

Spleen 2 - Dr. Mzhda

Idiopathic Thrombocytopenic Purpura (ITP)

- Purpura is local hge. to the skin.

Etiology by:

1. Increase capillary fragility duo to drugs.... steroids
2. Thrombocytopathies by aspirin tablets
3. Thrombocytopenia:
 - A- Platelets production decrease as in aplastic anemia, cytotoxic drugs
 - B- Increase platelets consumption as in IVC in sepsis, large haemangiomas
 - C- Increase platelets destruction as in autoimmune disease SLE, infection as IMN, ITP, drug reaction as quinine
 - D- Increase splenic sequestration of platelets as any condition causing splenomegaly as portal hypertention

Clinical features:

- Skin and mucus membrane purpura
- Epistaxis
- Menorrhagia
- Simple wound bleeding
- Urinary, GIT, haemarthrosis rare
- Intracranial he
- Torniquet test +ve
- 25% has palpable spleen
- Huge splenomegaly suggests the diagnosis is not ITP.

Investigations:

- 1- Increase bleeding time (clotting and prothrombing time is normal)
- 2- Platelet count < 60 000
- 3- Bone marrow biopsy shows plentiful supply of megakaryocytes

Treatment:

- Depend on behavior of the disease and age of the patient but no splenectomy during acute attaches of ITP.
- Children 75% regress by cortisone and azothiaprine
- Splenectomy in severe cases & girls approaching menarche
- Adult and sever if ITP persist > 6-9 months do splenectomy
- By splenectomy 60% cured, 20% improved, 15% no benefit

Erythrocytes disorders (haemolytic anemia)

Cases amenable to splenectomy:

- 1- Hereditary spherocytosis
- 2- Acquired autoimmune H. anemia
- 3- Thalassemia
- 4- Hereditary elliptocytosis
- 5- Pyruvate kinase deficiency

1- Hereditary spherocytosis

- Due to dysfunction or deficiency in one of the RBC membrane proteins which increase permeability to Na.
- Mendelian autosomal dominant
- Male=female
- Increase RBC permeability to Na so more fragile.

Clinical features:

- Mild jaundice, anemia, splenomegaly
- Pale, lassitude and undue fatigue
- RBC crisis sometimes occur, precipitated by acute infection even may cause death (pyrexia, abdominal pain, N, V, extreme pallor and jaundice)
- In adult may develop jaundice, attacks of biliary colic

On examination

- Spleen enlargement palpable, sometimes liver also palpable chronic leg ulcer in adults

Investigations:

- 1- Fragility test (N is 0.47% NaCl haemolysis occur but it is 0.60% or even more)
- 2- Reticular count
- 3- Faecal urobilinogen is high
- 4- Radioactive chromium 51
- 5- Sonography to show gall stones

Treatment:

- Splenectomy: at about 7 - years of age to decrease gall stone formation and chance for infections

2-Acquired haemolytic anemia:

- Due to complement or Ig on RBC which triggered by macrophages Fc receptors in spleen(culling effect),caused by:-
 - drugs as methyl dopa
 - disease like SLE
 - unknown causes

Clinical features:

- Common > 50 years of age, more in female has mild jaundice and anemia
- Spleen enlargement 50%
- Gal stones 20%

Investigation:

- Coombs test is positive

Treatment:

- No treatment-acute self limiting
- Treatment with cortisone
- Splenectomy if:-
 1. corticosteroid ineffective
 2. patient had complications from using cortisone
 3. cortisone contraindicated as in PU disease
- 80% respond to splenectomy

3-Hereditary elliptocytosis:

- When the erythrocytes circulate it elongated so less sequestered.

4-Thalassemia (Mediterranean anemia, Cooley's Anemia)

- Due to Hb synthesis defect
- Dominant trait
- Alfa ,beta, gamma type ,but mostly is beta chain result in Hb A decrease and has intracellular (Heinz bodies) so premature RBC destruction occur
- From heterozygous minor to homozygous major

Clinical features:

- Chronic anemia, jaundice, splenomegaly
- Major in children cause retarded growth, enlarged head, slanting eyes, depressed nose, leg ulcer, jaundice and abdominal distention

Investigations:

1. Small RBC, resist osmolysis
2. Nucleated RBC
3. Hb electrophoresis

Treatment:

- Blood transfusion
- Splenectomy by:
 - Repeated blood transfusion
 - Uncomfortable and painful splenomegaly

5-Sickle cell anemia:

- is hereditary haemolytic anemia
- autosomal codominant
- common in Africans origin in which normal Hb A replaced by Hb S
- As Hb S crystallized when O₂ tension decrease so elongate, increase blood viscosity and obstruct the flow in open and closed system of the spleen circulation.
- cause painful intermittent episodes of abdominal pain which may result in auto-splenectomy

Clinical features:

- 9% Africans origin
- Mostly asymptomatic
- Symptoms depend on which organ is involved like; joint pain, priapism, neurological abnormality, skin ulcers, abdominal pain due to intestinal bowel ischemia.

Investigations:

- Blood film show sickle shaped cells
- Hb electrophoresis

Treatment:

- Avoid hypoxia specially during general anesthesia
- Adequate hydration
- Splenectomy may be helpful
- In streptococcus infection may precipitate the acute attacks so treatment is by giving RBC and splenectomy

Schistosomiasis

- 75% is sch. mansoni, 25% is sch. haematobium
- Common in Africa
- Splenic hyperplasia occurs due to phagocytosis of the disintegrated worms, ova, toxins and liver fibrosis which may cause portal hypertension.

Clinical features:

- male > female
- splenomegaly features

Investigations:

- Urine and stool for ova
- Liver function test
- Hb decrease

Treatment:

- Medical treatment
- Splenectomy if spleen is bulky and painful

Tuberculosis

- Not uncommon
- Age 20-40 years

Clinical features:

- Asthenia, loss of weight, evening fever + splenomegaly
- May produce portal hypertension
- Cause cold abscess
- Investigation: culture and guinea-pig inoculation

Treatment:

- Anti-tuberculous drugs + splenectomy if there is no active disease in the body.

Gauchers disease

- Storage of abnormal lipoid(glucocerebroside) in reticuloendothelial cells
- Common in Jewish & Slavonic races
- Rare disease
- Spleen may reach 4-5 kg.
- Appear < 12 years of age so in early childhood
- Bone pain ,pathological #, jaundice, anemia, yellow –brunish discoloration of the skin of hands& faces
- Conjunctiva thickening (pinguecula)
- Early satiety and abdominal distension due to splenomegaly
- Diagnosis by Gaucher cells in bone marrow

Treatment :

- By splenectomy may be helpful
- Partial splenectomy in children

Amyloidosis

- Abnormal extracellular protein deposition
- Multiple organs involved
- Clinically may be asymptomatic or patient come with MOF (multiple organ failure)
- If associated with splenomegaly do splenectomy to relief symptoms of splenomegaly

Sarcoidosis

- Inflammatory disease characterized by non-caseating granulomatous lesions affecting the tissue.
- Common in young adults
- Involve mostly lung
- Signs & symptoms are nonspecific as malaise, fatigue
- 25% has splenomegaly, and spontaneous rupture of spleen may occur
- Treatment is by splenectomy when signs and symptoms are present.

Felty's Syndrome

- Due to immune complex coated the surface of the leucocytes
- Is chronic arthritis + leucopenia < 2000c/dl + splenomegaly
- If light arthritis associated with the leucopenia and splenomegaly called primary splenic neutropenia.
- Treatment by giving corticosteroids, methotrexate
- Splenectomy may be helpful.

Neoplasms

- Haemangiomas , sometimes haemangiosarcoma treated by splenectomy
- Lymphoma is the common one treatment by splenectomy

Hypersplenism duo to portal hypertention

- Cause decrease of platelets & granulocytopenia
- Treatment by splenectomy + shunt

Tropical splenomegaly

- Especially in children occur
- Duo to malaria, kalazar, schistosomiasis, malnutrition
- Abnormal response to malaria parasite and unusual spices of plasmodia
- Huge splenomegaly 2-4 kg.
- Anemia+ thrombocytopenia
- Treatment by splenectomy, but in endemic area with malaria give antimalarial drugs (proguanil)

Splenectomy

Indications:

1. Trauma-blunt or penetrating
2. Iatrogenic. e.g. mobilization of splenic flexure in operation for ca rectum.
3. As part of other operations eg. Carcinoma of the stomach.
4. Staging of lymphoma
5. Erythrocytes disorders as hereditary spherocytosis,acquired haemolytic anemia, auto immune haemolytic anemia(Mediterranean anemia)...etc
6. Haemoglobinopathies as thalassemia, sickle cell anemia, porphyria.....etc
7. Leukocytes disorders as Feltys disease....etc
8. Platelets disorders as ITP....etc
9. Bone marrow disorders as myeloproliferative disease
10. Cysts: parasitic or nonparasitic.
11. Neoplastic as angioma, primary fibrosarcoma
12. Infections as: bacterial.....TB, protozoal & parasitesschisotomiasis, tropical splenomegaly
13. Abcess
14. Storage & infiltrative disease as: Gaucher s disease, sarcoidosis, amyloidosis
15. Miscellanouse disorders as: Portal hypertension, splenic vein thrombosis

Preoperative preparation for elective splenectomy

1. 7-10 days before operation give vaccination for pneumococcal.....
2. Hb more than 10 mg/dl
3. Prepare 2-4 unit blood for huge splenomegaly cases
4. Treat thrombocytopenia
5. If patient receiving oral cortisone, shift to cortisone schedule by giving intravenous cortisone.

Post-operative complications

1. Hemorrhage
2. Gastric dilatation
3. Haematemesis
4. Left basal atelectasis + pleural effusion, pneumonia
5. Left subphrenic abscess
6. Damage to tail of pancreas may cause local pancreatic abscess, pancreatic fistula, pancreatitis
7. Wound infection
8. Thrombo-embolic phenomena causing deep vein thrombosis, portal vein thrombosis
9. Gastric fistula
10. Postsplenectomy septicemia by overwhelming postsplenectomy infection (OPSI) by streptococcus pneumonia, nisseria meningitides, haemophilus influenzae, salmonella, bacteroids