Budd-Chiari Syndrome

Rare condition which occurs when there is obstruction of the hepatic veins.

Aetiology

- Haematological:
  - Polycythaemia vera and other myeloproliferative disorders
  - Thrombophilic conditions e.g. deficiencies of Protein C, Protein S, Antithrombin III or Factor V Leiden
  - Antiphospholipid antibody syndrome
  - Essential thrombocytosis
  - Paroxysmal nocturnal haemoglobinuria
  - Post bone marrow transplant
- Reduced blood flow: Webs in vena cava, CCF, constrictive pericarditis, RA myxoma
- Obstetric: The condition can occur during pregnancy and post-partum
- Drugs: Combined oral contraceptives, HRT, urethane
- Chronic infections: Hydatid disease, amoebic abscesses, aspergillosis, syphilis, TB
- Chronic Inflammatory Conditions: IBD, sarcoid, SLE, Sjogren's, Behçet's disease, etc
- Malignancy: Hepatocellular ca, renal cell ca, Wilms tumour, adrenal ca, leiomyosarcoma
- Others: Trauma, Alpha 1-antitrypsin deficiency, idiopathic (30%)

Epidemiology

Most commonly assoc with haematologic disorders, pregnancy or tumours in USA. IVC webs are the main cause in most patients from eastern Asia, India, and South Africa.

Presentation

Most commonly it presents gradually with ascites ± jaundice ± renal impairment. But may be sudden abdominal pain, rapidly developing ascites, hepatomegaly, jaundice, ARF ± liver failure.

Differential Diagnosis

- Cirrhosis
- Portal hypertension
- Portal vein thrombosis
- Right-sided heart failure
- Hepatic veno-occlusive disease (common after BMT)

Investigations

- LFT (mild).
- CT/MRI - prominent caudate lobe
- Doppler USS to exclude hepatic v. or IVC thrombosis.
- Caval venography excludes caval webs and occluded hepatic vv
- Ascitic fluid - high protein
- Liver biopsy

Management

- Ascites should be managed with diuretics plus fluid and salt restriction.
- Surgical decompression e.g. transjugular intrahepatic portosystemic shunt (TIPS).
- Balloon angioplasty ±/ stent for inferior vena caval web or short hepatic vein thrombosis.
- Liver transplantation may be appropriate if there is decompensated liver cirrhosis.

Complications

Hepatic failure ±encephalopathy, portal hypertension, oesophageal varices ±haemorrhage, hepatorenal syndrome (renal failure in patients with advanced chronic liver disease).

Prognosis

Better prognosis if younger age at Dx, absence of a small amount of ascites, and low serum Cr.

Miscellaneous vascular disorders of the liver

Acute cardiac failure and shock

Reduced hepatic blood flow causes ischaemia primarily in the centrilobular area, and may cause necrosis there. Deranged liver function tests may result and in severe causes, jaundice.

Chronic venous congestion

A persistently ↑central venous pressure results in hepatic venous congestion and hepatomegaly. The associated histological changes are of centrilobular congestion with surrounding fatty change ('nutmeg liver'). The serum bilirubin level is usually increased and there may be a slight elevation in the ALP and in the transaminases. True cirrhosis of the liver with regenerative nodules is very rare, as is portal hypertension; it is very unusual to find oesophageal varices in patients with cardiac cirrhosis.

Hepatic arterial occlusion

This rare condition usually follows surgical trauma but has been found in association with arteritis and bacterial endocarditis. There is an acute onset of pain in the upper abdomen, tenderness over the liver, and progressive shock and liver failure. Most cases have a fatal outcome.

Septic venous thrombosis of the portal system

This results from infection anywhere in the abdominal cavity leading to pylephlebitis of the portal venous system. It may occasionally result from a systemic septicaemia or from inflammatory disorders of the bowel. The acute phase has features related to the underlying abdominal sepsis. This is followed by high fever, worsening abdominal pain, and rigors. There may be obvious evidence of septic embolization to the liver, with abdominal pain and hepatic tenderness with mild jaundice. Occasionally, multiple large intrahepatic abscesses may develop. The condition should be suspected in any patient with abdominal sepsis who develops an acute systemic illness with abdominal pain and deranged liver tests. Management consists of intensive antibiotic treatment directed particularly towards Gram-negative organisms and micro-aerophilic streptococci. In some patients portal venous thrombosis may develop.

Peliosis hepatis

Uncommon vascular condition characterised by randomly distributed multiple blood-filled cavities throughout liver. Size of the cavities usually ranges between a few millimetres to 3 cm in diameter. Associations include AIDS, chronic infections (incl TB), malignancy, renal transplants and drugs (steroids, tamoxifen, androgens, anabolic steroids). Generally asymptomatic but may present with ↑LFTs, jaundice, hepatomegaly, liver failure. It also occasionally affects spleen, lymph nodes, lungs, kidneys, adrenal glands, bone marrow and other parts of gastrointestinal tract.