Budd-Chiari Syndrome

Introduction

Budd-Chiari syndrome (BCS) is a rare condition which occurs when there is obstruction of the hepatic veins. It includes any condition in which there is obstruction to venous flow from the small hepatic veins to the inferior vena cava. It should be differentiated from veno-occlusive disease in which the sinusoidal epithelial cells of the hepatic venules are damaged; this mainly occurs after stem-cell transplantation.

Chronic BCS is thought to have a genetic basis: there is a high prevalence of myeloproliferative disease in these patients.\[1\]

Aetiology

- **Haematological:**
  - Polycythaemia vera and other myeloproliferative disorders.
  - Thrombophilic conditions - eg, 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: vena caval abnormalities (eg, webs, congenital absence of part of the vessel), right heart failure, constrictive pericarditis, right atrial myxoma.
  - Obstetric: the condition can occur during pregnancy and postpartum.
  - Drugs: combined oral contraceptives, hormone replacement therapy, urethane.
  - Chronic infections: hydatid disease, amoebic abscesses, aspergillosis, syphilis, tuberculosis.
  - Chronic inflammatory conditions: inflammatory bowel disease, sarcoid, systemic lupus erythematosus, Sjögren's syndrome, Behçet's disease (3.2% in one study\[2\]), mixed connective tissue disease.
  - Trauma.
  - Surgery.
  - Others: alpha 1-antitrypsin deficiency, idiopathic (30%).

Epidemiology

The global prevalence of BCS is unknown but is estimated at 1 in 100,000.\[3\] The epidemiology and presentation tend to vary in different parts of the world.

A Swedish review estimated a mean age-standardised incidence in 1990-2001 of 0.8 per million per year and a prevalence rate of 1.4 per million inhabitants.\[4\]

Idiopathic forms are common in South Asia, while hypercoagulable disorders are common causes in the West. The site of thrombosis is also different, with patients from South Asia presenting with combined obstruction of the hepatic veins and the inferior vena cava in contrast to isolated obstruction of the hepatic veins in the West.\[5\]

In Turkey, the most common causes are hydatid disease and Behçet's disease.\[6\]

Risk factors

In the Swedish series, the main risk factors identified were myeloproliferative disorders (38%), thrombophilic factors (31%) and oral contraceptives (30%). About half of the patients had a multifactorial aetiopathology.\[4\]

Presentation

The presentation can be very varied and so the diagnosis must be considered in any patient with acute or chronic liver disease.\[7\]

- It generally requires more than one hepatic vein to be obstructed for symptoms to be present.
- Presentation can be sudden with right upper quadrant pain and rapidly developing ascites, hepatomegaly, jaundice and acute kidney injury (± fulminant hepatic failure ± hepatic coma).
- Most commonly it presents gradually with ascites (jaundice is commonly absent).
- Just under 50% of such patients will also have renal impairment.
- Other examination findings may include dilated veins running over the abdominal wall and chest and splenomegaly.

Differential diagnosis
• Cirrhosis.
• Portal hypertension.
• Portal vein thrombosis.\[8\]
• Hepatic veno-occlusive disease (common after bone marrow transplantation).
• Right-sided heart failure.
• Appendicitis.
• Biliary atresia.
• Chronic granulomatous disease.
• Congenital hepatic fibrosis.
• Cystic fibrosis.
• Cytomegalovirus infection.
• Intussusception.
• Multicystic renal dysplasia.
• Nephrotic syndrome.
• Pancreatitis and pancreatic pseudocyst.
• Constrictive pericarditis.
• Syphilis.
• Toxoplasmosis.

Investigations\[7\]
• LFTs - these may show a mild elevation.
• Prothrombin time - this may be prolonged, which may be confusing if the condition is associated with a hypercoagulability state.
• Ascitic fluid - this usually has high-protein content (except if the onset is very acute) but the risks (eg, bacterial peritonitis) and benefits of paracentesis should be considered before this procedure is undertaken.
• Radio-imaging - MRI may show a prominent caudate lobe (the bit to the left of the portal vein when facing the patient). It is more sensitive than CT scan. 3D MRI angiography is a useful recent enhancement.\[9\]
• Doppler ultrasound may help to exclude hepatic venous or inferior vena caval thrombosis. One study identified altered hepatic and/or caval veins and caudate lobe hypertrophy as being the findings most commonly associated with BCS.\[10\]
• Caval venography excludes caval webs and occluded hepatic veins.
• Liver biopsy often shows centrilobular congestion.

Management
• Underlying conditions should be treated.
• Any underlying haematological condition should be treated (eg, with anticoagulation).
• BCS associated with chronic inferior vena caval thrombosis has been treated safely with warfarin.\[11\]
• Ascites should be managed with diuretics (spironolactone is first-line, followed by furosemide and chlorothiazide) plus fluid and salt restriction.
• Local thrombolysis with radiological support is preferred to generalised thrombolysis.
• Surgical decompression of liver is performed in cases of persistent congestion (eg, via transjugular intrahepatic portosystemic shunt (TIPS)) and gives excellent results, even in high-risk patients. Other shunts sometimes tried include mesocaval, mesoastral and portocaval.\[12\]
• There is indication for balloon angioplasty ± stent for inferior vena caval web and sometimes in hepatic vein thrombosis if the affected length of vein is not extensive. The percutaneous route may be used and has proved safe and effective.
• Liver transplantation may be appropriate if there is decompensated liver cirrhosis.\[13\] Reconstructing hepatic venous outflow post-transplant is sometimes a problem but successful venoplasty using autologous vein grafts has been reported.\[14\]

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

Prognosis
• Factors associated with a good prognosis include younger age at diagnosis, absence or a small amount of ascites, and a low serum creatinine level.
• Identification of the cause of the symptoms using modern radio-imaging techniques has had a major effect on prognosis.
• Interventional and medical treatment have helped to keep patients alive for up to eight years, after which liver transplantation is considered.
• Without treatment, BCS is often fatal.\[15\]
• With treatment, the five-year survival may be as high as 90%.\[16\]
• Portal hypertension and portal vein thrombosis carry a poor prognosis.\[10\]
• A case of fulminant liver failure associated with BCS, hepatic vein invasion and renal cell carcinoma has been reported.\[17\]

Further reading & references

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