

Budd Chiari Syndrome - A Case Report

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Abstract: *Budd-Chiari Syndrome (BCS) is a rare disorder characterised by hepatic venous outflow obstruction. Presentation can range from a fully asymptomatic disorder to a fulminant failure of the liver. BCS is an example of post-inusoidal portal hypertension. Management may be classified into three major categories: medical, surgical and endovascular.*

Keywords: Budd, chiari, syndrome, hepatic, veins, obstruction

1. Introduction

The word "Budd-Chiari" was created in the late 1800s after the work of George Budd, an internist, who identified three cases of hepatic vein thrombosis in 1845, and Hans Chiari, an Austrian pathologist, who reported the first pathological description of hepatic endophlebitis obliterated in 1899. The current definition of BCS includes a variety of conditions that induce obstruction of the hepatic outflow from small hepatic veins to the junction of inferior vena cava [1]. BCS consists of a heterogeneous group of disorders characterised by partial or full hepatic venous outflow obstruction. There is a rise in hepatic sinusoidal pressure secondary to hepatic venous outflow obstruction. This causes in portal hypertension and congestion of the liver. This leads to hypoxia and hepatocyte dysfunction. If the obstruction is serious and has not been resolved in a timely manner, hepatocyte necrosis is present. This gradually leads to hepatic fibrosis and cirrhosis [2].

2. Case Report

A 30-year-old man presented with bilateral lower extremity edema for the previous 1 week and abdominal swelling. He had prominent tachycardia (heart rate 124/min) and a distended abdomen with the presence of shifting dullness, suggestive of ascites. Laboratory studies revealed normal bilirubin and creatinine levels, increased prothrombin time and elevated liver enzymes. Abdominal ultrasound revealed moderate ascites with heterogeneous hepatomegaly and obliteration of hepatic veins. Contrast enhanced computed tomography (Fig 1) was performed and showed ascites, hypertrophy of caudate lobe with narrowing of the retro-hepatic IVC and lack of opacification of the hepatic veins, suggesting occlusion. Based on the findings, diagnosis of Budd-Chiari Syndrome was made.

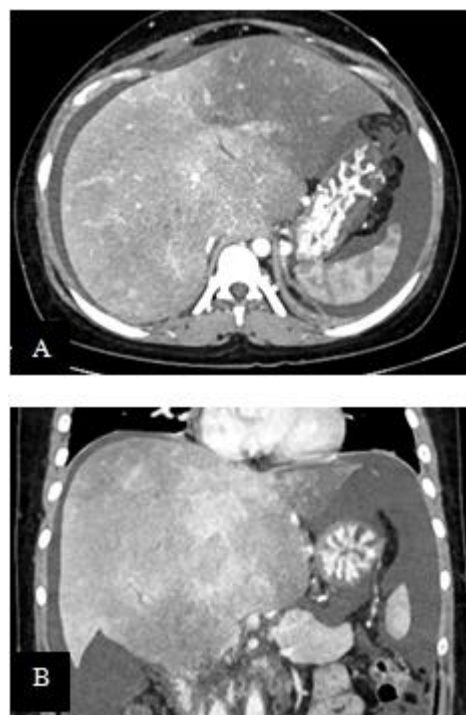


Figure 1

Fig 1: Contrast enhanced Computed tomography of abdomen Axial section (A) and coronal section (B) shows heterogeneous liver, ascites, hypertrophy of caudate lobe with narrowing of the retro-hepatic IVC and lack of opacification of the hepatic veins.

3. Discussion

The Budd-Chiari Syndrome (BCS) is an unusual disorder characterised by hepatic venous outflow obstruction; it has been documented to occur in 1 in 100,000 of the population worldwide. BCS is categorised as primary where obstruction of hepatic venous outflow is linked to a primary venous problem, such as thrombosis, stenosis, or webs, and secondary where associated with extrinsic compression, such as that caused by abscess, tumour, cyst, or hyperplastic nodules. The presentation and cause of the BCS which differ

based on the geographic region of the presentation[1]. It is important to have a high level of clinical suspicion when making a diagnosis; or else, there may be disruptions in management and therapy. Serum transferase levels may be more than five times the upper limit of the normal range, especially in fulminant and acute BCS forms [3]. Imaging also plays an important part in post-operative assessment and monitoring of shunts. This is performed by Doppler ultrasound and is complemented by Computed tomography (CT) and/or magnetic resonance imaging (MRI). Doppler ultrasound, frequently the first sample, has a sensitivity and accuracy of up to 85 per cent. Some radiographic findings seen in BCS include inhomogeneous parenchymal improvement, intrahepatic collateral, hyper vascular nodules, and caudate lobe hypertrophy. The caudate lobe has direct venous drainage into the IVC and is thus frequently susceptible to compensated hypertrophy. Liver biopsy is rarely used to diagnose BCS due to advances in imaging. Liver biopsy can show a wide range of histopathological findings from sinusoidal obstruction, cirrhosis inflammation and parenchymal necrosis. The extravagance of red cells to the liver cell plate and the Disse room is a distinctive function of BCS [4]. Treatment relies on the underlying issue and nature of the condition, including anticoagulant therapy, stent insertion, surgical or transjugular portosystemic shunt and liver transplantation. In our patient, BCS has been interpreted as secondary to chronic inflammatory disease / immunodeficiency. Angiography was performed and stent was inserted in the left hepatic vein and IVC with reduction of ascites and clinical progress. The BCS should be regarded as a diagnostic probability in the presence of heterogeneous liver and non-visualization of IVC/ hepatic veins [5].

4. Conclusion

BCS is a rare clinical disorder that needs precise, timely diagnosis and intensive treatment. Treatment will vary depending on the clinical presentation, cause and anatomical location of the problem. Health, surgical and endovascular treatment approaches for the control of this disorder are available.

References

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