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HAEMATOLOGY LECTURE NOTES 2014

TOPICS:

1. Anaemia
2. Microcytic Anaemia
3. Iron Deficiency Anaemia
4. Thalassemia
5. Normocytic Anaemia
6. Congenital Spherocytosis
7. Glucose-6-phosphate Dehydrogenase Deficiency
8. Sickle Cell Disease
9. Autoimmune Haemolytic Anaemia
10. Macrocytic Anaemia
11. Pernicious Anaemia
12. Bleeding Disorders
13. Abnormal Coagulation
14. Haemophilia A
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16. Anti-coagulant Therapy
17. Bleeding in liver disease
18. Massive transfusion/cardiopulmonary bypass
19. Abnormal Platelets
20. Thrombocytopenia (ITP)
21. Vascular Defect
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23. **Pancytopenia**
24. **Leukaemias**
25. **Acute Lymphoid Leukaemia**
26. **Acute Myeloid Leukaemia**
27. **Chronic Myeloid Leukaemia**
28. **Chronic Lymphoid Leukaemia**
29. **Hodgkin's Lymphoma**
30. **Non-Hodgkin's Lymphoma**
31. **Myeloma**
32. **Myeloproliferative Disorders**
33. **Polycythemia Rubra Vera**
34. **Essential Thrombocytopenia**

ANAEMIA

This is low haemoglobin levels.

Parameters for anaemia:

Hb <13.5 g/dl in men
Hb <11.5 g/dl in women

Classification:

1. Microcytic anaemia - MCV <76
2. Normocytic anaemia - MCV 76-96
3. Macrocytic anaemia - MCV >96

Classification depends on the value of **MCV**. Each type has different causes.

Signs & Symptoms:

1. Light headedness
2. Tinnitus (ringing in the ear)
3. Pallor
4. Fatigue
5. Weakness
6. Dyspnoea
7. Palpitation
8. Headache
9. Angina

Complications of anaemia:

1. Tachycardia
2. Murmurs (harsh systolic murmur)
3. Heart failure
4. Hyperdynamic circulation
5. Cardiomegaly

CLASSIFICATIONS OF ANAEMIA AND COMMON CAUSES

1. LOW MCV <76 (microcytic hypochromic)

- A. Iron deficiency
- B. Thalassemia
- C. Congenital sideroblastosis

1. NORMAL MCV 76-96 (normocytic normochromic)

- A. Anaemia of chronic disease (e.g. RA, SLE)
- B. Bone marrow failure e.g. aplastic anaemia
- C. Renal failure

- D. Pregnancy
- E. Haemolysis
- F. Acute blood loss

1. **HIGH MCV >96 (macrocytic)**

- A. B12 & folate deficiency
- B. Alcohol
- C. Liver disease
- D. Hypothyroidism
- E. Anti-folate therapy (e.g. Phenytoin or Trimethoprim)
- F. Leukaemia

A. MICROCYTIC ANAEMIAS

1. IRON DEFICIENCY ANAEMIA

Causes:

1. Chronic blood loss e.g. in bleeding: GIT bleed from use of aspirin/NSAIDs or peptic ulcer or menorrhagia, GIT malignancy
2. Hookworm infection - caused by *Ancylostoma duodenale* and *Necator americanus*. They attach to the walls of the small intestines causing blood loss which is occult and presents as anaemia. Investigations: FBC for eosinophilia and stool analysis for ova. **Treatment: Albendazole**
3. Diet e.g. vegetarian/vegan
4. Malabsorption e.g. Coeliac disease which causes both iron and folate deficiency
5. Inflammatory bowel disease (Crohn's disease)

Signs:

1. Koilonychia
2. Atrophic glossitis (sore tongue)

Investigation results:

1. Decreased MCV, hypochromia, anisocytosis, poikilocytosis
2. Decreased serum iron
3. Decreased serum ferritin (transport affected)
4. Decreased TIBC (total iron binding capacity) / (no iron)

Treatment:

- **Oral iron → ferrous sulphate for at least 3 months**
- **Any case with Hb < 8 → blood transfusion**
- **Any case with Ischemic Heart Disease and Hb < 5 → blood transfusion**

2. THALASSEMIA

This is common in Asian continent like Sri Lanka and Sub Mediterranean continent like Italy, Malta

There is underproduction or no production of Hb β peptide chain

β Thalassemia major = Hb is usually <9:

There is mutation in β - globin genes → decreased or absent β chain product

Signs & Symptoms:

1. Severe anaemia
2. Failure to thrive
3. Usually splenomegaly (because haemolysis usually occurs in the spleen)

Investigations:

- Hypochromic, microcytic anaemia (MCV <76)
- Target cells
- HbF raised
- HbA2 variable
- HbA absent

Treatment:

- **Transfusion depending on haemoglobin levels (if Hb <8)**
 - **Desferrioxime to prevent iron overload**
 - **Splenectomy**
 - **Folate supplements**
 - **Bone marrow transplants**
- If no transfusion → death may occur.
 - With transfusion → normal development but increased risk of iron overload.
 - If blood transfusion is not adequate in children → anaemia → decreased growth → skeletal deformity (bossing of the skull)
 - After 10 years of repeated transfusions it can lead to endocrine failure, liver failure, heart toxicity (siderosis) due to accumulation of iron.

β-Thalassemia minor: Hb usually >9

- MCV < 75 microcytosis
- Hb > 9 mild – moderate anaemia
- Hb A2 > 3.5 %
- Hb F slightly increased

Treatment: No intervention just observe if asymptomatic

B. NORMOCYTIC ANAEMIA

This is anaemia with normal MCV 76-96

CAUSES:

1. **Anaemia of chronic disease** e.g. polymyalgia rheumatica, rheumatoid arthritis, SLE etc.
2. **Bone marrow failure** e.g. usually autoimmune (aplastic anaemia)
3. **Renal failure** is due to reduced production of erythropoietin. In this case you treat anaemia with erythropoietin IM.
4. **Pregnancy** due to increased demand of iron and folate. Anaemia in pregnancy treat with iron sulphate.
5. **Haemolysis** usually causes normocytic anaemia but chronic repeated haemolysis can cause microcytic anaemia.

HAEMOLYTIC ANAEMIA**CAUSES:**

1. Genetic/Congenital causes (usually young patients or a child)
 - a. RBC Membranopathies e.g. Spherocytosis, elliptocytosis
 - b. Haemoglobinopathies e.g. Sickle cell anaemia, thalassemia
 - c. Enzyme defects eg. G6PD Deficiency
1. Acquired Causes
 - a. Immune e.g. Haemolytic disease of the newborn, blood transfusion reactions, autoimmune haemolytic anaemia, drug induced (penicillin, L-dopa)
 - b. Non-immune eg. Trauma (microangiopathic haemolytic anaemia), infection (septicaemia, malaria), paroxysmal nocturnal haemoglobinuria

CLASSIFICATION

1. Intravascular haemolysis (takes place in the vessels): free plasma haemoglobin, haemoglobinuria, ↓ haptoglobins, haemosiderinuria.

2. Extravascular haemolysis (takes place in the spleen): There is splenomegaly because the red blood cells are destroyed in the spleen.

Signs & Symptoms:

1. Jaundice

2. Haematuria
3. History of drug intake
4. Previous anaemia
5. Family history
6. Hepatosplenomagaly
7. Leg ulcers (sickle cell anaemia)

Investigations:

- Increased bilirubin (unconjugated)
- Reticulocytosis (<1% is normal) N.B reticulocytes are young RBC.
- Increased urinary urobilinogen → polychromasia
- Increased haptoglobins (binds free Hb)
- **Direct Coomb's test** → positive in immune type haemolysis

1. [CONGENITAL SPHEROCYTOSIS](#): This is an autosomal dominant condition.

Strong family history, inheritance pattern is 1:2.

There is splenomegaly due to extravascular haemolysis. In this condition the red blood cells fail to change their shape as they pass through the spleen. As a result they get haemolysed by the spleen. There is ↑ risk of gallstones.

Investigations:

1. Osmotic fragility test
2. Spherocytes in blood film

Treatment: folate, splenectomy

NB: Spherocytosis can either be congenital or autoimmune. To differentiate you need to do direct coombs test. If it is positive then it's an autoimmune.

[2. G6PD: X- linked](#) Seen commonly in males

Precipitated by: Primaquine, sulfonamides, ciprofloxacin, **broad beans**

Signs & Symptoms:

1. Rapid anaemia
2. Jaundice
3. Heinz bodies (characteristic in microscopy, common in men)
4. Usually episodic depending on the precipitating factor.

Investigation: Enzyme assay after several weeks after a crisis

Treatment: avoidance of precipitating factors, transfusion if severe anemia

[3. SICKLE CELL DISEASE](#)

This is a congenital haemoglobinopathy which is common in Afro-Caribbeans.

Haemoglobin S when deoxygenated it causes sickling so the red blood cells are more fragile which leads to haemolysis in the spleen.

Signs & Symptoms:

1. Painful crisis e.g. chest pain, bone pain (bone infarction), etc.
2. Infection e.g. chest infection, urinary tract infection
3. Anaemia which may require recurrent blood transfusion
4. Jaundice which indicates haemolysis
5. Crisis can be precipitated by menstrual period

SICKLE CELL CRISIS:

1. **Thrombotic crisis:** precipitated by cold, infection, ischemia, severe pain.
2. **Aplastic crisis:** due to parvovirus
3. **Sequestration crisis:** hepatosplenomegaly → RUQ pain, increased LFT, decreased Hb

Management of crisis:

1. **Prompt analgesia (IV opiates) e.g. morphine**
2. **Give oxygen**
3. **Cross match blood, Full Blood Count, reticulocytes**
4. **Check for signs of infection: Blood culture, Chest X-Ray, Mid-Stream Urine**
5. **Rehydrate by giving normal saline and keep warm**
6. **Antibiotics if febrile**
7. **Blood transfusion as required**

Investigations:

- Hb 6-8g/dl
- Reticulocytes 10 – 20 % (normal reticulocytes is <1%, high reticulocyte count generally means haemolysis of any type)
- Increased Bilirubin
- Electrophoresis to check the sickle cells

Management of chronic disease:

- **Chemotherapy (hydroxyurea) if frequent crisis (increase the level of fetal haemoglobin)**
- **Chronic blood transfusion keep HbS < 30 %**
- **Bone marrow transplant**
- **Splenic infarct → hyposplenism → patient will need antibiotics + immunization for streptococcus and pneumococcal pneumonia, N. meningitidis, H. Influenzae**

4. AUTOIMMUNE HAEMOLYTIC ANAEMIA (AHA)

1. Primary (idiopathic meaning cause not known)
2. Secondary (secondary to lymphoma or SLE or CLL)

Warm AHA: presents as acute or chronic anaemia. Haemolysis occurs at 37 degrees and above.

Treatment:

1. **Steroids**
2. **Splenectomy**

Cold AHA: chronic anaemia gets worse with cold often associated with Raynauds phenomenon

Treatment: keep warm, blood transfusion, chemotherapy, chlorambucil

C. MACROCYTIC ANAEMIA

This is anaemia with high MCV > 96 fl

Causes:

1. Decreased B12
2. Decreased Folate
3. Alcohol
4. Liver disease
5. Pregnancy
6. Haemolysis
7. Hypothyroidism
8. Anti-folate drugs (e.g. phenytoin, methotrexate)

Investigations:

- Hypersegmented polymorphs/neutrophils usually found in B12 deficiency.
- Target cells (liver disease)
- LFT (increased GGT in alcoholism)
- Serum B12
- Serum folate
- Bone marrow biopsy → if the cause can't be found by any of the above tests
- If the B12 is ↓ then schilling test → malabsorption from terminal ileum
→ intrinsic factor deficiency (pernicious anaemia)
- Oral radioactive B12 → check for the amount present in urine

CAUSES OF LOW B12:

1. Pernicious anaemia (an autoimmune disease)
2. Post-gastrectomy: deficiency in parietal cells lead to deficiency in intrinsic factor which means that B12 will not be absorbed in the GI tract.
3. Diet: Common in Vegans and Vegetarians

CAUSES OF LOW FOLATE:

1. Poor diet common in alcoholism
2. Increased need pregnancy
3. Haemolysis
4. Malignancies
5. Long term haemodialysis
6. Malabsorption like in celiac disease
7. Anti-folate medication like phenytoin, trimethoprim

NB: alcohol causes high MCV but there is usually no anaemia. Then you can check GGT. It is commonly raised in alcoholism.

1. **PERNICIOUS ANAEMIA:** This is an autoimmune disease with antibodies to parietal cells and to intrinsic factor. Parietal cells are in the stomach and they produce intrinsic factor.

Common features:

1. Tiredness
2. Weakness
3. Shortness of breath
4. Paraesthesia/weakness in the limbs due to spinal cord degeneration
5. Atrophic glossitis (sore red tongue)
6. Diarrhoea

Associations:

1. Thyroid disorders
2. Vitiligo
3. Addison's disease
4. Carcinoma stomach
5. Associated with atrophic gastritis which is usually shown by absence of mucosal folding and this usually leads to stomach cancer

Investigation results:

1. Decreased Hb, MCV > 110, decreased B12, decreased WCC, decreased platelets
2. Hypersegmented polymorphs/ neutrophils
3. Megaloblasts in bone marrow
4. Antibody to parietal cells in 90 % (positive results are **not diagnostic**)
5. Antibody to intrinsic factor in 60% (are **diagnostic** if present)

Treatment:

- **Hydroxycobalamine (B12) IM for 2 weeks every other day**
- **Maintenance → IM every 2 months for life**

Intrinsic factor binds to B12 in the stomach which protects B12 from being ingested by the hookworms in the intestine.

COMPLICATIONS:

Sub-acute combined degeneration of spinal cord:

Posterior lateral columns are affected

Triad: extensor plantars + brisk knee jerk + hyper-reflex at ankle

Also there is parasthesia and weakness or ataxia of the limbs (lower limbs)

1. **BLEEDING DISORDERS**

General approach

After injuries these occur in order to stop bleeding:

1. Vasoconstriction
2. Platelet aggregation
3. Coagulation cascade (fibrin)

Vascular & Platelet disorders ⇒ prolonged bleeding + purpura + bleeding from mucous membrane

Coagulation disorder ⇒ Usually there is bleeding into joints, muscles GI & GU

Therefore normal haemostasis requires the interaction of

- a. **Platelets**
- b. **Fibrin from clotting cascade**
- c. **Normal microvasculature**

Bleeding could be due to:

- a. **Platelets = too few or dysfunction**
- b. **Coagulation abnormality**
- c. **Microvasculature abnormalities**

A. **ABNORMAL COAGULATION**

COAGULATION TESTS

1. PT → test for extrinsic system 10, 7, 2, 1 (10 – 14 sec)
2. INR → 0.9 – 1.2 (PT control), increased INR in warfarin, vitamin K deficiency & liver disease
3. APTT → intrinsic system 12, 11, 9, 8 (35 – 45 sec), increased PTT (heparin, haemophilia (factor 8 affected))
4. Thrombin time → 10 – 15 sec, increased in heparin, increased in DIC
5. Bleeding time (normal 7 min) → commonest cause is Von Willebrand's disease.

NORMAL VALUES

Prothrombin Time (PT): 10-14 seconds

APTT: 35-45 seconds

Bleeding Time (BT): < 7 min

Thrombin Time (TT): 10-15 seconds

In the GMC exam make sure you use their values as standard normal as sometimes there are variations e.g. an APTT of 42 may be regarded as high and if you do not use their values you may get a question wrong.

Abnormality	Type of defect	Causes
High PT	Extrinsic pathway defect	warfarin, liver disease, vitamin K deficiency
High APTT	Intrinsic pathway defect	Heparin, haemophilia, Von Willenbrand's disease, lupus anti-coagulant (anti-phospholipid syndrome)
High PT & APTT	Multiple defect	Liver disease, DIC, warfarin
High TT	Abnormal fibrinogen production	Fibrinogen defect, excess fibrinogen degradation products
Low fibrinogen	Excess consumption of clotting factors and fibrinogen	Consumption coagulopathy e.g DIC or Liver disease
High Bleeding time	Abnormal platelet function	Von Willenbrand disease (Also causes high APTT), or acquired platelets dysfunction, Perform platelets studies if you suspect this.
High TT, APTT & PT	Multiple (aquired) defects	Deficiency or abnormal fibrinogen or heparin

Common causes of abnormal coagulation

1. Haemophilia A & B
2. Anti-coagulants e.g. warfarin
3. Liver (usually oesophageal varices)
4. Massive blood transfusion (due to dilution thrombocytopenia)
5. DIC (precipitated by sepsis, or severe bleeding e.g. placental abruption)
6. Von Willebrand's disease
7. Vitamin K deficiency (Obstructive jaundice, small bowel disease due to malabsorption)

General Management:

1. **Fresh Frozen Plasma** - indicated for treatment of
 - 1) Acute DIC with bleeding
 - 2) To improve haemostasis in decompensated liver disease
 - 3) Emergency reversal of warfarin therapy if no prothrombin complex concentrate (PCC) available
1. **Vitamin K**: phytomenadione used for reversal warfarin overdose
2. **Protamine sulphate**: used to reverse heparin
3. **Cryoprecipitate /fibrinogen concentrate** used if fibrinogen is less than 500g/L
4. **Anti-fibrinolytic** (e.g. tranexamic acid) is used if there used sometimes for treatment of life threatening bleeds following thrombolytic therapy.

1. HAEMOPHILIA A: Deficiency of Factor VIII, X- linked recessive (only males are affected)

Signs & Symptoms:

- Depends on severity
- After trauma → bleeding into joints (haemarthrosis) & muscles (haematoma)
- Haematoma usually in buttocks

Investigation: Increased APTT, decreased factor VIII assay

Management:

- Avoid I.M injection
- In major bleeding → give factor VIII

1. **HAEMOPHILIA B**: This is deficiency of Factor IX. It is also called Christmas disease.

Treatment: Give factor IX

1. ANTI-COAGULANT THERAPY:

Indications:

- Deep vein thrombosis
- Pulmonary embolism

- Atrial fibrillation
- Stroke prevention
- Prosthetic heart valve

WARFARIN:

- Oral
- Narrow therapeutic range
- Inhibits reductase which is responsible for generating active vitamin K

INR targets: (NORMAL INR: 0.9-1.2)

1. Prosthetic heart valves → 3 – 4.9
2. Atrial fibrillation → 2- 3
3. Pulmonary embolism → 2- 3 (3.5 if recurrent)
4. Deep vein thrombosis → 2-3

Warfarin overdose causes increased INR and therefore increased PT

Management:

- **INR 5-8 and asymptomatic: does not usually require specific treatment. Only withhold warfarin until INR less than 5.**
- **INR >8 and asymptomatic: give vitamin K**
- **If there is minor bleeding: give vitamin K**
- **If there is severe bleeding: Requires urgent correction. Prothrombin complex concentrate (II, VII, IX, and X) is the preferred treatment for life threatening bleeding.**

NB. Fresh frozen plasma should only be used if prothrombin complex concentrate not available.

Heparin: Can be standard heparin or low molecular weight heparin

Treatment for Heparin toxicity → stop heparin, give protamine sulphate

4. BLEEDING IN LIVER DISEASE

The liver is involved in synthesis of clotting factors II, VII, IX, X which are Vitamin K dependent factors and also vitamin K

Obstructive jaundice: prolonged PT due to vitamin K deficiency

Liver cirrhosis: Increased PT, APTT, and TT; low fibrinogen

Management: Give vitamin K (Fresh Frozen Plasma is more effective as it contains the clotting factors)

5. MASSIVE TRANSFUSION/cardiopulmonary bypass

This causes **dilutional** thrombocytopenia. Therefore if a patient starts bleeding after massive blood transfusion, for example 10 units of blood, think of dilutional thrombocytopenia.

Treatment: Transfuse platelets.

ABNORMAL PLATELETS**1. THROMBOCYTOPENIA - This is low platelets**

Normal Platelet count is 150-400 x 10⁹ /L

Causes**1. Increased platelet consumption****Immune**

- Idiopathic (ITP = Idiopathic Thrombocytopenic Purpura)
- Drug induced
- SLE

Non –Immune

- Massive blood transfusion leads to dilutional thrombocytopenia
- Hypersplenism (platelets are destroyed in the spleen)
- DIC due to increased consumption of platelets
- TTP

2. Reduced platelet production

- Myelosuppressant (drugs, alcohol, viral illness) e.g. aplastic anaemia
- Bone marrow infiltration/failure
- B12 or folate deficiency (they take part in the production of platelets)

3. Abnormal platelet function

- The common disease of this type of **Von Willebrand disease**.
- In this case **bleeding time is increased**.
- In the exam if you see bleeding time increased think of Von Willebrand first.
- The next investigation would be to do platelet function studies

GENERAL MANAGEMENT

1. **For immune mediated thrombocytopenia use steroid e.g. prednisolone with or without immunoglobulin**
2. **In DIC/massive transfusion = transfuse platelets to keep platelets above 75**

Idiopathic Thrombocytopenic Purpura ⇒ marrow failure, virus, DIC, lymphoma, hypersplenism

Signs & Symptoms:

- ITP usually follows an upper respiratory tract infection, common in children, patient present with bleeding from nose but patient is generally well.
- Bleeding
- Purpura (dependent pressure areas)
- Epistaxis
- No splenomegaly
- Everything is normal (Patient is clinically well)

Investigations:

- Anti – platelet Ig G +VE

Treatment: Steroids e.g. Prednisolone.

VASCULAR DEFECTS

Usually this is vasculitis

Congenital:

- Osler – Weber – Rendu syndrome which is also known as Hereditary haemorrhagic telangiectasia (nose bleeds and mucosal bleeds in GMC scenario)

Acquired:

1. Trauma
2. Vasculitis e.g. in Henoch – Schonlein Purpura
3. Scurvy (vitamin C deficiency causing bleeding from gums and usually peri-follicular bleeding)

1. MALIGNANCIES IN HAEMATOLOGY

- a. **Leukaemia**
 - i. **Acute/Chronic**
 - ii. **Lymphoid/Myeloid**
- b. **Lymphoma**
 - i. **Hodgkin**
 - ii. **Non-Hodgkin**

- c. **Myeloma**
- d. **Myeloproliferative disease**

A. LEUKAEMIAS

CLASSIFICATION

Acute or Chronic: LOOK AT THE AGE

If less than 40 years old it is likely to be acute leukaemia

If more than 40 years old it is likely to be chronic leukaemia

Lymphoid or Myeloid: LOOK FOR LYMPHADENOPATHY

If there is lymphadenopathy it is likely to be lymphoid leukaemia otherwise it is myeloid leukaemia.

N.B. splenomegaly alone means myeloid

LOOK FOR BLOOD FINDINGS: If neutrophils are in the blood it's myeloid, if lymphocytes then it's lymphoid leukaemia.

1. ACUTE LYMPHOID LEUKAEMIA:

- Patient is less than 40 years
- Anaemia
- Infection → zoster, CMV, measles, candidiasis, pneumocytosis
- Bleeding
- Splenomegaly (hepatosplenomegaly)
- Lymphadenopathy
- Orchidomegaly
- CNS involvement eg. meningitis

Investigations:

1. Blood film shows lymphocytosis
2. Bone marrow biopsy

Complication: Commonly lymphadenopathy

Treatment:

1) Supportive care → blood & platelet transfusion, I.V antibiotics

2) Chemotherapy (main treatment)

Poor prognosis if adult, male, presence of Philadelphia chromosome 9:22, CNS signs, WCC > 100*10⁹

2. ACUTE MYELOID LEUKAEMIA (AML):

- Anaemia
- Infection
- Bleeding
- Bone pain
- CNS sign (cord compression)
- Hepatomegaly
- Splenomegaly (very common)
- Lymphadenopathy
- Weakness and malaise
- Fever
- Gum hypertrophy

Investigations:

- WCC variable/high
- Blast cell in blood
- Bone marrow biopsy

Complications:

- Infection
- **TUMOUR LYSIS SYNDROME** due to chemotherapy
- Rapid cell death on starting chemotherapy, which causes a rise in serum urate, potassium and phosphate.
- **Treatment: Initial step is IV fluids, then allopurinol and high fluid intake**
- Big nodes may cause mass effect: Shortness of breath by compression of trachea
- Splenomegaly

Treatment: same as for ALL (chemotherapy)

3. CHRONIC MYELOID LEUKAEMIA (CML):

- Middle aged people, associated with Philadelphia chromosome 9:22

Signs & Symptoms:

- Chronic
- Weight loss
- Tiredness
- Gout
- Fever
- Bleeding
- Abdominal pain
- Massive splenomegaly
- Hepatomegaly
- Anaemia
- Bruising

Investigations:

- Raised WBC $>100 \times 10^9$
- Haemoglobin levels are low or normal
- Platelets variable
- Increased urate (causing renal stones or gout) & alkaline phosphatase

Natural history:

- Chronic
- Accelerated phase → increased symptoms, spleen size
- Blast transformation with features of leukaemia

Treatment: Chemotherapy (Imatinib mesylate, Hydroxyurea)

4. CHRONIC LYMPHOID LEUKAEMIA:

- > 40 years
- More common in men
- Bleeding + low weight + infection + anorexia
- Enlarged rubbery lymph nodes
- Late stage hepatosplenomegaly

Investigations:

- Lymphocytosis
- Normocytic normochromic anaemia
- Low or normal platelets

Complications:

- Auto immune haemolytic anaemia
- Infection
- Bone marrow failure
- Lymphadenopathy

Treatment:

- **Chemotherapy**
- **Radiotherapy**
- **Supportive Care e.g. transfusion or antibiotics**

HODGKIN LYMPHOMA:

Associated with Epstein Barr Virus

Common in young black patients

Malignant proliferation of lymphocytes (Reed Sternberg cells)

Signs & Symptoms:

- **Enlarged painless rubbery lymph nodes, especially cervical**
- Fever, weight loss, lethargy, night sweats
- Hepatosplenomegaly
- Anaemia

Investigations: Lymph node biopsy

Staging:

- I → single lymph node
- II → 2 or more regions on same side of the diaphragm
- III → both sides of diaphragm
- IV → beyond lymph node metastasis

Needs to be differentiated from TB which has tender matted cervical lymphadenopathy and night sweats.

Treatment:

- **Radiotherapy for I A & II A**
- **Chemotherapy for IIA – IV B**

NON HODGKIN LYMPHOMA:

- Weight loss, fever, anorexia, night sweats
- **No Sternberg cells**
- Rapid extranodal spread (skin nodes)
- **SKIN NODULES**

MYELOMA:

Malignant proliferation of plasma B-cell

Elderly >70 years of age (typical symptoms include chronic back pain, hypercalcemia, anaemia, proteinuria and raised ESR)

Signs & Symptoms:

- Bone pain in back, ribs, long bones
- Pathological fracture e.g. vertebral collapse
- Fatigue (from anaemia), bleeding
- Pyogenic infection
- Hyperviscosity (visual disturbance, headache)
- Proteinuria

Investigations:

- Serum electrophoresis
- Urine (Bence Jones protein with paraproteinuria)
- Bone marrow (plasma B cell)
- X-ray → punched out lesion (pepper pot skull)
- High Urea and hypercalcemia

Treatment:

- **Chemotherapy is the main treatment**

- **Analgesia**
- **Transfusion**
- **High fluid intake**
- **Bisphosphonates (control the increase in calcium, & bone pain)**
- **Radiotherapy locally, it decreases the pain if the pain is secondary to bone metastasis.**

1. PANCYTOPENIA

This is when all 3 types of cells are reduced, i.e. low haemoglobin, white cell count and platelets

CAUSES

1. Bone marrow failure e.g. aplastic anaemia
2. Hypersplenism
3. SLE
4. Megaloblastic anaemia

CAUSES OF BONE MARROW FAILURE

1. **Aplastic anaemia:** In this one all the 3 cell types are reduced i.e. low red blood cell (low Hb), low white cell count and low platelets. This is what differentiates aplastic anaemia from leukaemia in that WCC is increased in leukaemia.

Aplastic anaemia can be caused by drugs like cytotoxic cyclophosphamide, radiation, autoimmune, drugs (gold), viral hepatitis etc.

1. **Infiltration** (malignancy, Tuberculosis)
2. **Myelofibrosis**

Signs & Symptoms:

- Anaemia (from low haemoglobin)
- Infection (from low white cell count)
- Bleeding (from low platelets)

Treatment:

Supportive to increase blood count while undertaking definitive which is bone marrow transplant.

5. MYELOPROLIFERATIVE DISORDERS

Classification:

1. Increased RBC → **Polycythaemia Rubra Vera**
2. Increased WBC → **Chronic Myeloid Leukaemia**
3. Increased Platelets → **Essential thrombocythaemia**
4. Increased fibroblasts → **Primary myelofibrosis**

A. POLYCYTHAEMIA RUBRA VERA

Signs & symptoms:

- Itch after hot bath + plethoric face (red appearance of face)
- Angina - may cause electric shock like pain
- CNS signs
- Raynauds
- Gout (Increased turnover of cells = ↑ uric acid)
- Enlarged spleen (60%)

Investigations:

- Increased PCV (packed cell volume)
 - Increased WBC
 - Increased RBC
 - Increased Platelets
- Blood film

- Bone marrow biopsy

Treatment: Venesection, chemotherapy e.g. hydroxyuria

B. ESSENTIAL THROMBOCYTHAEMIA:

- Raised platelets
- Abnormal function & shape
- Bleeding / thrombosis

Treatment: Chemotherapy

***MYELOFIBROSIS**

- De-arranged haematopoiesis in spleen
- Systemic upset
- Bone marrow failure

Investigation: Myeloblasts and teardrop RBCs in blood film

Resource start date 2013-06-26 08:40

Resource end date 2023-06-27 08:40

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