least 10 days
4. Perform tonsillectomy at least 1 month after all clinical manifestations subside

Case 18

A 17-year-old girl presents with a fluctuant painful swelling above the right auricle. There is a visible pit in front of the right auricle. She claims that she had a previous similar attack with foul smell discharge coming out of the pit, which subsided after receiving antibiotics from her doctor.

The image below shows the lesion in this patient.



Questions

1. What differential diagnosis will you consider?
2. How would you manage this patient?

Discussion

1. **What differential diagnosis will you consider?**

Referring to the image above with the history mentioned we should think about the differential diagnosis considering the two main features.
Preauricular swelling or swelling above the auricle has many differential diagnoses, but the presence of the pre-auricular pit collapses our list to only a few.
he causes of preauricular swelling or swelling above the auricle are

- Preauricular swelling/infection.
- Parotid swelling/mass/tumor
- First branchial cleft cyst
- Duplication of ear canal
- Trauma

Thinking about the above-mentioned causes with the presence of pre auricular pit leaves us at one diagnosis which is pre-auricular sinus.

Then, we should think whether they are two separate pathologies? (pre-auricular sinus and another swelling) or one pathology?

Generally, we should consider it to be one pathology first, till proven otherwise.

Pre-auricular sinus and its origin:
Pre-auricular sinus is a common congenital anomaly described by Van Heusinger in 1864 as a result of incomplete fusion of the hillocks of His arising from the first and second branchial arches. It is characterized by a dimple or pit (hole) in front of the helix of the auricle leading to a blind track which can extend to the surrounding soft tissue and is lined with squamous epithelium. Obstruction of the mouth of this tract leads to formation of a cyst that can become infected.

The pre-auricular sinus can remain without any symptoms for long time or can be infected as mentioned above, in which case we should drain the abscess, we excise the opening, cyst and the whole tract to prevent recurrence.

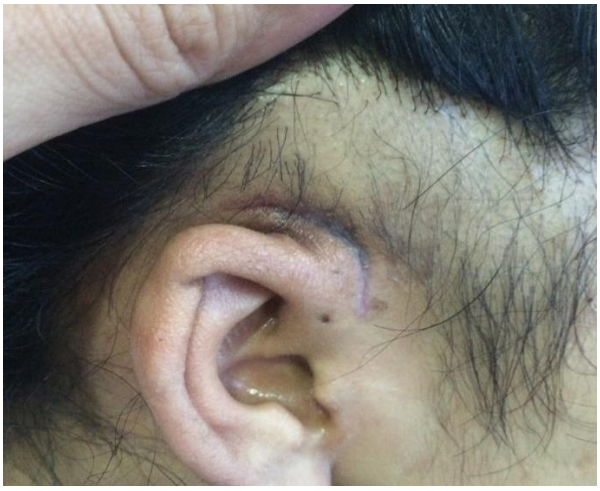
From the history and the photo above, we can tell clearly that the above-auricle swelling is an abscess related to the pre-auricular sinus. By introducing a small probe inside the sinus opening after sedating the patient we can reach the abscess site.

2. **How would you manage this patient?**

As long as the sinus is quiet, symptomless and no infection we do nothing.

Once infection starts, we should start antibiotics. As *Staphylococcus aureus* is the most common organism causing such infections, and other possible organisms are Proteus and Streptococcus, the antibiotic of choice is amoxicillin/clavulanic acid..

If the infection progresses and an abscess forms, drainage is mandatory, under cover of antibiotics.

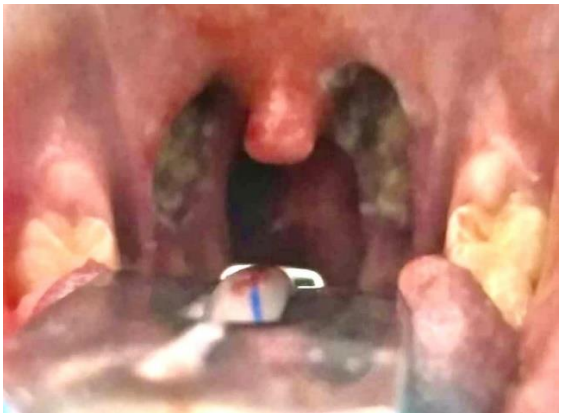


Then, after the patient recovers well and the healing process is complete, we excise the sinus, cyst and the tract completely.

Case 19

A 40-year-old male has had recurrent tonsillitis for the last few years. The patient has sleep disturbance with loud snoring, morning headaches and sleepiness or lack of energy during daytime.

The patient consulted his ENT specialist who performed a procedure on him. The figure below shows the oral cavity after the procedure.



Questions

1. What are the common indications of tonsillectomy?
2. What is Waldeyer's ring?

Discussion

The image obviously shows that this patient has had a tonsillectomy

1. **What are the common indications of tonsillectomy?**

1. Infections

- Recurrent acute tonsillitis with more than 6–7 episodes in one year, 5 episodes per year for two years, or 3 episodes per year for three years
- Peritonsillar abscess with the involvement of the tonsil

2. Obstruction

- Obstructive sleep apnea

3. Suspicion of malignancy

- Suspicious tonsillar neoplasm

4. As part of other surgeries

- e.g. Uvulopalatopharyngoplasty (UPPP) or Cautery-assisted palatal stiffening operation (CAPSO)

5. To access deeper structures (e.g., styloid process)

In this case, the patient has a history of recurrent tonsillitis and obstructive sleep apnea.

Is tonsillectomy common in this age?

No, it is common in young age where the hypertrophy of the tonsils (Waldeyer's ring) and infections are more common.

2. What is Waldeyer's Ring?

It is a ringed pattern of lymphoid tissues surrounding and guarding the entrance of aerodigestive tract and consists of:

- Nasopharyngeal tonsil (if symptomatically hypertrophied, called adenoid)
- Tubal tonsil (surrounds the Eustachian tube opening)
- Lateral pharyngeal bands (posterolateral wall of oropharynx)
- Palatine tonsil (lateral wall of oropharynx between anterior & posterior pillars)
- Lingual tonsil (base of tongue)
- Other small discrete lymphoid tissue on the soft palate

The ring drains into the upper deep cervical and retropharyngeal lymph nodes

As with any surgical procedure, there are risks and complications of tonsillectomy. Risks include:

- Hemorrhage (primary, reactionary, and secondary)
- Pain (can be severe especially with use of diathermy)
- Dehydration
- Airway obstruction
- Tongue, mucosal and dental injury
- Infection
- Oropharyngeal stenosis

The main complication is hemorrhage, and it is crucial to know the blood supply of the tonsil to be able to avoid and control it.

Blood supply of the tonsil arises from branches of the

- External carotid artery
- Inferior pole >> tonsillar branches of dorsal lingual artery, facial artery and the ascending palatine artery

- The superior pole >> tonsillar branch of the ascending pharyngeal artery and the lesser palatine artery.

The tonsil rains into the pharyngeal venous plexus

Obstructive sleep apnea (OSA)

Most common type of sleep-disordered breathing characterized by episodes of upper airway collapse, with cessation of the airflow.

Clinical features and associations

- Morning headache
- Daytime sleepiness
- Sore throat
- Apneas
- Snoring
- Insomnia
- Choking and gasping that awakens the patient from sleep
- Waking up as tired as they never slept
- Fatigue
- Memory and intellectual impairment
- Personality and mood changes
- Gastro oesophageal reflux disease
- Hypertension
- Sexual dysfunction
- High-arched palate
- Obesity and enlarged neck circumference and with receded mandible (causing difficult intubation)
- Grade III to IV hypertrophic tonsils
- Collapsed airway (must perform Muller's maneuver)

Investigations

1. Sleep study, or polysomnography
2. Pulmonary function tests may be needed

Treatment

- Continuous positive airway pressure (CPAP) is the gold standard for moderate and severe OSA
- Upper airway surgery such as septoplasty, turbinoplasty, reduction of soft palate redundancy, turbinates radiofrequency ablation (RF), uvulopalatopharyngoplasty, laser-assisted uvulopalatoplasty (LAUP), and radiofrequency probe (somnoplasty).

Case 20

A 67-year-old gentleman gives a 2-day history of fever, left neck pain followed by rapidly progressive neck swelling on the left side which became bilateral in one day. He also gives a history of extraction of the lower left second molar tooth recently.
On examination, he is febrile and has limited mouth opening, bilateral neck swelling, protruded elevated tongue and slight neck rigidity but no shortness of breath.

Questions

What is Ludwig's Angina?

Discussion

Considering this gentleman's clinical features and looking at the image, we should think of the differential diagnosis for such pathology.

First, we should consider deep space neck infection and its causes.

Identification of the site of the neck swelling and the space of the neck that is involved is of much value, as then we can diagnose the deep neck infection and extension and decide on how to manage it.

In this patient, Ludwig's Angina is highly likely, as the symptoms and signs are suggestive of this condition and the picture suggests the involvement of the submandibular spaces (submaxillary and sublingual spaces)

What is Ludwig's Angina?

It is an abscess which is characterized by a rapidly spreading cellulitis of the submandibular space (submaxillary and sublingual spaces), does not tend to form an abscess and potentially fatal. More than 70% of cases are from dental origin while other causes include mandibular fractures, diabetes mellitus, AIDS and penetrating wounds of the floor of mouth. As the oedema involves the floor of the mouth, it tends to push the tongue upwards and posteriorly against the palate resulting in respiratory obstruction.

However, we should also consider other differential diagnosis as:

- Acute parotitis
  Unilateral swelling at the parotid area
- Peritonsillar abscess (quinsy)
  Soft palate bulging, trismus and deviation of the uvula
- Retropharyngeal abscess

Midline bulging of posterior pharyngeal wall

Clinical features of Ludwig's angina

- Fever
- Trismus and neck rigidity
- Dysphagia and Odynophagia
- Edema & brawny induration of the floor of the mouth and the neck which starts unilaterally then progressively becomes bilateral
- Protruded and elevated tongue against the palate
- Possible signs of respiratory embarrassment due to obstruction

Management

Investigations:

- Full blood count, ESR and CRP
- Blood sugar level
- CT scan neck with contrast

Treatment

- Patient should be managed in hospital
- Early airway assessment, laryngoscopy should be done (tracheostomy may be needed)
- Combination of a broad-spectrum antibiotic and an anti-anaerobic intravenously for at least 2 weeks' duration
- Rapid Surgical drainage should be done

Case 21

A 25-year-old basketball player presents to the Emergency Department after a fall while playing basketball with his friends. On examination he has deformity of his right shoulder, flattening of shoulder contour and prominent bony landmark in right shoulder tip. Swelling is noted over the right pectoral area. Radial pulse and sensation are intact. No other associated injury is noticed.

Questions

1. State the possible differential diagnoses and give one most likely provisional diagnosis.
2. Give reasons for your provisional diagnosis.
3. State how you would confirm the diagnosis and give the expected findings.
4. What are the complications?

Discussion

1. **State the possible differential diagnoses and give one most likely provisional diagnosis.**

In this scenario these are the following points to be considered-

- The patient is 25-year-old basketball player. He is still young, and active who uses extreme movement of his upper limb for playing.

- He fell while playing. It can be taken as a history of trauma.

- He presented with deformity, flattening of shoulder contour and prominent bony landmark in his right shoulder. It can be taken as a sign of fracture or dislocation

- The flattening of shoulder contour means loss of rounded deltoid contour of shoulder. Deltoid contour is formed by underlying humeral head. When humeral head is no longer in its original position there will be loss of deltoid contour. Prominent bony landmark is secondary to loss of deltoid contour.

- Swelling over the right pectoral area may be due to two reasons, pathology of pectoral muscle or some other structure displaced to pectoral area. In this case most likely the structure is dislocated humeral head.

Based on the above points of history of trauma and deformity the first diagnosis you should consider is anterior dislocation of the right shoulder joint.

2. **Give reasons for your provisional diagnosis.**

The justifications for the diagnosis of anterior dislocation of the right shoulder have been mentioned above. In addition, anterior dislocation is the commonest type of shoulder dislocation.

For the differential diagnoses, you should consider fracture around the shoulder and other types of shoulder dislocations. In this scenario there is a history of trauma, you should focus on conditions of traumatic aetiology. The available clinical information does not point you towards considering infection or malignancy. Thus, your differential diagnosis should include the followings.

- Posterior dislocation
- Inferior dislocation
- Fracture of the proximal humerus
- Fracture of coracoid, acromion and glenoid

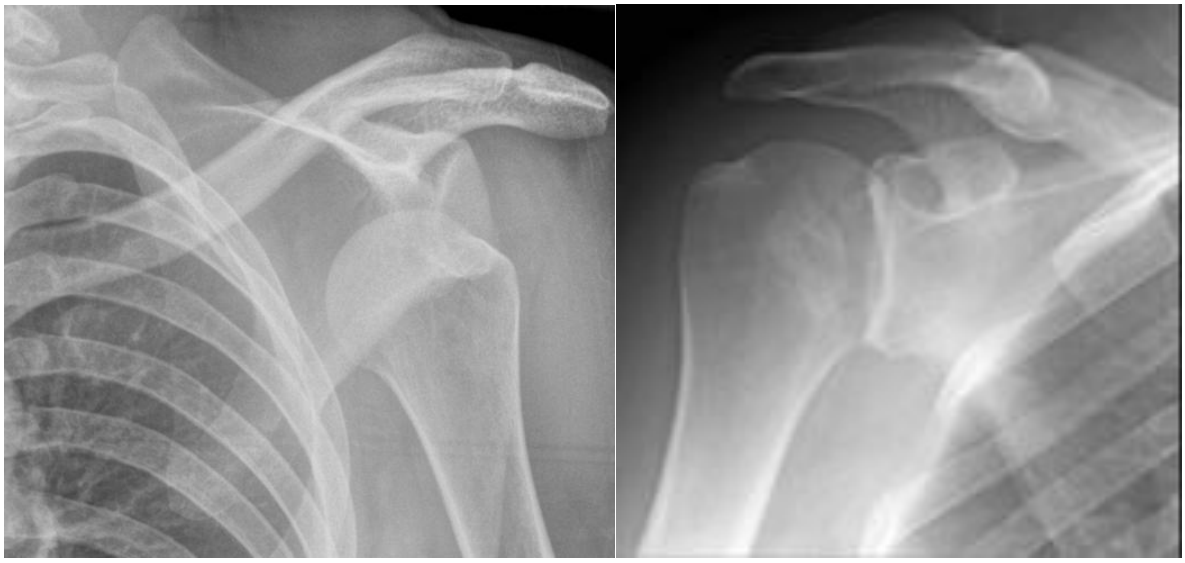
3. State how you would confirm the diagnosis and give the expected findings.

In Orthopaedics, X-ray is the most basic investigation. If X-ray does not provide adequate information, the next step is usually CT or MRI. Generally, CT is indicated and is useful in occult fractures while MRI is indicated in ligament or soft tissues injuries. Since the clinical features in this patient are obviously those of dislocation, you should be able to discuss the different radiological findings in different types of dislocation.

There are different view of shoulder X-rays but at the minimum you must know AP view and Lateral Y view to diagnose dislocation.

Anterior dislocation is easy to diagnose in AP X-ray. Humeral head is outside and below the glenoid socket. The humeral head is displaced antero-medially and over the scapula shadow.

In posterior dislocation the humeral head is above and posterior to the glenoid. In inferior dislocation, which is very rare, and humerus is pointing upward.



A: Anterior dislocation of a shoulder joint B: Posterior dislocation of a shoulder joint

Note the difference between anterior dislocation and posterior dislocation

(Sources: https://radiopaedia.org/articles/shoulder-dislocation)

When you discuss radiological findings of anterior dislocation, you should be familiar with two terms, Hill-Sachs lesion and Bankart lesion. Hill-Sachs lesion is compression fracture of posterior surface of humeral head due to repeated compression against the anterior surface of glenoid. Bankart lesion is a detachment of anterior labrum at the glenoid. Both lesions are due to recurrent anterior dislocation. It is better to diagnose with MRI.

4. What are the complications of shoulder dislocation?

Complications are best considered as immediate, early, and late complications. You can simply list down.

Immediate complications include,

Damage to the circumflex axillary nerve.

Arterial damage.

Irreducibility.

Late complications include,

Joint stiffness.

Recurrent dislocation.

Finally, you should understand the common methods of shoulder reduction.

There are many methods of reduction of shoulder dislocation. At least, you are expected to know one method.

These are historically well-known methods in reduction of shoulder dislocation.

Hanging-arm method (also known as Gravity method or Stimson method), Hippocratic Method and Kocher's method

Kocher's method

Nowadays Kocher's method is widely used in clinical setting. The steps in the Kocher method are as follows,

1. Traction-counter traction of the affected arm
2. External rotation and slight abduction
3. Adduction and internal rotation

Before reduction adequate sedation or short general anesthesia is recommended. Steady traction force should be given for about 5-10 minutes before you proceed to step 2. During reduction you may hear the clicking sound or get the feeling of relocation once it is reduced.

Case 22

A 35-year-old man developed severe back pain while working out in the gym. He lifted heavy weights in the gym and experienced worsening back pain as he went back home. It was difficult for him to stay a comfortable position. Next morning, he noticed that the pain was worsen, and he could not immediately get up from bed. The pain radiated down the back of right leg. He experienced loss of sensation in his perianal area and had episodes of urinary incontinence. He is brought to the Emergency Department, and you are the on duty medical officer.

Questions

1. What is the diagnosis?
2. What is the current condition of this patient?
3. What specific clinical examination would you perform on this patient?
4. How would you manage this patient?

Discussion

1. **What is the diagnosis?**

In this patient there are many strong pointers to narrow down the diagnosis. First, the chief complaint is low back pain. With this, one should immediately consider the differential diagnosis of low back pain. There are many differential diagnoses of low back pain. However, when you look at the age of the patient, he is middle age. If you narrow down the causes of low back pain in middle age it should not be too much.

The differential diagnosis of low back pain should include the followings.

Mechanical pain and muscle spasm

This is one of the most common pathologies; the pain is localized and there is no radiation of pain towards the lower limb.

Vertebral fracture

There will be an obvious history of trauma. Pain is localized in the spine. There will be local signs of fracture such as step, gap and hematoma or bruise.

Spinal infection

There will be signs of infection, such as fever. Pain is not related to physical activity.

Congenital abnormality

Most commonly symptoms start earlier than middle age.

Referred pain

You should exclude referred pain in every case of low back pain. The patient will manifest associated signs and symptoms of the primary disorder. For instance, renal colic, duodenal ulcer pain.

Prolapse intervertebral disc (PID)

The history of this patient is the typical presentation of PID. Points in favors of the diagnosis are- middle age, precipitating factor (weightlifting), radiation of pain to lower limb (sciatica). Therefore, after exclusion of other possibilities PID is the most likely diagnosis in this patient.

2. What is the current condition of this patient?

In this patient, in addition to low back pain his complaint is loss of perianal sensation and urinary incontinence. These two clinical signs together with low back pain suggest cauda equina syndrome.

What is cauda equina syndrome?

The cauda equina is the collection of nerves fibers at the end of the spinal cord, starting from L1-2 vertebral level. Cauda equina syndrome occurs when the nerve roots of the cauda equina are compressed and disrupt motor and sensory function to the lower extremities and bladder.

You should suspect cauda equina syndrome in a patient with low back pain who develops the following signs and symptoms:

- Saddle anesthesia (loss of sensation on the skin around anus)
- Bladder dysfunction (urinary retention or incontinence)
- Bowel incontinence
- Loss of reflexes in the extremities

One more question you would be asked is the precipitating factors.

Disk herniation is mostly gradual, aging-related wear and tear process. If there is association with risk factors, more chance of herniation in early decades. The risk factors include:

- Obesity
- Occupation. Weightlifting, physical labor.
- Smoking. Smoking reduces blood flow to disc.
- Family. Strong family is high risk.

3. What is the specific clinical examination for PID?

The Straight Leg Raising test. Here, I am not going to discuss SLR test technique in details. Please take a note that significant of SLR test is to confirm the sciatic nerve tension. Bow string sign is additional confirmatory test.

How would you investigate it?

- X-ray is not specific to diagnosis prolapses discs. However, it is important to rule out the other causes of low back pain such as fracture, renal stone. The disc space narrowing can be seen in X-ray, but it is not a specific finding.
- MRI (magnetic resonance imaging) is currently best imaging to diagnosis discs problems.
- CT (computed tomography) scan also can be used if MRI is contraindicated.
- CT myelogram is, nowadays, not a first choice due to its complications such as anaphylactic reaction and it is invasive.

Note. There is a possibility of asymptomatic disc herniation. Multiple level herniation. It is crucial to check MIR level of disc herniation and clinical finding to confirm the diagnosis.

4. How would you manage this patient?

Initially treatment should be started with conservative management. Bed rest for two to three days is recommended to relief pain immediately. Modifying daily activities to avoid stress on the back e.g. bending of the back, weight lifting is advisable.

Medications commonly used in low back pain are,

- Pain killer (NSAID or opioid)
- Muscle relaxant
- Cortisone injections (it is symptomatic treatment, symptoms relief in a few days but high risk of recurrent and complications steroid are need to be justify)

Physical Therapy

There are so many physical treatment modalities. Heat therapy cold therapy, wax therapy, USG therapy, wax therapy. Acupuncture, TENS. Therapeutic efficacy of all these physical modalities are variable.

Surgery

In refractory case, cauda equina syndrome, or severe prolapse, surgery is the treatment of choice.

Fenestration and discectomy (open), or microdiscectomy (arthroscopic) is the technique of choice. Recently implantation of an artificial disk becomes popular after discectomy.

Case 23

A 60-year-old retired teacher presents with bilateral knee pain. The pain is related to climbing stairs and walking long distance. This pain has gradually worsened over the past two years. She obtains temporary relief by pain killer and rest. She has no history of trauma and there is no sign of infection. There is no pain in other joints. On examination, she is obese, she has bilateral varus knee with, tenderness and crepitus over both knee joint. There is no other abnormality in other joints.

Questions

1. What are the differential diagnoses? And provisional diagnosis.
2. Discuss the pathophysiology of your provisional diagnosis.
3. Describe the carinal radiological signs of above conditions.
4. What are the management options?

The first possible question you should expect is-

1. **What are the differential diagnoses?**

In this scenario the chief complaint is knee pain without trauma, and without sign of infection. In that case you should start to consider degenerative condition like OA and RA ahead of other uncommon disorder. If the patient is teen age, you may want to think musculoskeletal tumor such osteosarcoma or GCT in the first place.

In this case the scenario is more on OA side rather than RA.

Points for OA in this case are as follows.

Age-65

Gradual onset related to activity, walking, climbing stairs.

Obese

Tenderness, crepitus and varus knee

Points against RA are as follows.

No other joint deformity

No valgus knee.

Therefore, this case is most likely osteoarthritis.

2. **Discuss the pathophysiology of OA.**

If you discuss OA, you should be able to recall the pathophysiology of OA. It is recommended to under overview of pathogenesis. In summary, steps are as follows:

Enzymes disrupting of the type II collagen causes softening and swelling of the cartilage. Which leads to fragmentation and fissuring of the cartilage, the process is understood as proteolysis. Finally massive loss of cartilage volume results in exposure of underlying bone.

In case of OA, diagnosis is usually with X-ray. You should remember important radiological signs in OA. The possible question is

3. **Describe the carinal radiological signs of above conditions.**

The typical radiological signs are-

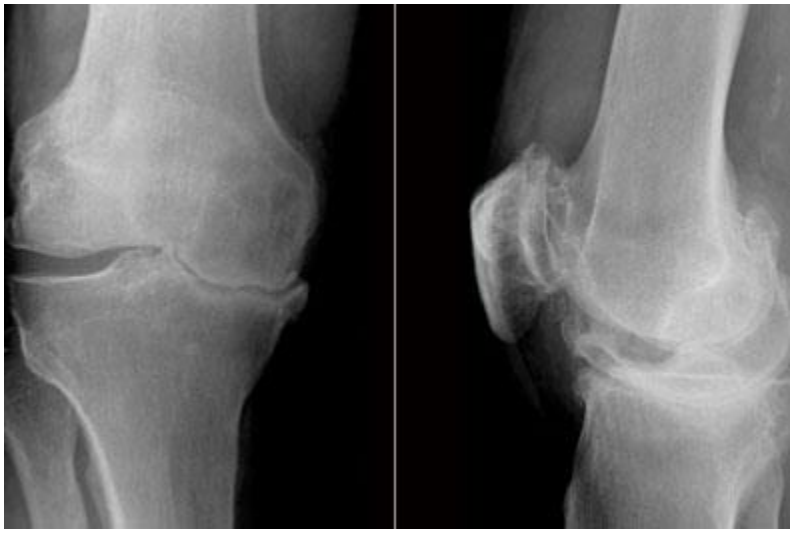
Joint space narrowing

Subchondral sclerosis

Subchondral cysts

Osteophyte formation

In the following X-ray you can appreciate all features except subchondral cysts, which is relatively uncommon finding.



(Sources: https://radiopaedia.org/articles/shoulder-dislocation)

.**What are the management options?**

The management of osteoarthritis is decided according to the severity of the symptoms.

In early case patient education and lifestyle modification work for most case.

Weight control, regular exercise, simple analgesics, topical agents, reduce climbing starts, avoid squatting and sitting on the floor are important lifestyle changes.

In moderate case, non-steroidal anti-inflammatory drugs, muscle relaxant, physiotherapy, injection Hyaluronic acid, glucosamine and chondroitin compound are common options.

In advance case

Joint debridement, partial or total joint replacement or osteotomy are indicated.

Case 24

A 70-year-old man presented with postprandial epigastric pain. He was prescribed antacids, which were ineffective; his weight then decreased by 11 kg in 3 months. The patient did not consume alcohol but was a 15-pack-year smoker. On Physical examination the patient was pale. His vital signs were within normal limits. On abdominal examination, Gentle, light palpation revealed epigastric tenderness on pressure manifested as resistance to palpation. A palpable nodule bulging into the umbilicus was noted. On deeper palpation of the abdomen, no palpable intrabdominal mass detected. The knee elbow palpation of his abdomen also didn't reveal any intra-abdominal or retro peritoneal mass.

His complete haemogram revealed a hemoglobin percentage of 6.8 g/dl. The WBC and the platelet counts were unremarkable. His hepatobiliary enzyme levels were not elevated. However, he was noted to have mild renal insufficiency with a serum creatinine level of 2 mg/dl. (normal range is 0.6 - to 1.3 mg/dl) and a serum Blood Urea Nitrogen (BUN) level of 30 mg/dl (Normal 8 to 20 mg/dl. The carcinoembryonic antigen level was within normal limits, whereas the carbohydrate antigen 19-9 level was elevated at 98 U/mL The anti-H pylori antibody was positive at 30 U/mL

Sister Mary Joseph nodes in the umbilicus with other gastrointestinal symptoms prompt the physician to make a provisional diagnosis of carcinoma of the stomach and spread of gastric carcinoma to around the umbilicus.

Questions

Is the clerking detail sufficient? What else do you want to explore in the complaint and history?

Discussion

Is the clerking detail sufficient? What else do you want to explore in the complaint and history?

The key to diagnosis may be in the clinical history. The importance of using the patient's complaints, history and physical examination lies with clinicians (here, students) as a basis for selecting relevant diagnostic testing, which leads to a timely and accurate diagnosis. This process protects patients from the risks of unnecessary testing and is cost-effective. The history in this clinical scenario is pointing to the gastrointestinal system. Therefore, we must explore more symptoms pertaining to this. First, we have to get the missing data and next then explore the existing data further.

- Did the patient have vomiting? How often and when? Was it after intake of food? How long after food, immediate or after some time?
- Was there haematemesis?
- History of diarrhoea/dysentery constipation (altered bowel habits). melena,
- Bloating of stomach after meals (postprandial fullness).
- Was there any attack of jaundice anytime? itchy skin, white-coloured stool?
- Coughing/hoarseness
- Haemoptysis?
- Recurrent thromboembolism
- Family history of any cancer including gastric carcinoma?

Exploration of the existing data includes:

- Pain: site, onset/ timing pre or postprandial epigastric pain (how many hours before or after food?), characteristics, (pressure/burning in the chest), severity, radiation (radiates to back, shoulder, retro-sternum), Associated symptoms,
- Exacerbating/relieving factors.
- Any reflux of food?
- Dysphagia (solid/fluid, all the time/initial time, exact location of difficulty, feel of vomiting);
- Loss of weight (LOW)? Gradual or sudden LOW? How many Kilo lost?

Next step is to list some more relevant physical examination recommended in this case.

- The patient may be anaemic, cachexic with or without signs of jaundice.

- Any visible gastric or intestinal peristalsis or abdominal mass? pyloric obstruction causes epigastric distension and visible gastric peristalsis. This patient has a palpable nodule bulging into the umbilicus. Palpate for any lymph nodes swellings (abdominal, axillary and left supraclavicular).
- The rationale of knee elbow palpation (KEP) of abdomen, bimanual palpation in this case has to be explained. What is KEP? How to do? What to elicit? This position is specially done to locate mass that is supposed to fall forward if it is intraperitoneal.
- Look for hepatomegaly (may result from liver metastasis or obstructive jaundice, congestion).
- Oedema of the lower limbs – recurrent venous thrombosis may occur due to neoplastic disease; Percussion to look for ascites (Fluid thrill and shifting dullness).
- The abdominal examination is incomplete if it does not include 'Rectal, scrotal and hernial orifice' examination. Lung examination - to look for presence of pleural effusion for any pulmonary metastases.

Next step is to correlate the available laboratory results and suggest other important lab tests that are lacking.

- The most common tumour markers in patients diagnosed with gastric cancer are CA 72-4, CEA, CA 19-9. The CEA in this patient is normal. CA 19-9 in this patient is elevated. Carbohydrate antigen 19-9 elevated (98 U/mL) → Elevated in gastrointestinal cancer & noncancerous condition like biliary tract obstruction CA 72-4 and AFP have not been assessed. (May be suggested later). These are clearly not ideal **tumour markers**. They lack day-to-day reproducibility and sufficient sensitivity and specificity to be useful as general screening tools. The clinical importance of the preoperative serum levels of these tumour markers in gastric cancer (GC) is not well known.
- Haemoglobin - 6.8 g/dL (anaemia); Frequent bleeding; impaired dietary iron absorption, chronic gastritis by *H.pylori* have to be considered.
- Mild renal insufficiency: A complete renal function test may be done. GFR <60 to 70 ml/min and/or the presence of increased urinary albumin excretion;

Hypertension/ DM; release of inflammatory cytokines has a toxic effect on kidney, and long-term chronic inflammation could have lead to the kidney damage

- Anti-H pylori antibody positive (30 U/mL) → Positive Helicobacter pylori: suggest this patient is having H pylori infection which may or may not be significant in this case. Measures IgG antibodies (enzyme-linked immunosorbent assay); Currently infected or have been infected; Could indicate acute and chronic gastritis, recurrent duodenal ulcer, gastric ulcer, gastric carcinoma; associated with development of gastric cancer

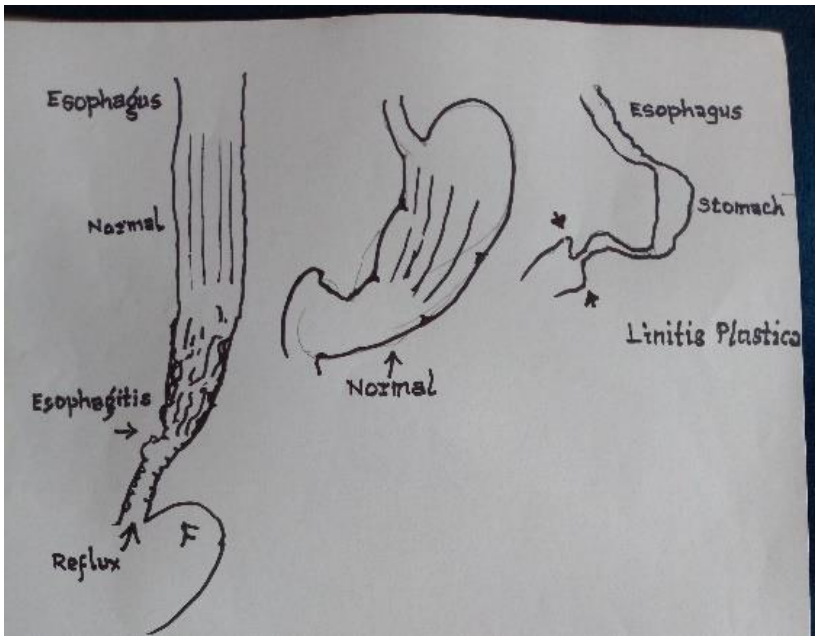
Suggestions of the following laboratory tests have to be made in this case:

1. Complete Renal function test including pH, electrolyte abnormalities, serum calcium, phosphate, potassium; metabolic acidosis
2. Urinalysis: to look for haematuria or proteinuria; cellular casts; Urine albumin: if increased suggesting CKD
3. Serum glucose: If hyperglycemia, it could contribute to diabetic nephropathy
4. HbA1c: if increased could suggest poor control of diabetes and correlates with complications of diabetes such as diabetic nephropathy

As the provisional diagnosis of gastric carcinoma was in his mind, the attending physician advised upper GI barium study and endoscopy in this patient.

Usually during upper GIT contrast x-ray, after ingesting barium contrast under fluoroscopy, the esophagus shows healthy long parallel mucosal folds. The patient is then asked to gulp plain water to wash off excess barium sticking to the esophagus. In the subsequent upper GIT study, barium will not be seen in the normal esophagus. In this patient, in the 1st diagram below on your left, a large extent of distal esophagus shows obviously abnormal irregular wall and irregularity of mucosal folds suggestive of abnormal gastroesophageal reflux (reflux oesophagitis). In the 2nd picture (on the middle) you can see normal capacious stomach and in the third diagram on your right, a small contracted stomach, with narrow rigid lumen, reduced capacity, loss of

distensibility and no peristaltic impressions, effacement of mucosal folds with no rugae pattern and narrow smooth antrum of stomach → shouldering effect (arrow head).



The endoscopist pointed out that (as shown below diagramatically)

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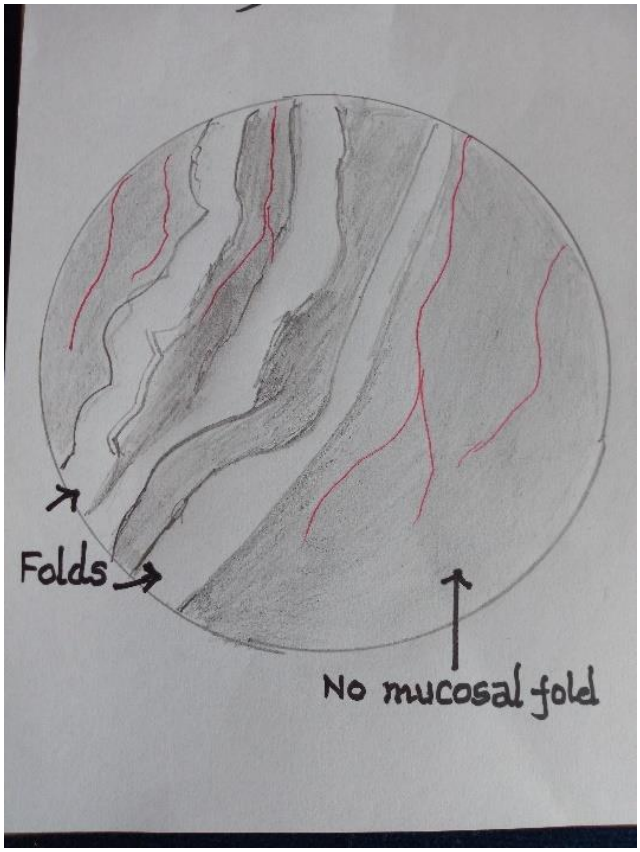
the greater curvature showed diffuse 'ironed-out' mucosa with a few swollen gastric mucosal folds. Endoscopy did not find intra-luminally projecting malignant growth. The physician referred the patient to a tertiary institution. In the tertiary hospital, the patient underwent 'repeat esophagogastroduodenoscopy' that revealed incompetent gastro-esophageal junction, reduced gastric capacity with outlet obstruction, and lack of peristalses.



Our next question is, are these findings scientifically discernible?

These endoscopy findings are to be correlated with the barium upper GI tract study. You should be in a position to understand that most of the gastric mucosa are 'ironed-out' and smooth with a few swollen gastric mucosal folds. This coexistence of atrophy and swelling may occur in the presence of gastric adenocarcinoma(GAG) /Chronic atrophic gastritis (CAG) and Helicobacter pylori infection. *The* reduced gastric capacity with outlet obstruction may be correlated with the clinical finding of reflux esophagitis and difficulty swallowing. The most important point to consider in this case is the gastric mucosal atrophy which is a crucial stage in the progress of gastric cancer, and the extent of atrophy is an important risk factor for gastric cancer. As the endoscopist did not find any intraluminal tumour, it is mandatory to perform endoscopic biopsy to obtain clear findings. Recent concept advocates multipoint

biopsy if the patient is not taking medications like aspirin as it increases gastric trauma and risk of bleeding. Recently, both chromoendoscopy combined with magnifying endoscopy and confocal laser microscopy have been important tools for the diagnosis and differential diagnosis of chronic atrophic gastritis. However, the diagnostic accuracy still depends on the standardized operations of accumulation of a large volume of pathological mucosa.

In the tertiary centre, endoscopic ultrasound (EUS) was done in this patient which indicated gastric wall thickening up to 9.3 mm mainly in the body of the stomach. The different five layers of the gastric wall had been disappeared. Low-level echoes were detected throughout the entire gastric wall. No lymphadenopathy noted. Multiple EUS-FNA of the gastric wall was performed parallel to the wall and a few larger and deeper biopsies were taken from the suspected areas of the stomach.

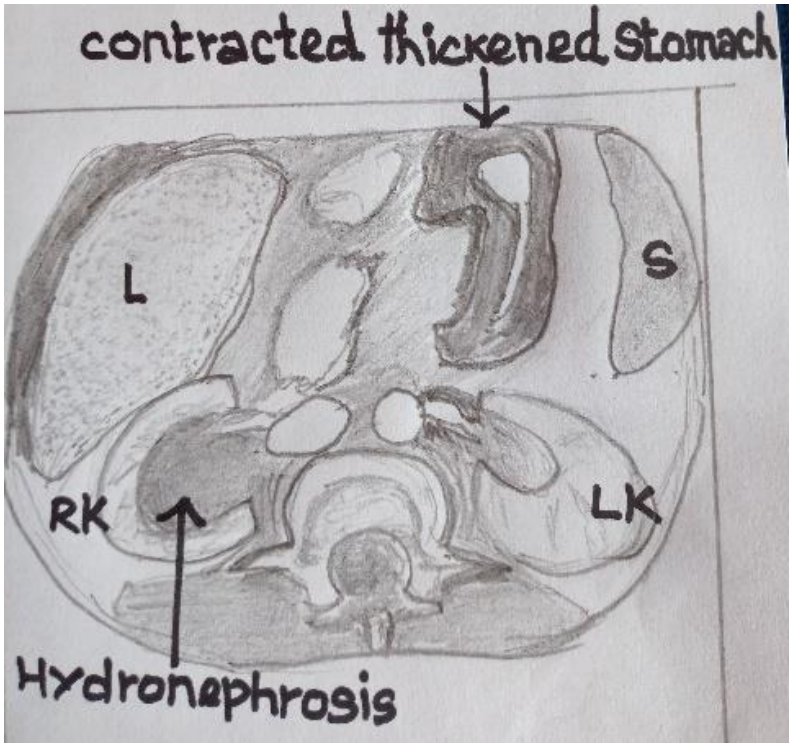
We have to know the current option on EUS. Is EUS-FNA of the gastric wall preferable than the conventional biopsy? Is EUS-FNA superior to conventional biopsy?

EUS is decisively useful in infiltrative gastric carcinoma. Endoscopic ultrasonography can assess the depth of penetration of the tumour or involvement of adjacent structures (lymph nodes etc). In this case, the thickness of the gastric wall as well. However, Ultrasonography and EUS are operator dependent. EUS is better than CT at assessing tumor depth (T stage) and perhaps lymph node involvement (N stage), particularly if fine-needle aspiration (FNA) is also performed. EUS-FNA superior to conventional biopsy. It is effective and safe as it reduces the complication of bleeding and perforation when a large or deep biopsy is warranted, However, regarding the yield and adequacy of biopsy, it's still slightly inferior to conventional biopsy (jumbo forceps, bite-on-bite technique). EUS-FNA mainly helps in diagnosing remote metastases, particularly if results may alter tumour staging and thereby the treatment received.

Once we arrived at the clinical diagnosis of Gastric carcinoma after correlation of all the above imaging findings, the most vital question is, how we will confirm the

diagnosis. Computed tomography scan and HPE of biopsy tissue are highly recommended.

Abdominal contrast-enhanced computed tomography was performed for this patient (diagram below). It suggested thickening of the wall of the gastric corpus. In addition, right hydronephrosis and a small amount of ascites fluid were detected in the abdomen. No enlargement of associated lymph nodes was detected.



We are almost reaching the final stage of our clinical approach in the diagnosis. Steps to help determine and identify the stages of stomach cancer may include:

- Imaging testing, such as CT scan
- Endoscopic ultrasound
- Laparoscopy
- Biopsies and HPE

According to the National Cancer Institute, the following stage groupings are used for staging stomach cancer:

Stage 0 carcinoma in situ, only in the mucosa; has not spread anywhere else.
Stage 1 The cancer is in mucosa and submucosa and has spread to up to six lymph nodes very close to the tumor. 0r the cancer is in imucosa and submucosa and has also invaded the muscle layer. It has not spread to lymph nodes or other organs.

Stage 2 The cancer is only in the submucosa and has spread to seven to 15 lymph nodes very close to the tumor or It is in the muscularis and subserosa and has spread to one to six nearby lymph nodes Or The cancer has gone through the outermost layer of the stomach wall. It has not spread to lymph nodes or other organs.
Stage 3 The cancer is in the muscle layer or subserosa and has spread to seven to 15 lymph nodes Or It has invaded nearby organs, such as the liver or spleen. Cancer cells have not spread to lymph nodes or to distant organs or It has penetrated through the subserosa and the serosa. The cancer has spread to one to 15 lymph nodes very close to the tumor.
Stage 4 The cancer has spread to nearby tissues and at least one lymph node, it has spread to more than 15 lymph nodes, or it has spread to distant parts of the body.

Final diagnosis based on the HPE findings of the biopsied tissues: Scirrhous Infiltrative 'Linitis plastica' type of gastric carcinoma with right urethral obstruction and metastasis to the peritoneal cavity causing ascitis.
The common microscopic features of Scirrhous Gastric Carcinoma show undifferentiated cancer cells or signet ring cells proliferate with abundant fibroblasts (fibrous tissues).

Special Note:
Linitis Plastica (LP): **LP is a** Scirrhous **distinct phenotype of primary gastric cancer (**SGC) **that often presents at an advanced stage, with a high rate of peritoneal involvement.**) is biologically aggressive, typically infiltrating into the gastric wall and accompanied by peritoneal metastasis LP, widely used term for Brinton's disease (also known as leather bottle stomach), is a type of adenocarcinoma. **a morphological variant of diffuse (or infiltrating) scirrhous stomach cancer.** It is also the term used to describe the condition of a rigid, non-distensible stomach which may be caused by

lymphoma and secondary metastases from particularly breast and lung carcinoma. It is not associated with H. pylori infection or chronic gastritis. The risk factors are undefined, except for rare inherited mutations in E-cadherin. The hereditary form of this cancer, Hereditary Diffuse Gastric Cancer, accounts for only 1–3% of gastric adenocarcinomas., Though not commonly associated with H. pylori infection or chronic gastritis, still mucosal atrophy may be found. A non-malignant condition such as a caustic injury to the stomach can be ruled out by definite history. . Majority of the cases of SGC are not detectable at an early stage because it dose not form a cauliflower like intraluminal mass, but, tumour cells migrate throughout the submucosa without severely affecting the mucosal lining of the stomach. LP grows rapidly and it is difficult to control it. Because of the rich lymphatic supply, the cancer rapidly disseminates beyond the reach of surgical resection. Imaging should be carefully scrutinised to see evidence of metastasis in the vessels, lymph nodes liver, lung, bone etc.

LP is a fatal disease. Despite recent advances in diagnostic techniques and therapies for GC in surgery, chemotherapy and radiotherapy, the prognosis of SGC remains poor. Sometimes the LP is not curable with gastrectomy and patients are on chemotherapy. But chemotherapy only reduces the occurrence of symptoms, does not cure the condition. Only 3 to 10% survival rate within five years

Research Note:

Researchers are continually working on new strategies for SGC methods including prevention, early detection and therapy based on its biological behaviour to improve the prognosis of SGC patients. Researchers are studying chemoprevention, methods of using diet and medications to lower the risk of getting cancer. They are studying the role of antioxidants such as vitamin C, beta-carotene, and vitamin E in cancer prevention. It appears that antioxidants may destroy free radicals, chemicals that cause genetic damage and cancer formation. Researchers are studying sentinel lymph node mapping methods. Lymph node mapping can indicate how far the cancer has spread through the lymph node system and identify cancerous lymph nodes for removal. Various studies are being done on the current development of diagnostic

techniques and therapy based on the findings of clinical and molecular investigations. In the area of immunotherapy, researchers are studying ways to boost a person's immune system to fight gastric cancer better. A novel target therapy based on the biological behaviour of SGC is imperative. Medications are also being studied that can detect fast and slow growing cancer cells. Researchers state that treating H. pylori infections with antibiotics appears to lower the risk of stomach cancer

Aspirin may lower the risk of stomach cancer and colon. Finally, researchers are looking for new ways to perfect the screening process for the early detection of stomach cancer. The concurrent recent accumulation of information of the molecular biology underlying the characteristics of SGC can facilitate in the establishment of a foundation for the diagnosis and development of treatment strategies for SGC.

Case 25

A 50-year-old male smoker presented at the tertiary health facility on self-referral. with complaints of a intermittent, chronic fever, cough, and shortness of breath. His fever was not continuous (fever free spells). He had Fatigue, Excessive drenching night sweats, unintended weight loss of 8 Kg within 6 months, with intermittent vague retrosternal discomfort for 6 months. He had left hypochondric and lumbar abdominal pain and low backpain. There was no previous history of abdominal surgery or reducible groin swellings. On general physical examination he was pallor, not cyanosed with no clubbing; He was febrile with a blood pressure of 139/87mmHg, pulse of 89 beats per minute, and respiratory rate of 22 breaths per minute. He was not having any obvious external evidence of infection except rashes on his chest, back (Fig.1), elbow and buttock and Painless bilateral cervical (Fig.2), axillary and inguinal lymphadenopathy. Examination of the extremities revealed 2+ pitting edema extending to thighs bilaterally. His abdomen was non-tender to palpation. He had no palpable peri-umbilical nodes. He was having palpable non-tender liver 1" below the right costal margin. There was massive splenic enlargement. On deep palpation of the abdomen There was however a distinctly palpable, hard, irregular solid mass occupying the mid-abdomen. The bowel sounds were normoactive. Laboratory values showed hemoglobin of 7.5g/dl with mean corpuscular volume of 71.2fl. His metabolic panel was significant for sodium of 134mmol/L. His liver function tests were significant for aspartate aminotransferase of 122U/L, alkaline phosphatase of 478U/L, and albumin of 2.3g/dl. No Mycobacterium Tuberculosis was detected on previous Gene-X-pert analysis of his sputum. Haematological and biochemical panels show Cytopenia, elevated CRP and LDH. His electrocardiogram showed T-wave inversions in leads V1–V4 and an S1Q3T3 pattern. On suspicion of Lymphoma initially a CXR was done (Fig 3).

The reader is invited to understand the importance of clinical history and features in this patient that help to primarily suspect an important disease, 'Lymphoma' which if undetected early becomes /serious or dangerous.

In such clinical background, while eliciting history, not to forget to include the following important information:

- type, pattern and duration of fever; **Fever** is one of several important symptoms of lymphoma. Pel-Ebstein **Fever, common in Hodgkin lymphoma, is a** particular pattern of **fever** named after two European doctors, Pieter Pel and Wilhelm Ebstein,
- nature of cough (dry or with lot of sputum/phlegm),
- hemoptysis;
- dyspnoea in rest or on exertion, orthopnoea;
- skin rashes, site, appearance, pattern, progression to papule/pustule, itchy, spread or static?
- elaborate on his smoking.
- weight loss: sudden or gradual, how many kilos etc.
- swelling in neck painful or painless, changing in size?

Note the alarming signs and symptoms in this patient:

1. the fever was intermittent, occurred and went off over several days or weeks without any obvious infection.
2. **Rashes** were mostly seen in the skin folds of elbow, chest, back, buttock. They appeared as reddish or purple scaly areas. (over the age of 55 years are at increased risk to developing lymphoma rash. A lymphoma rash, such as mycosis fungoides (MF), can be easy to confuse with other skin conditions, such as psoriasis or eczema, which can cause similar symptoms)
3. Painless swelling of the lymph nodes were seen and felt as **lumps under the skin**, in the neck above the clavicles), armpits and groin. An increase in the size, changed shape and consistency in the supraclavicular lymph node on the left with a high probability indicates the localization of the malignant process in the chest and or abdominal cavity. **Enlarged Supraclavicular Lymph Nodes** of Non-Hodgkin's **Lymphoma** Can Mimic Metastasis (of **Breast Cancer, ca lung, ca stomach etc)**
4. Unintended weight loss of 8 Kg within 6 months, (at least 10% of body weight over 6 months), Weight loss without trying, not on weight losing program

5. Excessive drenching night sweats even under the fan; drenching night sweats are often a manifestation of malignancy and, if persistent, should prompt the clinician to consider neoplastic disease.
6. When lymphoma starts in the thymus or lymph nodes in the chest, it may press on the nearby trachea, which can cause coughing, dyspnoea, or a feeling of chest pain or pressure.

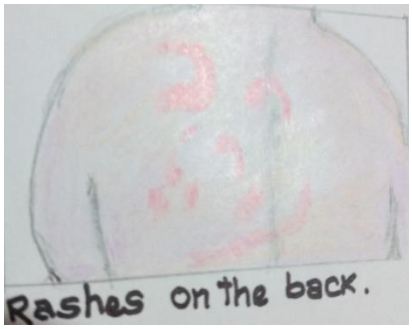
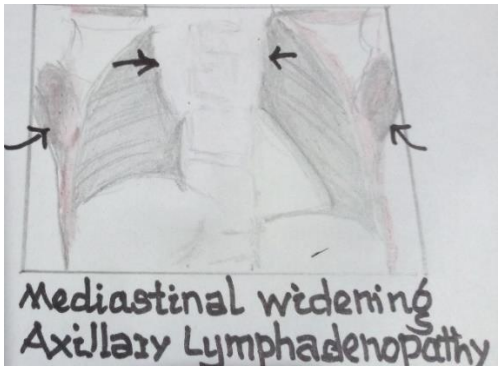


Figure.1

Figure.2: **Troisier's sign in the neck (Virchow's lymphnode)**



(Figure 3: The reader should be able to recognize this CXR and
appreciate the mediastinal widening and axillary lymphadenopathy
Note the upper mediastinal widening due to a mediastinal mass (10 cm × 6 cm) (arrows); the inferior edge of upper mediastinal mass the angles of which is obtuse. The superior border/margin of the mediastinal mass is not visible. The hilar vessels cannot be seen through the mass ("hilum overlay sign" absent). A positive cervicothoracic sign is present. **These three findings** confirm that this mass is located in the superior-anterior-middle mediastinum. Bilateral subcutaneous axillary lymphadenopathy can be detected.
The reader has to ponder over the question: What are the other clinical features to look for in such anterosuperior intrathoracic mass?

Ultrasonography of the neck and abdomen (Figure-4a and b) were performed.

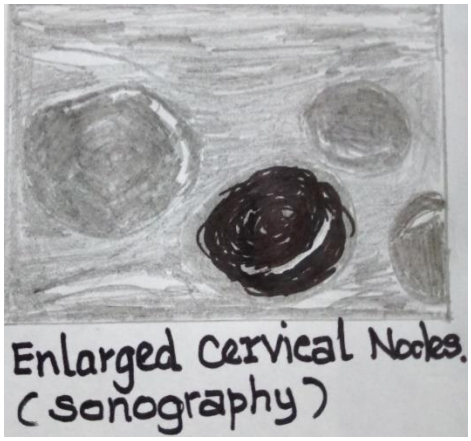


Figure **-4a.** Ultrasound image of cervical lymph nodes

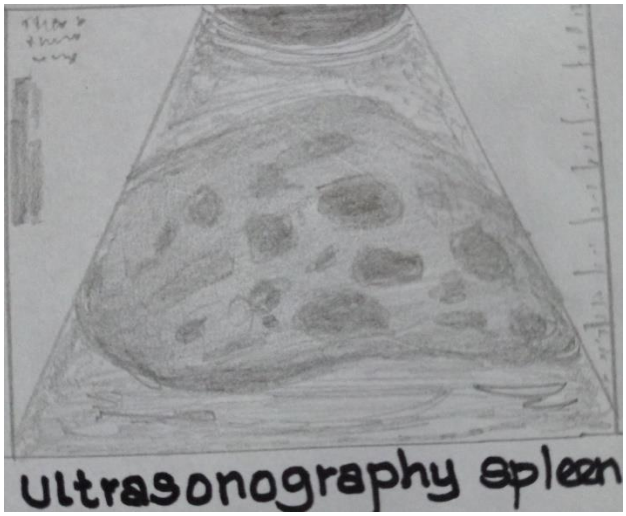


Figure **4-b-Ultrasonogrphy. Splenic-involvement**

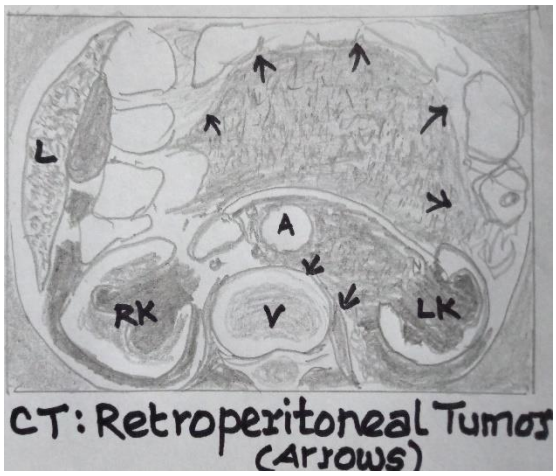


Figure-**5: CT: Retroperitoneal mass causing bilateral hydronephrosis**

The reader/student should take keen interest to appreciate the following points in Figures 4-5 illustrated above:

- **Splenomegaly**. The upper limit of normal adult splenic length is traditionally cited at 12 cm, but lengths upwards of 14 cm can be seen in normal, taller males… Soft tissue density of the enlarged spleen can be very well seen on ultrasonography (US). Associated hepatomegaly should be looked for.

- **Massive splenomegaly** is variably defined, including when the spleen is 5 standard deviations above the mean normal volume (about 943 cm^3) [4], heavier

than 1.0 kg or 1.5 kg, longer than 18 cm or extending into the pelvis or across the midline. This patient has moderate splenomegaly.

- The enlarged spleen in lymphoma may have foci of infarction, necrosis, infection, abscess, infiltration and calcification.
- Ultrasonography of cervical lymph nodes (LN) show the alteration of the size, shape, architecture and vasculature. The importance of transverse scan of LN should be realized. Vertical diameter of LN should not exceed its transverse diameter. The loss of hilum and roundness of LN are important abnormal findings.
- In multifocal splenic involvement in lymphoma US image shows several small hypoechoic splenic deposits in a patient with histologically proven lymphoma
- **Figures 5:** CECT demonstrated the huge retroperitoneal mass and the retroperitoneal position is said to be the most common location of lymphoma. The retroperitoneal lymphoma has been noted to have greater tendency for forming composite soft tissue encasing the major vessels.
- Encasement of Aorta and left renal vein. Entrapped left ureter causing hydro-ureter and hydronephrosis (Figures 5).

As patient could not afford the cost for further imaging investigations, excisional Biopsy of the cervical LN was done and the final diagnosis of Non-h*odgkin's lymphoma* was confirmed.

The mainstay of treatment of NHL is non-operative multimodal therapy and regardless of location, NHL is highly responsive to cytotoxic chemotherapy, except those with high grade biological profile. Early stage, slowly growing retroperitoneal NHL can be treated successfully with radiotherapy, especially, the nodular or follicular type. External beam radiotherapy however may result in debilitating retroperitoneal fibrosis, radiation enteritis or pyelonephritis. The first line chemotherapy for diffuse B cell lymphoma is Rituximab in combination with Cyclophosphamide, Doxorubicin, Vincristine and Prednisone (R-CHOP). Surgical resection is deployed in the treatment of retroperitoneal lymphoma, especially, a diffuse B-cell lymphoma. with a huge mass lesion with extrinsic compression of

adjoining organs. This may involve an En bloc resections of the involved organs. A microscopically free margin (R0) resection is the goal of surgical resection.

The reader/student should try to answer the following:

- What are the different levels of cervical lymph nodes?
- Differential diagnosis of enlarged supraclavicular lymph nodes
- What are the other imaging investigations recommended in lymphoma?
- What is SVC and IVC syndrome? Did this patient have IVC syndrome?
- Discuss on the best available staging system for lymphoma
- Discuss the risks and complications of retroperitoneal lymphoma
- What is primary retroperitoneal tumour?
- When is an MRI may be preferable over CT scan in patients with lymphoma

Which of the following is not indicative of a primary retroperitoneal tumour?

- Anterior displacement of retroperitoneal structures such as the renal vein, the inferior vena cava (IVC), and the duodenum.
- Crescentic deformation of the IVC.
- Rounded edges of adjacent solid retroperitoneal organs such as the kidneys.
- Distortion of the renal parenchyma into a beaked configuration.

NOTES TO THE READER

The CT diagnosis of diseases in the retroperitoneal lymph nodes is based mainly on an evaluation of the size of the nodes in the transverse plane. Opinions on the normal size of the nodes vary, however. Obtaining the diameters of the lymph nodes on lymphograms, US scan, CT scan may slightly differ. The upper limit for the diameter is not the same for all lymph nodes, but varies with the position of the node in the para-aortic chains, ranging from 7 to 15 mm, with increasing diameters in the caudal direction. The transverse diameter of abdominal lymph nodes should not exceed 1.5 cm. The product: transverse diameter X antero posterior diameter should not exceed 2cm.

More nodes may be seen on MRI than on CT. MRI criteria for normal retroperitoneal and pelvic lymph node size are , the 95th centile values for maximum short axis diameter (MSAD) of pelvic lymph nodes are common iliac and obturator 4 mm, external and internal iliac 5 mm and hypogastric 6 mm. In the retroperitoneum the 95th centile MSAD values are retrocrural, high left para-aortic, paracaval and interaortocaval 3 mm, post-caval 4 mm and low left para-aortic 5 mm.

Takeaway

Ideally, a lymphoma diagnosis requires a lymph node or involved-tissue biopsy. We may also need blood tests and imaging tests. The official evidence based confirmed diagnosis will allow to create the best treatment plan for our needs.

RESEARCH /SELF STUDY ACTIVITIES:

1. Is Follicular lymphoma (FL) common?
2. Is mantle cell lymphoma prevalent in our region? If not, which one is more prevalent?
3. In people with HIV in Malaysia, which is the most common type of NHL?
4. **Is t**he PET scan advantageous in the diagnosis of lymphoma?
5. Which grade of lymphoma have the risk of local recurrence rather than distant metastases?
6. Researchers are studying chemotherapy with autologous or allogeneic hematopoietic stem cell transplantation (HSCT) for the treatment of individuals, particularly younger individuals, who have relapsed or refractory follicular lymphoma.
7. Research is underway to develop additional anti-CD20 monoclonal antibodies for treating follicular lymphoma. These medications would be for individuals who did not respond to therapy with rituximab or obinutuzumab, or could not tolerate those medications, or that may prove more effective.
8. Additional medications that are being studied for relapsed follicular lymphoma include ibrutinib (Imbruvica®), lenalidomide (Revlimid®), BCL-2 inhibitors, and inhibitors of programmed cell death protein 1 (PD1) or PD1 ligand 1

(PDL1). More research is necessary to determine the long-term safety and effectiveness of these therapies.

9. Drugs are also being studied that can stimulate the immune system to act in order to enhance the effectiveness of rituximab. These drugs are sometimes called immunostimulatory drugs.

Disclaimer

Case-26

A 41-year-old obese female nonsmoker presented with history of nonspecific abdominal pain The pain was in the epigastric region radiating to the back. No history of intolerance to fatty food, nausea or vomiting, diarrhoea or staetorrhoea, melena, or bleeding. Her blood parameters showed hemoglobin of 11.8 g/dl, mean corpuscular volume 92 fL, total leukocyte count of 7.7 x 10^9/L with 58% neutrophils, 9% monocytes and 22% lymphocytes, 10% eosinophils and platelet count of 367 x 10^9/L. The family doctor advised her a plain x-ray of the abdomen. (AXR) Fig-1.

Fig:1 Plain x-ray of the abdomen AP view, subhepatic arrows point to a few nodular opacities suprajacent to the right floating rib suggestive of multiple gall stones (GS).

The reader/student has to ask, whether all GS will be casting shadow on plain x-rays of abdomen.

Majority of the gallstones are radiolucent. Only a small percentage of gallbladder calculi are visible on radiographs (~10%). The number, size and shape appearing on the plain x-ray of the abdomen are not the true nature of the gall stones as we are able to see only the radiopaque part of the calculi. Although a larger proportion of gall stones are visible on CT, ultrasound remains the best first-line imaging test for identifying calculi in the gallbladder.

As there is a strong possibility of cholelithiasis, the family doctor further advised her sonogram of the abdomen (Fig-2) …….

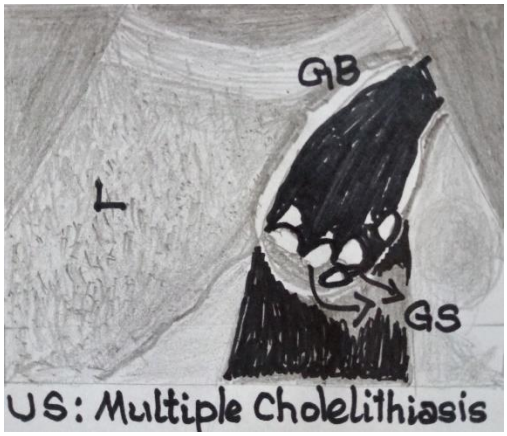


Fig 2. Ultrasound image shows multiple hyperechoic stones (arrow) in the lumen of the gallbladder (Labelled) with obvious posterior acoustic shadowing

The reader has to understand that sometimes the calculi move causing pain that may radiate to the shoulder; may move down and block the cystic duct, common bile duct and or ampulla of Vater, may even enter into the pancreatic duct, blocking it, may lead to pancreatitis. We have to look also for evidence of cholecystitis whenever there is cholelithiasis.

The attending physician got a second opinion from the Radiologist on the AXR and ultrasonography. who mentioned about the multiple tiny, mobile particles changing their position and finally settling in the dependent position of the lumen of the GB (sludge formation). This indicated stasis of bile in the GB due to previous or current infection. As the patient was only mildly symptomatic, she ignored the surgical treatment suggested by the family doctor and got back to home. After two years, the patient presented herself to the tertiary hospital for admission with acute severe abdominal pain, progressively increasing breathing difficulty, distended abdomen, a weight loss of 5 kilograms, history of frequent indigestion and altered bowel habits on and off. She had a dry cough and increased dyspnea on lying down partly relieved by sitting up. There was loss of appetite, nausea and vomiting. Two days before this admission, she had upper abdominal pain with fever. There was no history of trauma to the chest and abdomen.

On admission, she was coherent, febrile, had mild pallor, no lymphadenopathy, clubbing, cyanosis, mild icterus, pulse was 98/min, regular, adequate volume, and

blood pressure was 110/70 mm Hg. Signs of chronic liver disease, such as palmar erythema, spider nevi, and parotid glands enlargement, were not detected. The examination of respiratory system revealed tachypnea (respiratory rate 28/min), stony dull note on the left lower cheat, absent vocal fremitus, and absent breath sounds on the left. Abdominal examination revealed diffuse tenderness, epigastric rigidity and massive ascites with no palpable mass. Liver and spleen span were normal. Examination of rest of the systems was normal.

Her biochemical tests showed blood urea 15.7 mmol/L, serum creatinine 53.04 µmol/L , serum sodium 137 mmol/L, serum potassium 4.1 mmol/L, serum calcium 2.1 mmol/L, serum thyroid-stimulating hormone 1.96 mIU/L, serum 10.65 ukat/L, serum lipase 19.2 ukat/L, and serum lactate dehydrogenase (LDH) 386.4 U /L. The liver function test revealed serum bilirubin 16.58 µmol/L, alanine transaminase 20.5 U/L, aspartate transaminase 116.8 U/L, alkaline phosphatase 134.2 U/L, total proteins 55.8 g/L, serum albumin 24 g/L, serum globulin 31.8 g/L and INR 1.82. Her lipid profile was within normal limits except for low high-density lipoprotein cholesterol (HDL-c) which was 0.818 µmol/L. Serum C-reactive protein (CRP) was 2100 mg/L at 48 hours after the onset of symptoms. Her urine routine examination was normal, and culture was sterile. Her viral markers (hepatitis B surface antigen, anti-hepatitis C virus antibodies, anti-HIV l and ll antibodies) were nonreactive. Her antinuclear antibody and rheumatoid arthritis tests were negative. Stool for occult blood was negative. A set of Radiology imaging were done for her including AXR, CXR, ultrasound abdomen and an urgent CT scan of abdomen to confirm the clinical diagnosis of acute pancreatitis.

The reader must recognize in the AXR (Fig 3] an important "Colon cut off" sign. Distended transverse colon, abrupt absence of gas at Splenic flexure with narrowing and Collapse of descending colon starting from Splenic flexure.

Fig 3: AXR focussed view shows colon cut off sign.

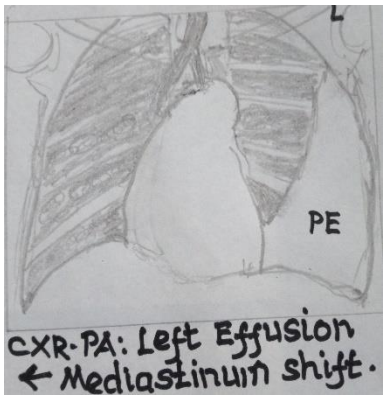


Fig 4: Chest PA Xray film showing large left sided pleural effusion with mediastinal shift to the right

Her transabdominal US scan showed a large swollen heterogeneous, pancreas involving more of head and partly the body compatible with evolving necrosis. **(**acute pancreatitis). There was inflammatory hypoechoic fluid collection in the supra-pancreatic, peripancreatic and left anterior pararenal spaces due to acute pancreatitis.

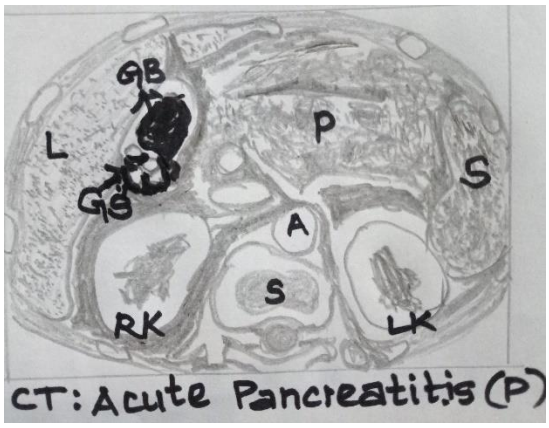


Fig 5: Acute necrotizing pancreatitis. CT shows heterogeneous, non-enhancing parts of pancreatic head, neck, and body. Note, multiple stones in the gallbladder. Also present is the peripancreatic inflammation fluid extending up to the splenic region.

Left-sided underwater seal intercostal drainage tube was inserted after consulting chest physician and gastroenterologist. **Hemorrhagic pleural and ascitic fluid** *were tapped on the 1st day of admission. The fluid specimen was sent for biochemical and pathology examination. No pathogenic organisms were isolated. Ziehl–Nelson staining did not show AFB.*

She received octreotide for 2 weeks, in addition to piperacillin-tazobactam 4.5 g thrice a day intravenously. Her intercostal tube drainage gradually decreased

A magnetic Resonance imaging evaluation was planned. However, patient refused and on the 20th day of admission, left the hospital against medical advice due to financial constraint.

FINAL DIAGNOSIS:

Acute biliary necrotizing Pancreatitis

Discussion:

Diagnostic Assessing of Etiology, Imaging & Severity

The standard work-up of the cause of acute pancreatitis include imaging tests (Ultrasonography, Endoscopic Ultrasonography, Computerised Tomography, Magnetic Resonance Imaging, Magnetic Resonance Cholangiopancreatography, and Endoscopic Retrograde Cholangiopancreatography) mainly driven by individual preferences. However, based on current available evidence and recommendations

according to established guidelines, an abdominal US is advised in all patients presenting with acute pancreatitis, both at first presentation and in recurrent episodes of otherwise idiopathic pancreatitis. Depending on expertise, availability, and local practices, further testing by means of EUS or MRCP is indicated as a next step if US is negative but the clinical suspicion for a biliary etiology is high. Additional imaging (i.e., state-of-the-art multidetector CT, EUS, and/or MRI/MRCP) is especially warranted in patients over 40-50 years of age with "idiopathic" acute pancreatitis or repeated episodes of acute pancreatitis to exclude a pancreatic neoplasm as possible cause of the pancreatitis.

Acute pancreatitis is a serious disease with varying severity. The recently revised Atlanta Classification 2012 on acute pancreatitis (RAC) classified the severity of acute pancreatitis clinically (on the basis of presence or absence of organ failure) and morphologically (on the basis of presence or absence of tissue necrosis). Morphologically (i.e., on imaging), two types of pancreatitis are discriminated: interstitial pancreatitis (no tissue necrosis) and necrotizing pancreatitis (tissue necrosis).

Interstitial pancreatitis:

Interstitial pancreatitis is usually a self-limiting disease with a short hospitalization stay and represents the most common form of acute pancreatitis (46, 53). These patients typically recover uneventfully without complications. On imaging, interstitial pancreatitis may reveal a minimal increase in size of the pancreas, focally or diffusely. Morbidity from interstitial disease ranges about 10% with mortality less than 3%, primarily due to co-morbid disease (95). Our case here is **Necrotizing pancreatitis**

Necrotizing pancreatitis:

Necrotizing pancreatitis is associated with a protracted clinical course, long hospital stay with a high morbidity (30-80%), and a mortality rate up to 20-30%. The 2012 revised Atlanta Classification distinguishes three subtypes of necrosis depending on involvement of pancreatic parenchyma alone (rare), peripancreatic tissues (extrapancreatic necrosis or EXPN, more common), or the combination of both (combined necrosis, most common). Pancreatic parenchymal necrosis tends to occur early in the course of the disease, within the first 48-72 h after symptom onset. CT

criteria for the diagnosis of pancreatic parenchymal necrosis are dependent on the detection of areas lacking enhancement, which may be focal or diffuse. Lack of pancreatic enhancement corresponds with decreased blood perfusion of the pancreatic gland and correlates well with necrosis. Pancreatic parenchymal necrosis is ideally detected on scans performed >72 h after the onset of an attack of acute pancreatitis. Scans done within this timeframe may be falsely negative or equivocal.

Biliary pancreatitis:

To make this diagnosis a biliary component must exist as a causative factor. The diagnosis is straightforward when gallstones are seen at abdominal US; gallstones appear as intraluminal, echogenic, mobile foci that are gravity-dependent and create a clean posterior acoustic shadow. A repeat abdominal US is advised in those with "idiopathic" acute pancreatitis as gallstones may be missed or migrated on the initial evaluation. Because of the superior sensitivity an abdominal US should be performed in every patient presenting with acute pancreatitis early in the disease course to rule out gallstones as possible etiology. However, acute biliary pancreatitis may also be due to microlithiasis or biliary sludge (defined as stones smaller than 2 mm), which can be difficult to diagnose by abdominal US, but may be responsible for recurrent episodes of acute pancreatitis. Biliary sludge is a viscous suspension of bile fluid that includes small stones, cholesterol monohydrate crystals, or calcium bilirubinate particles. Most patients who have biliary sludge are asymptomatic. Yet, biliary sludge is detected with increasing frequency in patients who have acute, otherwise idiopathic, pancreatitis. On CT gallstones appear as single or multiple filling defects within the gallbladder. Gallstones may have varying densities on CT depending on the composition. Stones may be densely calcified, rim calcified or laminated or have a central nidus of calcification. Stones also may present as a soft-tissue density or a lucent filling defect within the bile. Some stones may contain gas. In about 25% of cases, stones are isodense to fluid and therefore not identifiable on CT (99). Both cystic and common bile ducts must be examined closely to pick up migrated gall stones. MRI is an excellent, but costly alternative for US for depicting stones (larger than 4-5 mm) in the gallbladder or common bile duct.

Cross-sectional imaging may show ‘choledochal ring’ sign, defined as hyperenhancement of the common bile duct wall relative to the pancreatic parenchyma (difference of more than 15 HU), has been reported to be indicative for a biliary cause of acute pancreatitis Some other CT features were significantly associated with biliary pancreatitis, including pericholecystic fluid or fat stranding, pericholecystic increased attenuation of the liver, increased gallbladder wall enhancement, and gallbladder wall thickening. it’s the author’s opinion that initial severity assessment should be based on clinical scoring systems and correlated with imaging parameters.

Vascular Complications

The reader must be aware of vascular complications arising from acute pancreatitis that include portosplenomesenteric venous thrombosis, arterial pseudoaneurysms, and hemorrhage due to vessel wall erosion by extravasated proteolytic pancreatic enzymes, commonly involving the splenic artery, the pancreaticoduodenal or the gastroduodenal arteries, which are closely related to the pancreas. Splenic vein thrombosis occurs most common and may result in complications such as gastric or esophageal varices and splenomegaly (left-sided portal hypertension). Massive acute hemorrhage secondary to bleeding pancreatic collections or arterial pseudoaneurysm has an associated mortality rate of 10-35%. An easily overlooked complication on abdominal CT in patients who are bedridden because of their illness (i.e., not unique to acute pancreatitis) is the occurrence of deep vein thrombosis in the iliacofemoral veins that may lead to pulmonary embolisms. In contrast to portosplenomesenteric vein thrombosis, this finding urgently necessitates the initiation of anticoagulant treatment**.**

Synopsis of management

The presence of extended necrosis (more than 30%), especially in the midgland is associated with increased need for intervention; signs of infected necrosis requires empirical antibiotics or invasive intervention; massive hemorrhage or detection of an arterial pseudoaneurysm necessitates for angiographic coiling or surgery; deep vein thrombosis or detection of pulmonary emboli is an indication for anticoagulant therapy; acute cholecystitis needs percutaneous drainage or cholecystectomy, bowel ischemia or perforation is an indication for emergent surgery; and findings of ACS requiring

percutaneous drainage of ascites or surgery. Gallstone-induced acute pancreatitis is a prevalent condition that is associated with an unacceptably high mortality rate. Early endoscopic intervention, including endoscopic retrograde cholangiopancreatography and endoscopic sphincterotomy within 24 to 72 h of hospital admission, can be used to remove stones in the common bile duct (CBD) and establish biliary drainage. ERCP is usually successful in 95% of patients, and CBD stones can be detected and removed in most of them. The morbidity associated with the endoscopic procedure was 3-5%, and no death is expected.

RESEARCH / FURTHER SELF STUDY ACTIVITIES

The student must think….and explore the following areas:

- What is the evidence based final diagnosis?
- Colon cut-off sign and its clinical significance?
- What is the cause of pleural effusion? Can it recur? Can it be very massive?
- Why is there ascites?
- Why both pleural and ascitic fluid are hemorrhagic?
- If pleural effusion is recurring very quickly, what to do?
- Significance of Sentinel loop, colon cutoff sign
- **Causes of elevated serum amylase and lipase, amylase-lipase ratio and its significance**
- When to do CT abdomen?
- When to do other special imaging tests?
- What are the differential diagnoses?
- Acute vs. chronic pancreatitis: Symptoms and treatments
- How to lower our risk of future pancreatitis attacks?
- What is the ICD 10 code for acute pancreatitis?

Disclaimer

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this do not constitute legal or other professional advice on any subject matter. *Please consult with your own resources, physician or healthcare specialist regarding the suggestions and recommendations made in this to determine what may be best for your individual needs.* Information and illustrations are taken from many (re)sources and are duly acknowledged. The information contained within this article does not refer to a particular patient. *Although the author has made every effort to ensure that the information in this paper was correct at press time in regard to the subject matter covered, the publisher and the author assume no responsibility for errors, inaccuracies, omissions, or any other inconsistencies herein.* The contents of this links in any form of reproduction, permanent storage, or retransmission are subjected to the copyrights of them and recommended to follow them. The use of this article implies your acceptance of this disclaimer

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Case 27

A 2-year-old male presents with petechiae and increased bruising for the past 2 weeks, initially on both his legs, then spreading to his abdomen and arms. There is no history of trauma. Mother mentions that he had fever and cough about 4 weeks ago that resolved over several days. He does not have gingival bleeding, nose bleeds, or joint swelling. He has not consumed any traditional medication.

On physical examination, he appears well but has petechiae and bruises of variable stages of healing over both legs, arms and trunk. He also has some buccal petechiae. Vital signs are stable. There is no lymphadenopathy or hepatosplenomegaly. Other systemic examinations are unremarkable.

His full blood count reveals: white cell count 7×10^9/L, haemoglobin 13 g/dL, haematocrit 37% and platelet count 5×10^9/L. Peripheral blood film shows that red blood cells and white blood cells are normal in morphology. No blast cells seen. The platelets are reduced and appear big (11.4 fL) and no abnormal clumping seen.

Differentiating petechiae and purpura

Petechiae are pinpoint non-blanching spots
Purpura are larger non-blanching spots that are > 2mm
Both these lesions do not blanch when pressure is applied - this is in contrast to other common rashes in children such as viral exanthemas and urticaria.

Glass test can be used to differentiate whether a rash is blanching by applying a slide glass firmly against the rash. If the rash does not disappear it is non-blanching.

Questions

1. What is the most likely diagnosis in this child?
2. What is its pathogenesis?
3. What is the treatment?

Looking at the presentation of this child with preceding history of respiratory tract infection, with the physically examination revealing a well-child, with no lymphadenopathy or hepatosplenomegaly but with only thrombocytopenia associated

with large platelets seen on the peripheral blood film, the diagnosis is most likely immune thrombocytopenia purpura.

Other possible diagnosis are:
1. Henonch scholein purpura (HSP), but the classical findings of HSP is palpable purpura over the extensor surface of the lower limbs with arthralgia or arthritis which are not present in this child
2. Acute leukemia can be a differential diagnosis but usually the child will be more ill with recurrent fever, bone pain, lymphadenopathy and hepatomegaly. In the blood investigation, it commonly shows involvement of more than one cell line.
3. Meningococcaemia; it is crucial to be able to identify and exclude meningococcaemia. However, in meningococcaemia, the presentation will be more acute and the child will be toxic-looking. have high grade fever, and the full blood count will show leukocytosis in addition to thrombocytopenia.

One of the most common causes of a low platelet count is pseudothrombocytopenia from platelet clumping. In about 1% of individuals, ethlenediaminetetra-acetic acid (EDTA) (purple-coloured cap bottle), the anticoagulant causes platelet clumping, resulting in pseudothrombocytopenia. In this instance, the blood sample for full blood count should be collected in citrated bottle (blue-coloured cap bottle)
Thrombocytopenia is defined as a platelet count of less than 150×10^{9} /L at essentially at any age

Platelet size may also help determine the cause of thrombocytopenia. Tiny platelets (mean platelet volume [MPV] <7 fL) are seen in Wiskott-Aldrich syndrome and X-linked thrombocytopenia. Giant platelets (MPV >11 fL) are seen primarily in Bernard-Soulier syndrome. Most other causes of thrombocytopenia will result in platelets with a normal platelet size and volume, although thrombocytopenia resulting from increased platelet turnover often results in a modestly increased MPV (big platelet).

Platelet cells normally have a life span of 9 to 10 days, and younger platelets are generally larger than older platelets. A mildly elevated mean platelet volume performed by an automated counter suggests a destructive process because a predominance of young platelets would increase the average size

The diagnostic criteria for Immune Thrombocytopenia Purpura

ITP is a diagnosis by exclusion. ITP has been defined by an International Working Group (IWG) consensus panel as a platelet count less than 100×10^9 /L in the absence of another disorder known to cause thrombocytopenia.

In practice, the diagnosis of primary ITP is made when a well-looking, otherwise healthy child presents with acute-onset bleeding symptoms, such as petechiae, purpura, epistaxis and, rarely, severe hemorrhage, without other findings, such as congenital anomalies / dysmorphism, lymphadenopathy, hepato-splenomegaly and other cytopenias. Secondary ITP can be diagnosed in a patient with isolated thrombocytopenia who is found to have an underlying associated condition such as HIV infection, hepatitis C virus (HCV) infection, or systemic lupus erythematosus (SLE).

The pathophysiology of immune thrombocytopenia

The pathogenesis of ITP is multifactorial. Primarily, ITP is caused by the production of autoantibodies against platelet antigens (specifically glycoprotein IIb/IIIa and Ib/IX).

Autoantibody-coated platelets induce Fc receptor-mediated phagocytosis by mononuclear macrophages, primarily but not exclusively in the spleen.The spleen is the key organ in the pathophysiology of ITP, not only because platelet autoantibodies are formed in the white pulp, but also because mononuclear macrophages in the red pulp destroy immunoglobulin-coated platelets

Platelet autoantibodies primarily mediate platelet destruction but have been shown to also impair platelet production by binding to megakaryocytes.

Classification of ITP

ITP: within 3-month from the diagnosis

Persistent ITP: between 3 to 12 months from diagnosis

Chronic ITP: lasting more than 12 months.

Treatment for ITP

For most affected children, ITP is a benign, self-limiting disease process, with low risk for serious bleeding.

In children with no bleeding or skin manifestations only, observation is the recommended therapy, regardless of platelet count.

Treatment is generally indicated if there is:

i. Life threatening bleeding episode (e.g. intracranial haemorrhage) regardless of platelet count.
ii. Platelet count < 20 x 109/L with mucosal bleeding.
iii. Platelet count < 10 x 109/L with any bleeding.

Treatment of ITP

Oral Prednisolone 2 mg/kg/day for 14 days then taper off over 5 days (regardless of response) (maximum daily dose 80mg)

- Oral Prednisolone 4 mg/kg/day for 3 - 4 days (maximum daily dose 200mg)
- IV Immunoglobulin (IVIG) 0.8 g/kg/dose for a single dose, round up to nearest bottle to avoid wastage

Treatment if the child presents with severe bleeding or intracranial hemorrhage (ICH)

The incidence of ICH is very low in children with ITP, it is around 0.1-0.5%. The risk is highest with platelet count < 20 x 109/L, history of head trauma, aspirin use and presence of cerebral arteriovenous malformation.

Emergency treatment of ITP with severe bleeding, i.e. severe epistaxis or gastrointestinal bleed causing drop in Hb or ICH includes:

IV Methylprednisolone 30 mg/kg/day for 3 days.
IVIG 0.8g - 1g/kg as a single dose – calculated to nearest bottle of IVIG.
Platelet transfusion 8 - 12 units/m2 BSA (2 to 3 fold more than usual number of units)
Consider emergency splenectomy if other modalities fail.
Neurosurgical intervention maybe indicated in ICH for clot evacuation or extradural drains insertion.

Splenectomy may be performed concurrently if patient is going for neurosurgical intervention.

Splenectomy:

- Splenectomy is rarely indicated in childhood ITP.
- Splenectomy should be avoided if at all possible before 5 years of age and within 1 year of disease onset.
- Before considering splenectomy, reassess the diagnosis of ITP by excluding alternative diagnoses, including inherited thrombocytopenia, bone marrow failure, drug-induced thrombocytopenia, immunodeficiency syndromes (e.g. common variable immune deficiency [CVID]), myelodysplastic syndrome and other connective tissue diseases.
- Usually 6-weeks prior to splenectomy vaccination with pneumococcal; H influenza type B; and meningococcal vaccines. Pneumococcal booster should be given every 5-years.
- Post splenectomy the child will be given penicillin prophylaxis for life.
- Up to 70% of patients may achieve complete remission after splenectomy.

Platelet transfusions in ITP can transiently raise the platelet count in many patients with ITP. However platelets should not be routinely given in ITP, as most patients with ITP are not at risk for major haemorrhage, even without platelet support. In the event of life-threatening haemorrhage or intracranial hemorrhage (ICH), platelet transfusion may be used.

Preferred is random platelet transfusion rather than apharesis platelet transfusion.

Second-line treatment for ITP

Eltrombopag is indicated for treatment of thrombocytopenia in patients with chronic ITP who have shown insufficient response to corticosteroids, immunoglobulins, or splenectomy. This drug was approved by the FDA in 2008. In August 2015, the FDA expanded the indication for eltrombopag to include treatment of chronic ITP in patients 1 year of age and older who have not achieved an appropriate response with other medical therapy or splenectomy. The Joint Working Group guidelines place a priority on Thrombopoietin-receptor agonists (TPO-RAs)

as second-line treatment with rituximab being considered a third-line agent reserved for patients who have failed a TPO-RA.

Eltrombopag increases platelet production to maintain haemostatic platelet levels by uniquely binding to the transmembrane domain of the thrombopoietin (TPO) receptor. This stimulates differentiation and proliferation of cells in the megakaryocyte lineage. Eltrombopag is known to interact with polyvalent cations in certain foods, drinks and medicines. This interaction can significantly impair the absorption of eltrombopag into the body. Polyvalent cations to be avoided include calcium, aluminium, iron, magnesium, selenium and zinc as these can significantly reduce the absorption of eltrombopag. Eltrombopag must be administered at least 2 hours before or 4 hours after polyvalent cation-containing antacids, dairy products (or any foods, drinks, or medicines containing ≥50 mg calcium) and other products containing polyvalent cations, such as mineral supplements.

Advice before discharge

Precaution with physical activities especially small children.

- Avoid contact sports.
- Seek immediate medical attention if significant bleed or signs and symptoms of intracranial bleed. To explain to parents to look for headache, persistent projectile vomiting, blurred of vision.
- Avoid aspirin /NSAIDs and any over-counter medication.
- Avoid hot rub like methyl-salicylate rub.

Case 28

Examination of the Newborn

Mrs. Ong just delivered her first born male boy by elective LSCS. She is excited to go home as soon as possible. She is asking:

- Is my child healthy?
- When can we be discharged from hospital?

Question

1. What assessments would you need to do in order to be able to answer these questions?

In addition to her antenatal history, birth history and past family history, the doctor should obtain further information as follows:

<u>To decide if fit for discharge, the certain criteria should be fulfilled:</u>

The baby is able to breast feed well and mother able to take care of the baby

Normal physical examination for the baby

All relevant screening results (G6PD enzyme and TSH result) traced and checked

Hearing assessment (otoacoustic emissions test) cariied out

Pulse oximetry test:done: differential cyanosis detected by placing pulse oximetry on the right hand and the foot between 24-48 hours. Lower saturation in the leg than in the right hand suggest R to L shunt. (Currently not compulsory in Malaysia)

The baby has bladder and bowel open.

Received the relevant injection and immunization. In Malaysia all newborn will receive IM vitamin K , IM hepatitis B vaccine and also intradermal BCG vaccination

Assess gestational age as soon as possible if premature baby. (to be discharged only when weight more than 1.8kg)

Assessment of the baby would include the following:

- Baby's general condition & anthropometry

including colour, breathing, behaviour, activity, posture and cry.

anthropometry

vital signs

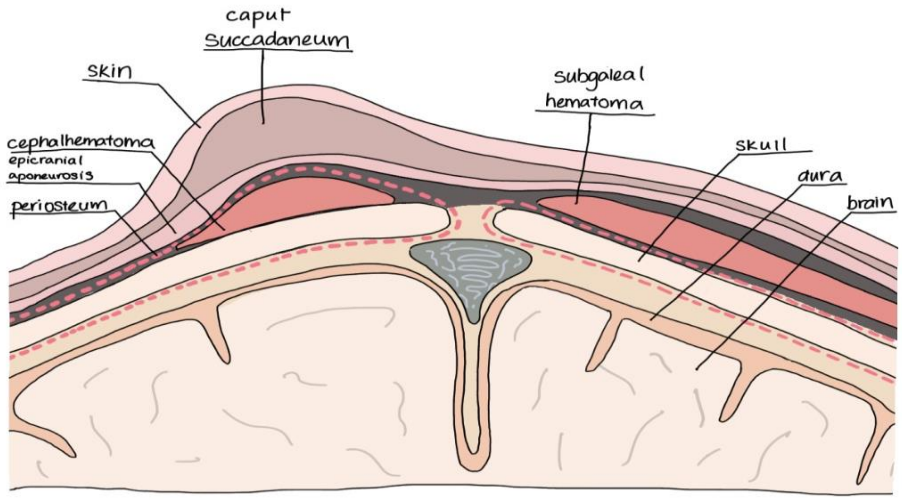
- Head and skull.

Size, shape and suture line

Some common findings on examination of head and skulls of a new born baby	
Cranial moulding	An abnormal head shape that results from pressure on the baby's head during childbirth. It resolves within few days
Caput succedaneum	A diffuse subcutaneous fluid collection with poorly defined margins (often crossing suture lines) caused by the pressure on the presenting part of the head during delivery. It does not usually cause complications and resolves over the first few days
Cephalhaematoma	A subperiosteal haemorrhage which occurs in 1-2% of infants. The haemorrhage is bound by the periosteum, therefore, the swelling does not cross suture lines (in contrast to a caput succedaneum). Cephalhaematoma is more common with instrumental delivery and may cause jaundice, therefore, bilirubin should be monitored and mother should be advice to bring the baby to healthcare provider once the baby appears jaundiced.
Subgaleal hemorrhages	It occurs between the aponeurosis of the scalp and periosteum and forms a large, fluctuant collection which crosses suture lines. They are rare, but may cause life-threatening blood loss. Usually the baby will be admitted for monitoring of head circumference and signs & symptoms of active bleeding.

Craniosynostosis	A condition in which one or more of the fibrous sutures in an infant skull prematurely fuses, changing the growth pattern of the skull which can result in raised intracranial pressure and damage to intracranial structures. Surgical intervention is required with the primary goal being to allow normal cranial vault development to occur by excision of the prematurely fused suture

Illustration of the conditions mentioned above



- Fontanelle

  Anterior fontenelle is about 3-6cm in diameter and posterior fontenelle not larger than 1.5cm

- Skin

  Colour – pallor / cyanosis / erythema / jaundice/ cutis mamorata

  Bruising / lacerations may be secondary to trauma during childbirth

  Facial birthmarks: Salmon patch, port wine stain, café-au-lait

It is very important to document any birthmarks or bruising/lacerations from birth trauma found on initial examination in case there are any child protection concerns in the future.

- Face

  Appearance – note any dysmorphic features

  Asymmetry – e.g. facial nerve palsy

- Nose

  Patency of nasal passages – infants are obligate nasal breathers, therefore, will present with respiratory distress and cyanosis at rest if they have bilateral choanal atresia.

- Eyes

  Inspect the eyes for evidence of erythema or discharge

  Inspect the sclera for any discolouration

  Position and shape (e.g. any ptosis, epicanthic folds, upslanting, coloboma, aniridia)

  Assess red reflex:

  Use ophthalmoscope to look for red reflex.

  An absent red reflex requires immediate ophthalmology referral as it may suggest congenital cataracts or retinoblastoma

- Ears

  Inspect the pinna – asymmetry / prominence / accessory auricles/ skin tags or pits.

- Mouth and palate

  Look for micronagnatia

  Clefts of the hard or soft palate

  The full palate should be examined by visual inspection. And palpation for submucosal clefts. By insertion examiner finger with sterile glove, you also can assess the sucking reflex of the baby.

- Neck and clavicles

  Look for short neck, neck webbing or neck swelling (Turner syndrome with short, webbed neck and cystic hygroma)

  Palpate for clavicular fracture – secondary to traumatic birth (e.g. shoulder dystocia)

- Upper limbs

  Inspect for symmetry – ensure equal in size and length

Inspect fingers – ensure correct number and morphology. Polydactyly: Preaxial (from little finger), preaxial (from thumb)

Inspect palms – should have two palmar creases on each hand, single palmar crease may suggest Down's syndrome

Looks for Erb's palsy, Klumpke paralysis

- Chest

Chest wall deformities (pectus excavatum & carinatum), chest wall expansion

Respiratory system & cardiovascular system

Observe for respiratory failure, auscultate for abnormal breath sound & murmur.

Pulse oximetry: To check for preductal and postductal oxygen saturations for detection of critical congenital heart disease in newborn infants. Both readings should both be ≥95% and within 3% of each other.

- Abdomen

Inspect for evidence of abdominal distension& inguinal hernias

Palpate the abdomen: liver may be palpable but should not be more than 2cm below costal margin. Kidney occasionally can be palpated on deep palpation. Bladder should not be palpable.

- Umbilicus

Inspect for any discharge or hernias/ astroschisis/ omphalocele/ discharge

- Genitalia

Note any ambiguity of genitalia – e.g. congenital adrenal hyperplasia (in girls, boys with CAH will have normal genitalia)

Males:

Check for position of meatus (exclude hypospadias or epispadias).

Size of penis – should be at least 2cm.

Hydroceles – by transilluminates test.

Palpate scrotum to ensure both testes are present – unilateral undescended testis is common and should be followed up over time; bilateral absence is considered a disorder of sexual development and should be investigated

Females:

Inspect labia – ensure they are not fused.

Inspect clitoris – ensure it is normal size.

Vaginal discharge – white discharge is normal due to maternal oestrogens

- Lower limbs

Inspect limb symmetry – should be equal in size and length

Assess tone in both lower limbs

Assess movement in both lower limbs

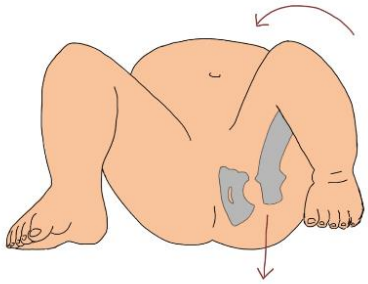
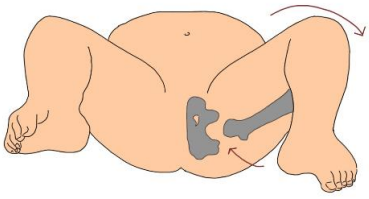
Palpate femoral pulses – Weak, absent or delayed femoral pulses are a sign of coarctation of the aorta.

Foot – e.g. talipes equinovarus, pedal oedema (which may indicate Turner's syndrome)

Ensure correct number of digits on each foot

- Hips

Barlow's and Ortolani's test are carried out as part of the routine newborn examination to detect development dysplasia of the hips. Hips should be examined individually with all clothing and nappy off.

Barlow's test 	Test is performed by adducting the hip whilst pushing the tigh posteriorly. If the hip is unstable, the femoral head will slip over the posterior rim of the acetabulum. If the hip is dislocatable the test is considered positive.
Ortholani's test 	Is used to confirm posterior dislocation of the hip joint. Flex the hips and knees of a supine infant to 90 degreesThen with your index fingers placing anterior pressure on the greater trochanters, gently and smoothly abduct the infant's legs using your thumbs. The test is positive if a clunk is heard when the femoral head relocates anteriorly into the acetabulum

- Back and spine

  Inspect the spine for:

  Scoliosis,hair tufts, naevus, birthmarks, sacral pits

  Hair tufts and sacral pits can be associated with underlying neural tube defects (spina bifida).

- Anus

  Inspect the anus for patency

  Meconium should be passed within 24 hours – delay is suggestive of obstruction or Hirschsprung's disease

- Reflexes

  Assess the newborn's reflexes:

  Palmar grasp reflex – When an object is placed in the infant's hand and strokes their palm, the fingers will close and they will grasp it with a palmar grasp. [8]

  Sucking reflex – Causes the child to instinctively suck anything that touches the roof of their mouth.

  Rooting reflex – Present at birth and disappears around four months of age, as it gradually comes under voluntary control. A newborn infant will turn its head toward anything that strokes its cheek or mouth to help breastfeeding.

  Stepping reflex – When the soles of their feet touch a flat surface they will appear to walk by placing one foot in front of the other.

  Moro reflex – Support the infant's upper back with one hand, then drop back once or twice into your other hand. The legs and head extend while the arms jerk up with the fingers extended. The arms are then brought together and the hands clench into fists, and the infant cries. Asymmetry may be due to hemiparesis, brachial plexus injury or fractured clavicle.

To complete the examination,

Share the results of the assessment with the parents, explaining the reason for any referrals you feel are required

Ask if the parents have any further questions

Thank the parents

Offer to dress the baby or allow parents to do so (depending on their preference)

Wash hands

Document your findings and suggest any relevant investigations or referrals

In Malaysia after delivery, the baby will be examined daily for first 4 days,

Then alternate day till Day 7 and then weekly till 28 weeks.

Case 29

A 50-year-old man presents to the Emergency Department with severe left sided chest pain. During initial assessment, the patient collapses. He is not responsive to call, not breathing and has no pulse. He has history of hypertension and dyslipidaemia for 5-years.

Questions

1. What possible emergency condition does the patient have now?
2. What will you do next?
3. Explain the mechanism of blood flow during cardiopulmonary resuscitation (CPR).
4. Is blood flow to the brain and heart adequate during CPR?
5. ECG monitoring shows either irregular wide-complex tachycardia or regular wide-complex tachycardia without pulse. What will be the diagnosis?
6. Explain the mechanism of defibrillation.
7. After the 2nd defibrillation, you get the normal heart rhythm on the monitor. What will you do?
8. You have resuscitated the patient at the Emergency Department. How will you proceed to the next step?
9. If ECG recording shows a straight line, what will be the diagnosis?
10. How do you manage the asystole?

Discussion

1. **What possible emergency condition does the patient patient have now?**

There will be only one condition called **"Cardiac arrest."**

2. **What will you do next?**

You need to start resuscitation measure immediately.

Start BLS (Basic Life Support) algorithm. Start CPR with chest compression and breathing (30:2 ratio).

Check cardiac rhythm at the cardiac monitor whether it is **shockable** or **non-shockable** rhythm.

3. **Explain the mechanism of blood flow during CPR.**

There are two models of mechanism of blood flow when we do chest compression.

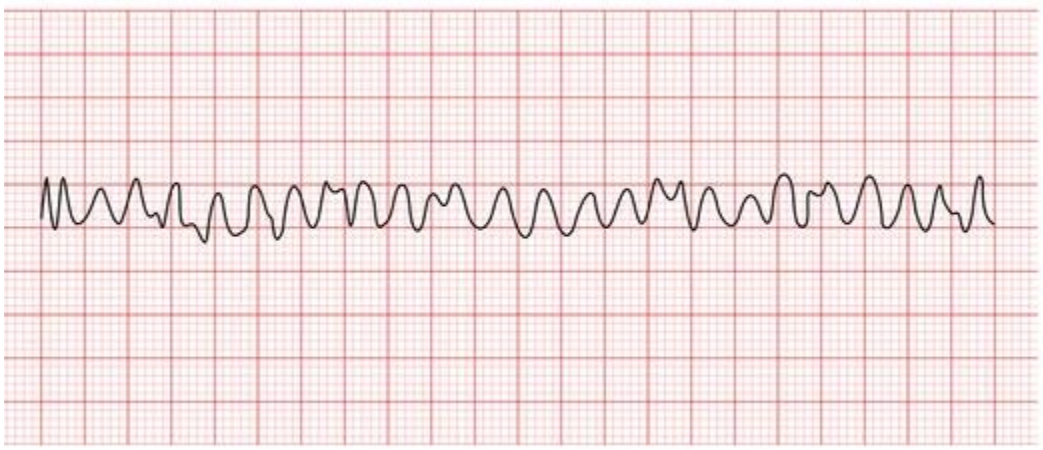
1. Cardiac pump model: The heart is pressed or squeezed between the sternum and the spine. During compression, the atrio-ventricular valves close and unidirectional blood flow occurs towards the great arteries. During relaxation intracardiac pressure falls, the atrio ventricular valves open, and blood is drawn to the heart from the lungs and veins.

2. Thoracic pump model: chest compression uniformly increases pressure throughout the thoracic cavity. Then, forward blood flow is achieved. During relaxation or chest recoil, there will be increased negativity in thoracic pressure which improves ventricular filling and coronary blood flow.

4. **Is blood flow to the brain and heart adequate during CPR**?

Even the best CPR provider can achieve only approximately 30% normal blood flow to the brain and heart.

5. **ECG monitoring shows either irregular wide-complex tachycardia or regular wide-complex tachycardia without pulse. What will be the diagnosis?**



The above ECG shows ventricular fibrillation

Either of these is a **shockable rhythm** (ventricular fibrillation); you need to defibrillate using manual defibrillator as soon as defibrillator is ready.

6. **Explain the mechanism of defibrillation**.

Defibrillation is the therapeutic use of electric current delivered in large amounts for a brief period to the heart. Defibrillation shock temporarily depolarizes ("stuns") an irregularly beating heart and allows natural pacemaker in the sinoatrial node of the heart to resume the normal heartbeat.

Currently, Manual defibrillators are biphasic, and you can use 120 to 200 Joules of shocking energy (depends on the manufacturer's recommendation). If it is a monophasic defibrillator, you can use 360 Joules. But if you are not sure of the type of defibrillator, you can use maximum dose.

Delay defibrillation will reduce the chance of conversion to normal rhythm. That is why AHA recommended immediate defibrillation as soon as defibrillator is ready. You must restart CPR immediately after the defibrillation shock. After 2 minutes, recheck the pulse again. If still in ventricular fibrillation, you need to shock again.

Then, you can give 1st dose of adrenaline 1mg IV just before or after the 2nd Shock

7. **After the 2nd defibrillation, you get the normal heart rhythm on the monitor. What will you do?**

Check the carotid pulse again. If you can feel the pulse, it means the heart is beating spontaneously.
It is called Return of spontaneous circulation (ROSC).

8. **You have resuscitated the patient at the Emergency Department. How will you proceed to the next step?**

If the patient has ROSC, you must maintain his cardiovascular function and oxygenation. It is called post cardiac arrest care.
You must make sure patient has adequate ventilation and good oxygen saturation. You can give oxygen via face mask or nasal prong or intubate and ventilate if no spontaneous breathing.
You need to support the cardiovascular function by giving fluid and vasopressor drugs if needed.
Then you need to check his 12 lead ECG. If ECG shows STEMI (ST elevation MI) or is highly suggestive of myocardial infarction, immediate referral to cardiologist for the reperfusion therapy is needed.
Finally, the patient should be managed in the advanced critical care unit. If patient is still unconscious, need to consider targeted temperature management (Therapeutic hypothermia).

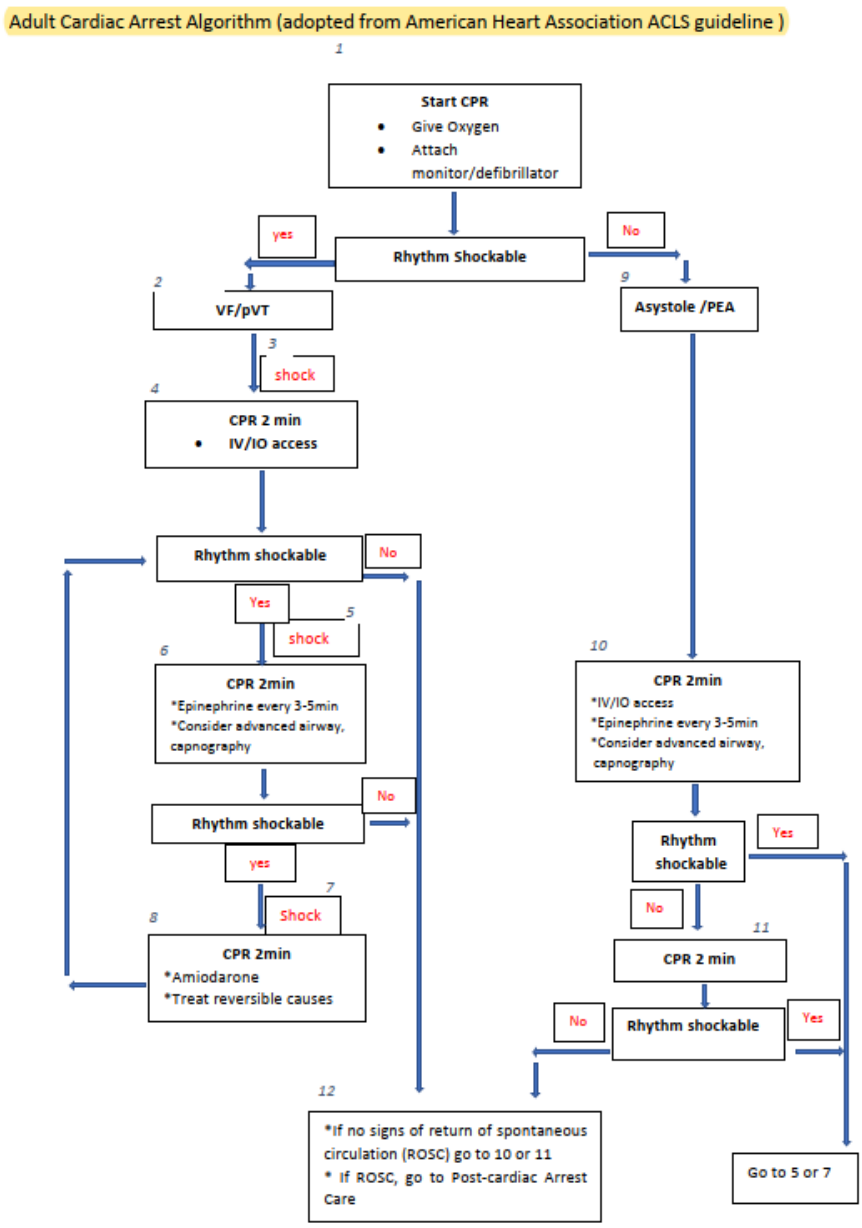
9. **If ECG recording shows a straight line, what will be the diagnosis?**

It is called asystole. Sometimes, you may see the cardiac rhythm on the monitor and no pulse will be felt. This condition is called pulseless electrical activity (PEA).
Both conditions are non-shockable rhythms.

10. **How do you manage the asystole?**

Follow Advanced Cardiac Life Support (ACLS) algorithm and do CPR.

Remarks: student needs to know more about the sequences of resuscitation and drugs used in ACLS.



The following are used in ACLS cardiac arrest algorithm.

CPR quality

- Push hard (at least 2 inches [5 cm]) and fast (100-120/min) and allow complete chest recoil.
- Minimize interruptions in compressions.
- Avoid excessive ventilation.
- Rotate compressor every 2 minutes, or sooner if fatigued.
- If no advanced airway, 30:2 compression-ventilation ratio.
- Quantitative wave form capnography
  - If PETCO2 <10 mmHg, attempt to improve CPR quality
- Intra-arterial pressure

- If relaxation phase (diastolic) pressure < 20 mmHg attempt to improve CPR quality.

Shock Energy for Defibrillation

- Biphasic: Manufacture recommendation (eg, Initial dose of 120-200 J); if unknown, use maximum available.
  - Second and subsequent doses should be equivalent, and higher doses may be considered.
- Monophasic: 360J

Drug Therapy

- Epinephrine (Adrenaline) IV/IO dose: 1mg every 3 to 5 minutes
- Amiodarone IV/IO dose: First dose: 300mg bolus. Second dose: 150 mg.

Advanced airways

- Endotracheal intubation or supraglottic advanced airway
- Waveform capnography or capnometry to confirm and monitor ET tube placement
- Once advanced airway is in place, give 1 breath every 6 seconds (10 breaths/minute) with continuous chest compressions

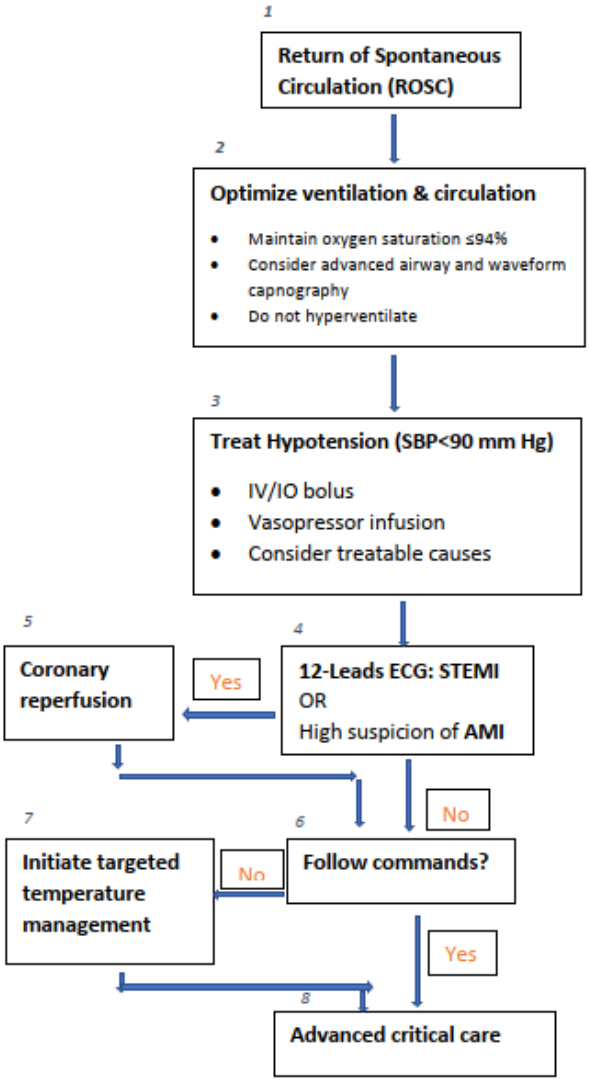
Return of Spontaneous Circulation (ROSC)

- Pulse and blood pressure
- Abrupt sustained increased in PETCO2 (typically ≥ 40 mmHg)
- Spontaneous arterial pressure waves with intra-arterial monitoring

Reversible Causes

- Hypovolemia
- Hypoxia
- Hydrogen ions (acidosis)
- Hypo-/hyperkalemia
- Hypothermia
- Tension pneumothorax
- Tamponade, cardiac
- Toxins
- Thrombosis, pulmonary
- Thrombosis, cardiac

Adult Immediate Post-Cardiac Arrest Care Algorithm *(adopted from AHA ACLS guideline 2015)*



Doses/Details

Ventilation/oxygenation:
Avoid excessive ventilation.
Start at 10 breaths/min and titrate to target $PETCO_2$ of 30-40 mm Hg.
When feasible, titrate FIO_2 to minimum necessary to achieve $SPO_2 \leq 94\%$.

IV bolus:
Approximately 1-2 L normal saline or lactated Ringer's

Epinephrine (Adrenaline) IV infusion
0.1 to 0.5 mcg/kg per minute
(In 70-kg adult: 3-35mcg per minute)
Dopamine IV infusion:
5-10mcg per minute
Norepinephrine IV infusion:
0.1 to 0.5 mcg/kg per minute
(In 70-kg adult: 3-35mcg per minute)

Reversible Causes
Hypovolemia
Hypoxia
Hydrogen ions (acidosis)
Hypo-/hyperkalemia
Hypothermia
Tension pneumothorax
Tamponade, cardiac
Toxins
Thrombosis, pulmonary
Thrombosis, cardiac

Case 30

A 54-year-old lady presents to the Emergency Department with palpitations. Her Blood pressure is 120/80 mmHg, pulse rate is 160 beats/minute, respiratory rate is 24 breaths/min, and oxygen saturation is 96% on room air.

Questions

1. How will you manage this patient initially?
2. What is the diagnosis on arrival?
3. How do you treat this patient?
4. What is the action of adenosine and why do you need to inject with IV push?
5. If the patient becomes restless, her blood pressure is dropping, and experiencing chest pain during the examination, what will be your clinical diagnosis?
6. How do you treat unstable tachycardias?
7. What is the synchronised cardioversion?

Discussion

1. **How will you manage this patient initially?**

First, you have to decide that these symptoms are caused by increased heart rate. Sometimes, slight increase in heart rate doesn't cause palpitations, shortness of breath or chest pain. Assess the appropriateness of increased heart rate and clinical presentation before starting the action

Then, Check for the vital signs. In this case, heart rate and respiration rate are increased but BP and oxygen saturation are within the normal range.

Then, look for the underlying causes and treat. For example, high fever, septicaemia, acute exacerbation of bronchial asthma and some medical conditions can be the cause of increased heart rate. In these cases, the underlying causes will be treated first before acting on the increased heart rate.

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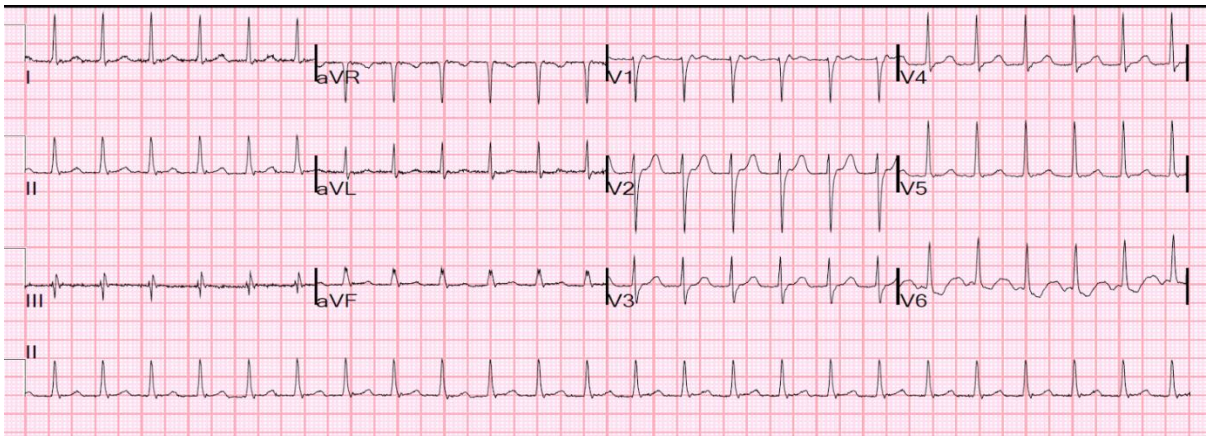
1. Maintain the patent airway and assist the breathing if necessary
2. Give Oxygen if hypoxaemic: means ≤ 94% oxygen saturation
3. Use cardiac monitor to identify rhythm, monitor blood pressure and oxymetry

Her ECG is given below:



In this ECG, heart rate is 150 beats/min. It shows narrow complex tachycardia without p-waves. Do not mistake T-waves in this ECG for P-waves.

2. **What is the diagnosis on arrival?**

Apart from tachycardia, her vital signs are in the normal range. So, the patient is considered stable and not in life threatening state.
Generally, ECG diagnosis is supraventricular tachycardia (SVT)
Clinical diagnosis will be stable tachycardia due to SVT

3. **How do you treat this patient?**

1. Set IV access and 12 lead ECG (which is already available)
2. Try vagal manoeuvres such as carotid sinus massage, ~~or~~ Valsalva manoeuvre or cold-water drinking
3. If vagal manoeuvres are not effective, give IV adenosine 6mg IV push. The dose can be increased to 12 mg if tachycardia is not terminated with 6mg.
4. If it is stable irregular narrow-complex tachycardia, it is called atrial fibrillation. It is better treated by cardiology expert as it does not require urgent management.
5. You can consider referral to cardiologist.

4. **What is the action of adenosine and why do you need to inject with IV push?**

Adenosine produces transient atrioventricular nodal block when injected as an intravenous bolus. This is of therapeutic value in the conversion to sinus rhythm in a majority of paroxysmal supraventricular tachycardias, which involve the atrioventricular node in a re-entrant circuit.
Adenosine is remarkable for its rapid metabolism and brevity of action, with a half-life of a few seconds. That is why you need to give by IV-push
It commonly produces subjective symptoms, particularly chest discomfort, dyspnoea, and flushing, which are of short duration only.

Sometimes, when the heart rate is so fast, you may not be able to differentiate regular or irregular rhythms. When IV adenosine blocks the AV node, then you can see the AF waves (in irregular rhythms) or SVT is reverted to regular sinus rhythm. So, adenosine has both therapeutic and diagnostic effects when using in very fast atrial tachycardias.

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5. **If the patient becomes restless, her blood pressure is dropping, and experiencing chest pain during the examination, what will be your clinical diagnosis?**

It is known as unstable tachycardia regardless of whether it is narrow QRS complex or wide complex. These symptoms are caused by low cardiac output.

Common symptoms of low cardiac output due to very fast heart rate are:

1. Hypotension
2. Acutely altered mental status
3. Signs of shock
4. Ischaemic chest discomfort
5. Acute heart failure

It is called unstable because of higher risk of getting cardiac arrest.

6. **How do you treat unstable tachycardias?**

Synchronized cardioversion is the main stay of treatment for unstable tachycardia. Before treating with synchronized cardioversion, you have to consider sedation for the patient. There is exception only for the regular narrow complex tachycardia. You can consider IV adenosine.

7. **What is the synchronised cardioversion?**

Defibrillation is non-synchronized random administration of shock during a cardiac cycle.

Cardioversion is a synchronized administration of shock during the R waves or QRS complex of a cardiac cycle.

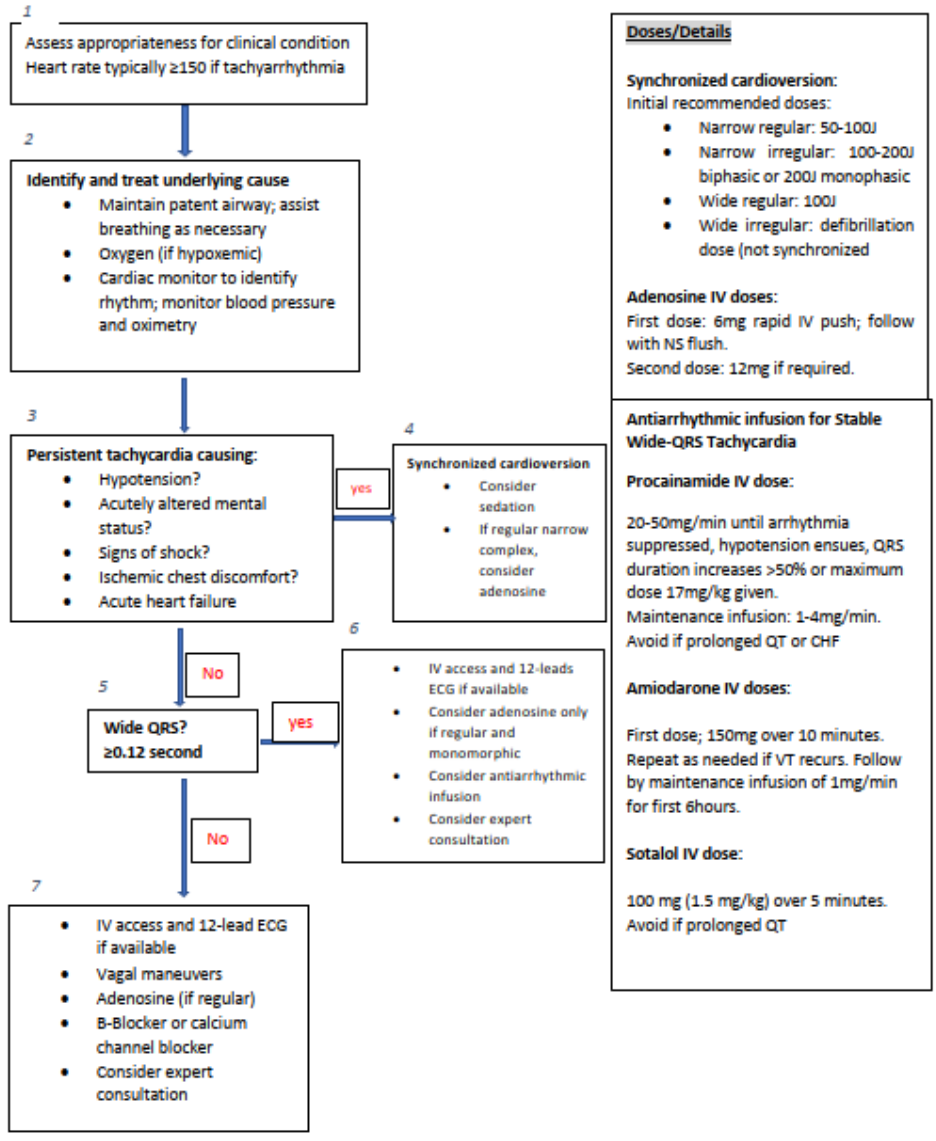
During defibrillation and cardioversion, electrical current travels from the negative to the positive electrode by traversing myocardium. It causes all the heart cells to contract simultaneously. This interrupts and terminates abnormal electrical rhythm. This, in turn, allows the sinus node to resume normal pacemaker activity.

If ECG shows wide QRS complex tachycardia and there are no signs and symptoms of low cardiac output, this condition is called stable wide-complex tachycardia.

Usually regular wide-complex tachycardia is ventricular tachycardia (VT) unless proven otherwise.

For stable VT, you can treat with antiarrhythmic drug infusion and consider for the expert consultation.

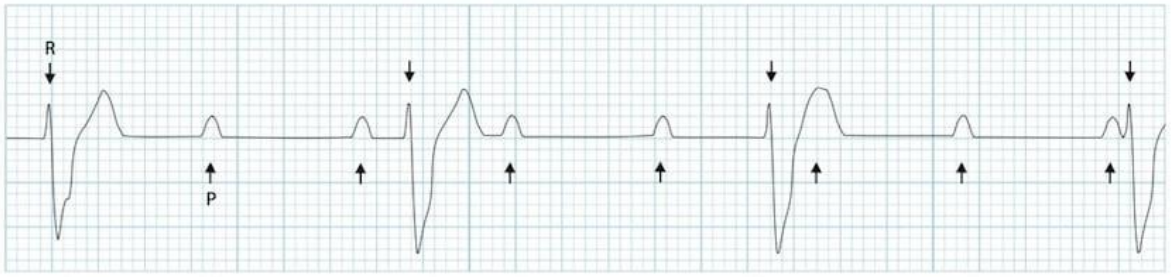
Adult Tachycardia with a Pulse Algorithm (Adopted from AHA ACLS guideline)



Case 31

A 62-year-old man presents to the Emergency Department with central chest pain for 2-hours' duration. He also has nausea and vomited once before arrival to the ED. He has a history of hypertension and ischaemic heart disease. On arrival to ED, His blood pressure is 90/60 mmHg, pulse rate is 40beats/min, and oxygen saturation is 94% on room air.

You take ECG and it shows as below:



Questions

1. Describe your initial management plan for this patient on arrival.
2. What will be the ECG diagnosis and clinical diagnosis for this patient?
3. The patient's condition needs urgent or immediate intervention. What will you do?
4. How do atropines increase the heart rate?
5. If a patient presents with bradycardia without any symptoms, what will you do in the Emergency Department?

Discussion

1. Describe your initial management plan for this patient on arrival

First, you have to decide that these symptoms are caused by a very slow heart rate. Then, check for the vital signs, look for the underlying causes, and treat.

Maintain the patent airway and assist the breathing if necessary

Give Oxygen as the patient's SpO_2 is 94%.

Use cardiac monitor to identify rhythm, monitor blood pressure and oxymetry

2. What will be the ECG diagnosis and clinical diagnosis for above patient?

ECG shows:

1. Regular p-waves about 100/minutes and regular QRS complexes of 38/min
2. There is a complete dissociation between p-waves and QRS complexes

The ECG diagnosis is complete heart block or 3rd degree AV block.

Because of his presenting symptoms and vital signs, the patient looks unstable.

Common symptoms of low cardiac output due to very slow heart rate are:

- Hypotension
- Acutely altered mental status
- Signs of shock
- Ischaemic chest discomfort
- Acute heart failure

It is known as "unstable" because of higher risk of progressing to cardiac arrest.

So, the Clinical diagnosis will be unstable bradycardia secondary to complete heart block.

3. The patient's condition needs urgent or immediate intervention. What will you do?

In this condition regardless of several types of bradycardia, you should initiate urgent treatment as soon as possible.

1. Start IV atropine; first dose is 0.5mg bolus and repeat every 3-5 minutes (maximum 3 mg)

2. If Atropine is ineffective, you need to start either one of the following procedures.
    A. Do Transcutaneous pacing (TCP)
       OR
    B. Use dopamine infusion (infusion rate is 2-20mcg/kg per minute. Titrate to patient response)
       OR
    C. Adrenaline infusion (infusion rate is 2-10 mcg per minute. Titrate to patient response)
3. Then, consider expert consultation and plan for transvenous pacing.

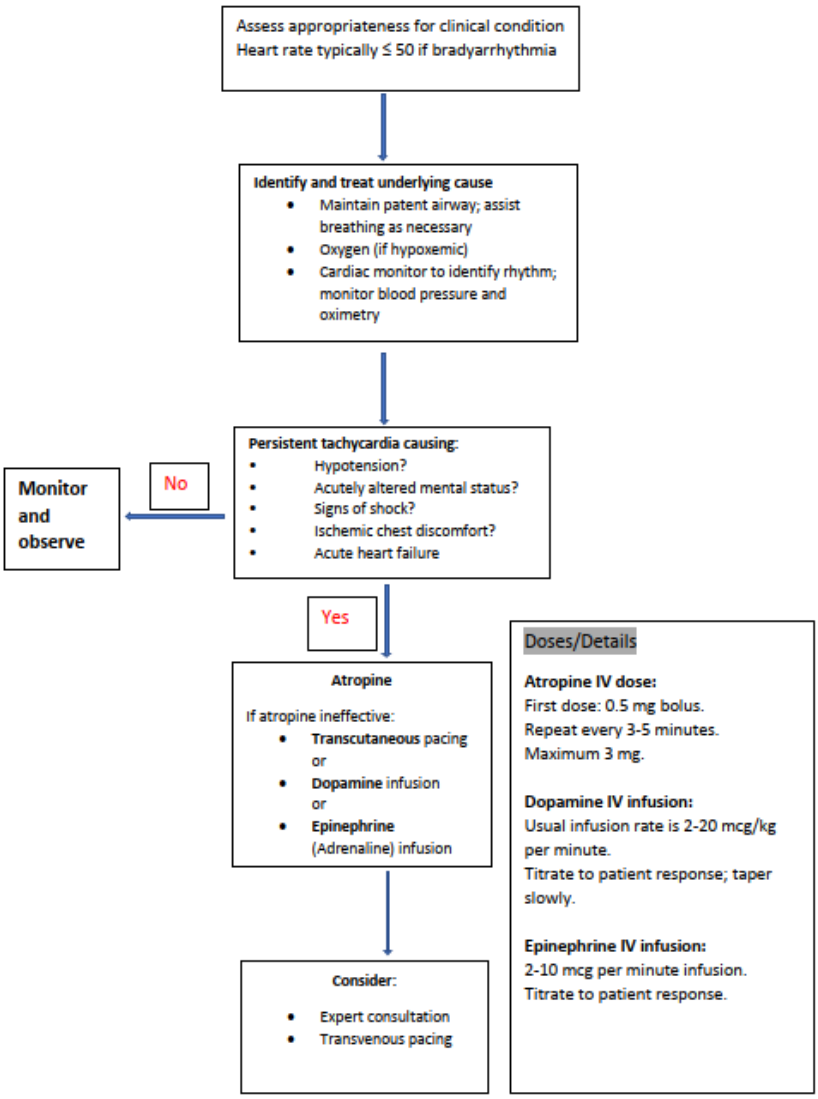
4. How do atropines increase the heart rate?

Atropine (anticholinergic drug) increases the heart rate and improves the atrioventricular conduction by blocking the parasympathetic influences on the heart.
By blocking the actions of acetylcholine, muscarinic receptor antagonists very effectively block the effects of vagal nerve activity on the heart. By doing so, they increase heart rate and conduction velocity.

5. If a patient presents with bradycardia without any symptoms, what will you do in the Emergency Department?

This would be asymptomatic bradycardia or stable bradycardia. You can observe the patient and refer to the cardiologist for further management.

Adult Bradycardia With a Pulse Algorithm (Adopted from AHA ACLS guidelines 2015)



Case 32

Rahman is a 33-year-old Bangladeshi who works at a motor shop in Sungai Besi as a mechanic. He was brought by his employer to the Emergency department of Hospital KL due to a fracture of his left foot sustained after an accident at the workplace. He was unable to ambulate, and his left foot becomes swollen and tender.

Further inquiry reveals the following:

Occupational History: Works as a mechanic at Sungai Besi from 2019 and holds a valid working visa.

Family History: He lives with his wife, who also works as a helper to the family of his employer. Both of his parents are alive & healthy in Bangladesh.

Social History: Never been smoking or consuming alcohol.

Pertinent Physical Findings:

General Survey- Alert, conscious, wheelchair-bound due to difficulty in moving his left foot, in severe pain due to swollen left foot.

Vital Signs

• Blood Pressure: 130/72

• Pulse Rate: 108 beats per minute

• Temperature: 37 degrees Celsius

• Respiratory Rate: 30 breaths per minute

• SpO2: 97.5%

Cardiovascular system:

Normal rate, regular rhythm, no murmur, no other abnormality detected

Respiratory system:

Normal breath sounds.

GIT & Nervous System:

No significant finding

Xray of the left foot was taken and revealed: Complete displaced fracture of the lateral malleolus.

The patient was admitted for 4 days, and the broken bone was placed in a cast. He was sent home with a walker.

The patient was advised to file for a medical leave for at least 6 weeks and to follow up in between for physiotherapy.

Rahman disclosed that he may not be able to comply with the doctor's advice due to financial difficulties. He cannot afford to incur absences at work due to NO WORK NO PAY POLICY of the shop. He receives a monthly salary of RM 1,200.

One way to help Rahman is thru the Social Security Organization (**SOCSO**). SOCSO is a Malaysian-based government department under the Ministry of Human Resources that is responsible to provide social security protections to all employees including medical and cash benefits, provision of artificial aids and rehabilitation to employees to reduce the sufferings and to provide financial guarantees and protection to the family.

Question

1. Discuss the eligibility of Rahman to the benefits of SOCSO.
2. Who is responsible for paying SOCSO contribution of Rahman? What is the rate of the contribution and how much are the monthly dues?
3. What are the different schemes under this insurance program? Discuss the scheme in which Rahman's case will be considered or classified.
4. What are Rahman's benefits under this scheme?
5. Is he eligible for the Temporary Disablement Benefit? If so, calculate how much will he receive based on the MC given by the attending physician.
6. Where will you refer this patient for his physiotherapy?

Discussion

1. **Discuss the eligibility of Rahman to the benefits of SOCSO.**

   Though SOCSO is primarily for Malaysians, it has also extended its coverage to all legal foreign workers in Malaysia, effective 1 January 2019, (including domestic servants- effective June 16, 2021) whereby they will be covered by Employment Injury (EI) Scheme under the Act 4. The EI Scheme protects employees against occupational accidents or diseases arising out of and in the course of their employment as well as commuting accidents

2. **Who is responsible for paying SOCSO contribution of Rahman? What is the rate of the contribution and how much are the monthly dues?**

   The rate of contribution for foreign workers is 1.25% of the insured monthly wages (capped at RM 4,000) and to be paid by the employer.

   Rahman salary RM 1,200 X 1.25%= 15 ringgit /month

3. **What are the different schemes under this insurance program? Discuss the scheme in which Rahman's case will be considered or classified.**

There are 3 different social security protection schemes: employment injury scheme, invalidity scheme and self-employed employment injury scheme. Rahman can be covered only under the employment injury scheme. This scheme protects employee against accident or occupational disease arising out of and in the course of employment

4. **What are Rahman's benefits under this scheme?**

   Rahman will enjoy the same benefits as Malaysians except for except the dialysis, vocational and return to work programs.

   Below are the seven benefits under this Employment Injury Scheme:

a. *Medical benefit*

   Employees suffering from employment injuries or occupational diseases may receive free medical treatment at SOCSO's panel clinic or Government clinic/hospital until

they are fully recovered. For serious injuries, medical care may be obtained from the Government hospital and the employee is eligible for second class ward treatment. Specialist treatment will be provided, if required. Employers or employees can claim reimbursement of expenses incurred in respect of medical treatment at SOCSO's non-panel clinic.

b. Temporary Disablement Benefit

Temporary Disablement Benefit is paid for the period the employee is on medical leave certified by a doctor for not less than 4 days including the day of the accident. However, Temporary Disablement Benefit will NOT be paid for the days for which the employee works and earns wages during this period.

c. Permanent Disablement Benefit

Employees who suffer from permanent disability due to employment injury can apply for this benefit. The daily rate of Permanent Disablement Benefit is 90% of the employee's average assumed daily wage, subject to a minimum of RM30.00 per day or a maximum of RM118.50 per day.

d. Constant-attendance Allowance

This allowance is paid to an employee who is suffering from total permanent disablement and is so severely incapacitated as to constantly require the personal attendance of another person, certified by the Medical Board or Special Medical Board or the Appellate Medical Board. The allowance is fixed at RM500 per month.

e. Dependents' Benefit

If an employee dies because of an employment injury, his dependents are entitled to this benefit. The daily rate is 90% of the average assumed daily wage subject to a minimum of RM30.00 per day and a maximum of RM118.50 per day.

f. Funeral Benefit

Funeral Benefit will be paid to the eligible person if the employee dies because of employment injury or while he/she receives Permanent Disablement Benefit.

g. Rehabilitation Facilities

Physiotherapy, Occupational therapy, Reconstructive surgery, supply of prosthetics, orthotics and other appliances, supply of orthopedics apparatus such as a wheelchair, crutches, hearing aid, spectacles, special shoes, and others.

Based on the injury suffered by Rahman, he will be eligible for medical benefit, temporary disablement benefit and physiotherapy.

5. **Is he eligible for the Temporary Disablement Benefit? If so, calculate how much will he receive based on the MC given by the attending physician.**

   Yes.

   The daily rate of Temporary Disablement Benefit is 80% of the employee's average assumed daily wage. The minimum rate is RM30.00 per day while the maximum rate is RM105.33 per day.

   Rahman receives 1,200 per month, approximately about RM 40/day

   Temporary disablement benefit 80% of RM 40= RM32/day X 30 days= RM960

   Rahman will receive RM 960 in a month.

   His Medical leave is six weeks so he will receive RM 1,440.

6. **Where will you refer this patient for his physiotherapy?**

Rahman can be referred to a SOCSO panel rehabilitation center near his residence in Sungai Besi.

Case 33

Steve, a 60-year-old male retiree consults his general practitioner about his chronic respiratory complaints which started nine months ago and have worsened since then. He complains of shortness of breath on exertion with nonproductive cough. He has not had fever nor chest pain. He is a non-smoker and has no other past medical or surgical history of note. His previous consultation was infrequent and was for upper respiratory tract infections and gastroenteritis. The patient's occupational history reveals that he has worked in different industries in the past 40 years. (cattle farming, coal mining and shipbuilding)

Physical Examination:

On general examination, digital clubbing is observed. Pulmonary examination reveals **bibasilar crackles with "Velcro" quality.**

Diagnostics: *Pulmonary function test results*

Item	Measured Value	Predicted Value	% of predicted
FVC (L)	2.80	3.69	70.7
FEV1 (L)	2.39	3.11	85.4
FEV1/FVC (%)	85.3	84	101.1

Questions

1. What is your impression of the case?
2. Which form will you fill up to notify this case to the DG of Department of Occupational Safety and Health (DOSH)?
   Fill out the Section D of the form based on the case scenario.
3. What is the penalty if you fail to notify this case to DOSH Malaysia?

Discussion

1. **What is your impression of this patient's illness?**
    a. Probably due to an occupational disease, as he has worked in multiple industries for forty years.
    b. Provisional Diagnosis is Asbestosis based on the following:

- **Occupational Exposure** (asbestos fibers from the shipbuilding industry) (Asbestos is a naturally occurring mineral which can resist corrosion and high temperatures, which makes it an ideal material for use in the shipbuilding industry. It is used to insulate boilers, and steam pipes.) Though it was not mentioned in the history when he started working in this industry, 40 years is more than enough to accommodate the considerable latency period (usually at least 20 years) between exposure and onset of clinically apparent disease.
- **Symptomatology:** Progressive dyspnoea with non-productive cough in the past nine months, non-smoker with no known past medical conditions.
- **Physical Findings:** Lung auscultatory findings of bibasilar crackles with "Velcro" quality suggest pulmonary fibrosis. Digital clubbing reflects chronic poor oxygenation which can be caused by ~~a~~ compromised lung function.
- **Diagnostic Tests:** *Pulmonary Function Test* Although the predicted percentage of FEV1/FVC ratio is almost normal (98.5%), Both his FVC (forced vital capacity) and FEV1 are lower than predicted values which is consistent with a restrictive pattern of a lung disease.

2. **Which form will you fill up to notify this case to the DG of Department of Occupational Safety and Health (DOSH)?**
   Fill out the Section D of the form based on the case scenario.

The relevant form is JKKP 7

JKKP 7

REPORT FOR OCCUPATIONAL POISONING / OCCUPATIONAL DISEASE OCCUPATIONAL SAFETY AND HEALTH
(NOTIFICATION OF ACCIDENT, DANGEROUS OCCURANCE, OCCUPATIONAL POISONING AND OCCUPATIONAL DISEASE) REGULATION 2004

Part D of the form is given below

Part D

Description of work that led to occupational poisoning/disease (Please describe any work done by the affected person which might have led to them getting the disease is thought to have been caused by exposure to an agent at work, e.g.a specific chemical - please state what that agent is)

Signature of Notifier

Date

3. What is the penalty if you fail to notify this case to DOSH Malaysia?

Either or both a fine up to 10 000 Ringgit Malaysia and imprisonment up to a year, upon conviction.

The relevant regulations are given below

Guidelines on Safety and Health (Notification of Accident,
Dangerous Occurrence, Occupational Poisoning
and Occupational Disease) Regulations 2004
[NADOPOD]

Section 32 of the Occupational Safety and Health Act states that an employer shall notify the nearest DOSH office of any accident, dangerous occurrence, occupational

poisoning or occupational disease which has occurred or is likely to occur at the place of work.

Registered Medical Practitioner. Every medical practitioner or medical officer attending to, or called in to visit, a patient whom he believes to be suffering from any of the occupational poisoning or occupational disease listed in Third Schedule or Table 16, must report the matter to the Director General within 7 days using the approved form (JKKP 7).

10.3. (a) Section 51 of the Act and regulation 13 of the Occupational Safety and Health (Notification of Accident, Dangerous Occurrence, Occupational Poisoning and Occupational Disease) Regulations 2004 prescribes penalties for failure to report and keep records. The penalties for conviction is a fine not exceeding ten thousand Ringgit Malaysia or to imprisonment for a term not exceeding one year or to both.

Case 34

A- 32-year-old woman presents with recurrent headaches for the past 1 year. The headaches have worsened in the past few months during the lockdown. Her husband, a tour guide lost his job during the Covid 19 pandemic. The headaches are associated with insomnia, low energy, and generalized body pains. She has been treated with several medications after numerous visits to several clinics, but the symptoms persist. It has not been associated with blurring of vision, fever, neurological deficit, or trauma. She has also had a CT scan of the brain last month and it was reported as normal.

Today, she is very anxious for some treatment as her husband is "fed up" with her headaches. She finds it difficult to cope with the housework and kids; and her husband is always angry with her. She is worried that her husband will become very agitated with her if she returns without any medications. Upon clarification of '*what happens when your husband becomes angry*?', she starts crying and reveals the story of her husband's anger; shouting and occasionally slapping her. After the consultation, she does not want to proceed with any intervention as she feels that she can still cope with her situation. She is given some advice and a follow up appointment.

1. What is intimate partner violence (IPV)?
2. What are the signs and symptoms of intimate partner violence?
3. How do perpetrators establish power and control over the victims?
4. What is cycle of abuse?
5. What are the essentials in the management of domestic violence cases?
6. In this scenario, the patient was not keen for intervention yet. Is the doctor's management appropriate?

Discussion

1. **What is intimate partner violence (IPV)?**

According to World Health Organization, intimate partner violence (IPV) or domestic abuse is defined as any behaviour within an intimate relationship that causes physical, emotional, or sexual harm to those in the relationship. The abuse includes physical and sexual violence, emotional abuse (ongoing humiliation or intimidation), economic restrictions (confiscating earnings), and other controlling behaviours (monitoring movements, restricting access to information or assistance). Most survivors of IPV report that the emotional abuse is often worse than the physical abuse. Multiple lifetime victimisation is common. A personal history of child abuse is a risk factor for being a victim or perpetrator, and perpetrators are likely to come from violent families. Studies worldwide show that one out of seven women has experienced domestic violence (DV) and that 20–40% of women will be victimized at least once during their lifetime. A World Health Organization (WHO) multi-country study has shown that the lifetime prevalence of DV ranges from 15 to 71%.

2. What are the signs and symptoms of intimate partner violence?

- Bruises and marks on the inside of the arms, back-injuries that point to a defensive position over the face
- Injuries to the chest and stomach, reproductive organs, and anus
- The illness or injuries do not match the cause given
- Delay in requesting treatment/care
- Injuries and bruises of various ages, indicating injuries occurring regularly over a period
- Repeat injuries, someone who is ‘accident prone’
- Injuries during pregnancy and repeated reproductive health problems: repeat miscarriage, early delivery, sexually transmitted diseases
- Psychological problems
- Suicide attempts or signs of depression
- Repeat and chronic medical complaints, pelvic problems and pains, psychological diseases

Partner's behavior

- Extreme and irrational jealousy / possessiveness
- Attempts to control time spent with the healthcare providers
- Speaking on behalf of the patient
- Insisting on staying close to the patient, who hesitates to speak in front of the partner

(*Reference: Addressing domestic violence in primary care: what the physician needs to know, Libyan J Med. 2014; 9: 10.3402/ljm. v9.23527)*

3. How do perpetrators establish power and control over the victims?

The power and control wheel, developed by the Domestic Abuse Intervention Programs in Duluth, Minnesota, shows the various tactics used by the perpetrators to establish power and control.

Figure 1: Power and Control Wheel developed by Domestic Abuse Intervention Programs in Duluth, Minnesota

(Reference: Domestic Abuse Intervention Programs, United States of America, 'Wheels' <https://www.theduluthmodel.org/wheels/>

4. What is cycle of abuse?

IPV often happens in cycles. The cycle of abuse happens when the perpetrator threatens violence, abuses the partner, apologizes, and promises to change, before starting the cycle again. The perpetrators may not be abusive all the time; the mix of both violent and "honeymoon" phases is what makes the victims confused and difficult to break away from the relationship. It is a tactic for the perpetrators to maintain power and control.

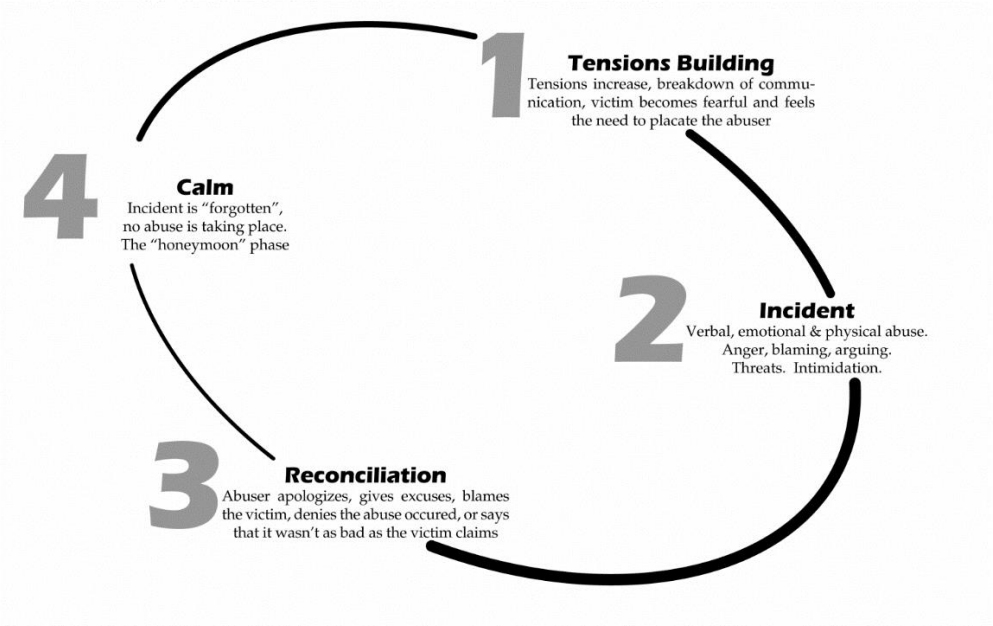


Figure 2: Cycle of Abuse

Image Retrieved from https://commons.wikimedia.org/wiki/File:Cycle_of_Abuse.png

5. What are the essentials in the management of domestic violence cases?

Assess: The degree of danger, presence of danger indicators, the mental status of the survivor

Safety: Assessment of safety of the victim at home needs to be established. Discuss a safety plan and revisit it with the survivor at each encounter

Support: Talk in private; make eye contact; assure confidentiality, use encouraging statements and show empathy

Options: Discuss options and provide information about legal options and community resources (e.g., women's shelters, support groups, legal advocacy, NGOs)

Strengths: Recognize the survivor's strengths.

Documentation: Record the patient's words, describe the observed behavior and injuries when present (pictorial documentations and photographs after obtaining patient's approval). Include also in documentation the assessment of the mental status, danger severity and follow up plans
Continuity: Show willingness to continue care of the survivor and always offer a follow-up appointment. Check for barriers to access and discuss solutions
(Reference: Addressing domestic violence in primary care: what the physician needs to know, Libyan J Med. 2014; 9: 10.3402/ljm. v9.23527)

6. In this scenario, the patient was not keen for intervention yet. Is the doctor's management appropriate?

Change is a long process and is always associated with frequent setbacks. Survivors should not be forced into decision making but rather supported at their own pace of readiness. By insisting on making changes, the doctor recreates power and control dynamics (as seen in the domestic abuse).

Therefore, assessing the survivor's readiness to change is necessary and as physicians, we can help the survivor move from one stage to the other towards action.

The stages of change are:

1. Pre-contemplation – The survivor is not aware of the situation or is still justifying abuse.
2. Contemplation – The survivor is considering change but is not ready to act yet.
3. Determination – A decision has been made by the survivor to make changes.
4. Action – The survivor is actively taking steps to address the DV.

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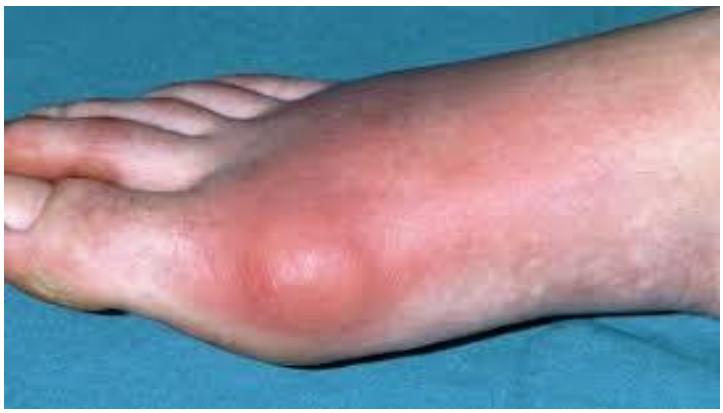
If any patients with domestic violence or IPV need help or support, Women's Aid Organisation (WAO) can be reached out at WAO Hotline at 03 3000 8858 or SMS/WhatsApp TINA at +6018 988 8058.

Punithavathy Shanmuganathan
Loh Keng Yin

Case 35

A 54-year-old -man presents to the clinic with sudden onset severe pain, tenderness and swelling of the first metatarsophalangeal joint of the right foot for the past one day. The pain is worsening, and he has difficulty walking. He has had 2 similar episodes within the last year, that resolved with medication. There is no history of trauma. He has underlying hypertension and diabetes mellitus for the past 2 years. His current medications are as follows:
Perindopril 4 mg OD
Metformin 1g BD
Simvastatin 20 mg ON
Blood tests reveal a raised serum urate level. On examination, his right foot is as shown below:



1. What is the most likely diagnosis for the above findings? State the criteria to fulfill the diagnosis as stated.

2. What is the important test that can be performed to confirm the diagnosis? State the expected findings of the test.

3. What are other investigations that must be done as a part of work up of a patient presenting with the abovementioned condition?

4. What is the non-pharmacological advice that you will give this patient?

5. What are the medications that can be used to treat the above acute condition?

6. What are the indications to initiate a prophylaxis treatment for this condition?

7. What is the medication that is frequently used as prophylaxis?

8. What are the side effects of the medication frequently used for prophylaxis?

Discussion

1. **What is the most likely diagnosis for the above findings? State the criteria to fulfill the diagnosis as stated**.

The most likely diagnosis is acute gouty arthritis.
Gout is characterized by painful joint inflammation, most commonly involving the first metatarsophalangeal joint, resulting from accumulation of monosodium urate crystals. The disease was described by Hippocrates in the fifth century BCE. It is associated with many serious comorbidities such as hypertension, chronic kidney disease, obesity, diabetes mellitus, and cardiovascular disease.

Gout presents for the first time typically with acute monoarthritis of the foot or ankle. It is self-limiting and lasts about 1-2 weeks. This acute episode is followed by complete resolution of symptoms and signs of joint inflammation. If hyperuricemia persists, the result is recurrent flares. Their occurrences gradually become more frequent and prolonged and may involve multiple joints (polyarticular gout), including those of the upper limbs.

Presence of two of the following criteria is required to make a diagnosis of gout:
- A clear history of at least 2 attacks of painful joint swelling with complete resolution within 2 weeks
- A clear history of podagra
- Presence of tophus
- Rapid response to colchicine within 48 hours of starting treatment.

2. **What is the important test that can be performed to confirm the diagnosis? State the expected findings of the test?**

Demonstration of monosodium urate crystals (negative birefringent) in synovial fluid confirms the diagnosis of gout and is the gold standard in the diagnosis of gout. It has 100% specificity. Polarised light microscopy is the standard method for detecting these crystals in the synovial fluid.

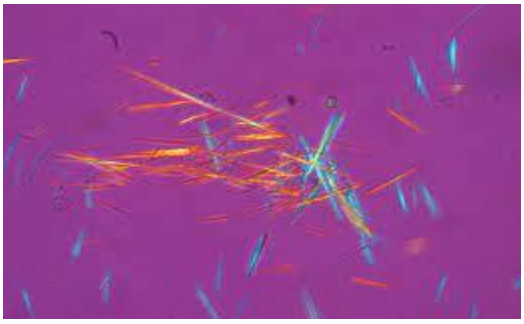


Figure 1. Negatively birefringent crystals under light microscopy

3. **What are other investigations that must be done as a part of work up of a patient presenting with the abovementioned condition?**
(As per recommendations in Clinical Practice Guidelines (CPG) Management of Gout, 2008, Malaysia)

Full blood count	To exclude infection or myeloproliferative disorders
Serum creatinine /urea	To exclude renal diseases or renal disease secondary to urate nephropathy
Serum urate	The level of urate cannot be used to confirm/exclude gout. A normal serum urate level does not exclude gout.
Blood glucose	To detect the presence of diabetes mellitus
Fasting lipid profile	To detect hypertriglyceridemia
Urinalysis	To detect renal disorders
Skeletal X ray	In acute gouty arthritis - usually normal In chronic tophaceous gout-erosive bone lesions characterized by punched out lesions
Renal imaging	Plain abdominal radiography detects only 10% of all urate stones Ultrasound is the investigation of choice as it would detect both radio-opaque and radiolucent stones as well as obstructive nephropathy

4. **What is the non-pharmacological advice that you will give this patient?**

- Health education and behavioral intervention should be offered
- Weight reduction in those who are obese/overweight. Aim to achieve an ideal body mass index (BMI)
- Restriction or elimination of alcohol intake (all types of alcohol - beer, wine, and liquor)
- Consumption of low-fat dairy products
- Adequate intake of fluid of 2-3 L per day
- Limiting intake of
    - purine-rich food especially of animal origin, except omega-3 polyunsaturated fatty acid-rich fish
    - high-fructose corn syrup
- avoidance of medications that increase risk of gout if feasible

5. **What are the medications that can be used to treat the above acute condition**?

In gout flare, the following may be used as monotherapy:

- Low dose colchicine
- Nonsteroidal anti-inflammatory drugs
- Corticosteroids

A combination of the above may be used if monotherapy response is inadequate.

6. **What are the indications to initiate a prophylaxis treatment for this condition?**

The following are indications for initiating hypouricaemic drug in a patient:

- Frequent and disabling attacks of gouty arthritis (3 or more attacks per year)
- Clinical/ radiographic signs of erosive gouty arthritis
- The presence of tophaceous deposits
- Urate nephropathy
- Urate nephrolithiasis
- Impending cytotoxic chemotherapy or radiotherapy for lymphoma or leukaemia

7. **What is the medication that is frequently used as prophylaxis?**

Allopurinol, a xanthine oxidase inhibitor, is a first-line agent to prevent recurrent gout. The starting dosage is 100 mg per day, and 300 mg per day is a common maintenance dosage. In patients with chronic kidney disease, low initial doses are recommended with slow titration to achieve target levels.

Febuxostat is a xanthine oxidase inhibitor that was approved by the FDA in 2009. Febuxostat is considerably more expensive than allopurinol. This drug may be used for patients with gout who have a contraindication to allopurinol or intolerance to allopurinol.

8. **What are the side effects of the medication frequently used for prophylaxis?**

Severe life-threatening adverse effects can occur with allopurinol.
Adverse effects include:

- Rash
- Bone marrow suppression
- Aplastic anemia
- Agranulocytosis
- Granulomatous hepatitis
- Jaundice
- Life threatening hypersensitivity syndrome which includes fever, rashes, hepatitis, eosinophilia, and renal impairment.

Certain ethnic groups have a higher risk of a severe hypersensitivity skin reactions when starting allopurinol therapy. Increased incidence has been seen in patients of Han Chinese, Thai descent and in Koreans with chronic kidney disease stage 3 or greater.