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Case Report

Inferior vena caval web causing Budd Chiari syndrome

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ABSTRACT

Budd Chiari syndrome (BCS) is a condition resulting from obstruction of hepatic venous outflow at various levels, regardless of the cause of obstruction. Manifestations can range from asymptomatic state to fulminant hepatic failure or cirrhosis depending on how acutely the obstruction developed. IVC web is an unusual yet potentially treatable cause of BCS. This case report throws light on a rare cause of BCS, which is an IVC web and how endovascular technique helped in patient recovery.

Keywords: Inferior vena cava, Budd Chiari syndrome, Hepatic venous outflow obstruction

INTRODUCTION

Budd-Chiari syndrome (BCS) is a rare condition resulting from hepatic venous outflow obstruction (HVOO) at any level from small hepatic veins to the IVC and right atrium.[1] The majority of cases of BCS are associated with hypercoagulable states causing thrombosis in hepatic veins, with myeloproliferative disorders accounting for nearly 50% of cases. The classical triad of BCS consist of abdominal pain, tender hepatomegaly and ascites. Here we discussed a rare cause of BCS caused by an IVC web.

CASE REPORT

This is the case of a 38 year old gentleman, with no known comorbidities presented with complaints of insidious onset of progressive abdominal distention of 2 weeks duration and swelling of bilateral lower limbs of 10 days duration. There was no history of abdominal pain, vomiting, loose stools, constipation, fever, melena, haematemesis, fever, jaundice, altered sensorium, oliguria, frothy urine, dyspnoea, palpitation. On examination, patient was moderately built and nourished. Vitals reported BP-130/80 mm of Hg and pulse rate- 86 per min regular. Icterus and bilateral lower limb pitting oedema was present extending up to the thigh (Figure 4). No stigmata of chronic liver disease present. Cardiovascular system- JVP was elevated with absent hepatojugular reflex. S1 S2 was normal with no murmur. Respiratory system- normal vesicular breath sounds bilaterally with breath sound intensity reduced in right infra scapular area, infra-axillary area with dull note in these areas on percussion. Per abdomen- distended, tense, non-tender with dilated veins both above and below diaphragm which filled from below upwards. Traube's space was dull. Liver and spleen- not palpable by dipping palpation. Genitalia- hydrocele, penile oedema and scrotal excoriation was present. Nervous system examination was within normal limit. Investigations observed total count 10,800 N50L29M21, Hb- 13, platelet count- 2.3 lakhs. PT INR 15.4/1.24, aPTT-40/30, RBS- 156, RFT- 23/1, Na+/K+- 128/4.1, LFT- total bilirubin/direct bilirubin- 2.2/0.9, total protein /albumin-5.6/2.5, AST/ALT/ALP- 69/96/100. Ascitic fluid study showed high SAAG (1.7) ascites with total count of 300 cells with 50% polymorphs and 50% lymphocytes. Ascitic fluid total protein was 1.37, sugar- 126, albumin 0.8. So, considering the causes of portal hypertension, hepatic and pre-hepatic causes were unlikely as the patient did not have features of cirrhosis, splenomegaly or variceal bleed.

Among the post- hepatic causes, patient lacked features of pericarditis, tricuspid regurgitation and right heart failure. Hence, the strong possibility considered was BCS. We then proceeded with the ultrasound Doppler which showed echogenic thrombus noted in IVC of maximum thickness 1.9 cm in intra hepatic and infra hepatic portion. Hepatic vein showed normal colour flow, normal liver echoes with gross ascites and bilateral pleural effusion. CECT abdomen, showed inferior vena caval thrombus involving the hepatic and suprarenal segment with extension to middle hepatic vein, with hepatic attenuation difference, mild caudate hypertrophy with early caudate enhancement, suggestive of acute BCS confirming our clinical diagnosis. Our next task was to find out the cause for BCS. For which we did thrombophilia work up including APLA panel- Beta 2 glycoprotein, lupus anticoagulant, anti-cardiolipin antibody, s. homocysteine, protein C, protein S, ANA-IF, all were negative. Work up for myeloproliferative disorders- peripheral smear examination- normal and BCR-ABL and JAK2 mutation-negative.

Interventional radiology opinion was sought. Suction thrombectomy and thrombolysis using urokinase was planned. Venography was performed via right femoral access and was able to demonstrate IVC thrombus, seen as total cut off of infra renal IVC with numerous collaterals along azygous system draining to Superior Vena-cava (Figure 1). This was the reason for the elevated JVP despite thrombosis of IVC. Suction thrombectomy was done. Check venogram showed web at IVC- right atrial junction (Figure 3). Via combined common femoral vein and internal jugular vein access double balloon angioplasty of web was done. Check venogram showed partially opened up stenosis with significant forward flow (Figure 2). Post procedure there was significant reduction in ascites and scroto-penile oedema and lower limb oedema (Figure 4). There was plan for further check venogram and venoplasty, but venoplasty was abandoned as the patient symptomatically improved and also due to financial constraints. Patient fully recovered of his symptoms during follow up.

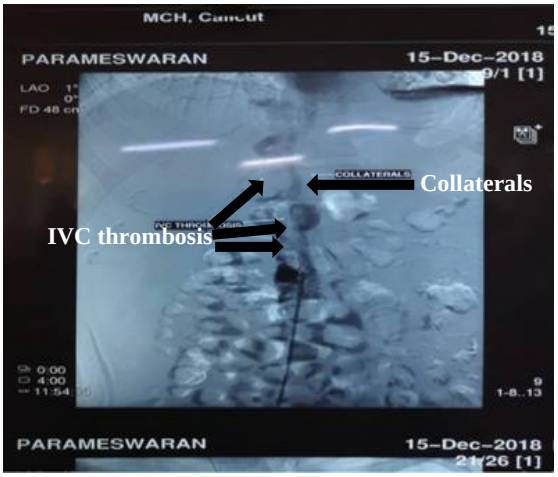

Figure 1: IVC thrombosis.

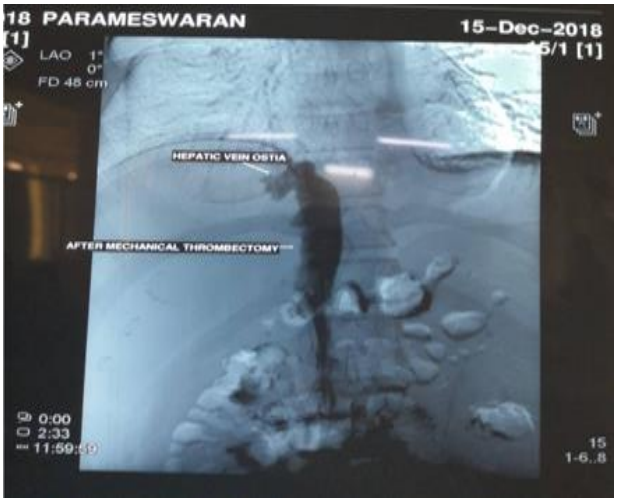

Figure 2: After mechanical thrombectomy, showing significant forward flow.

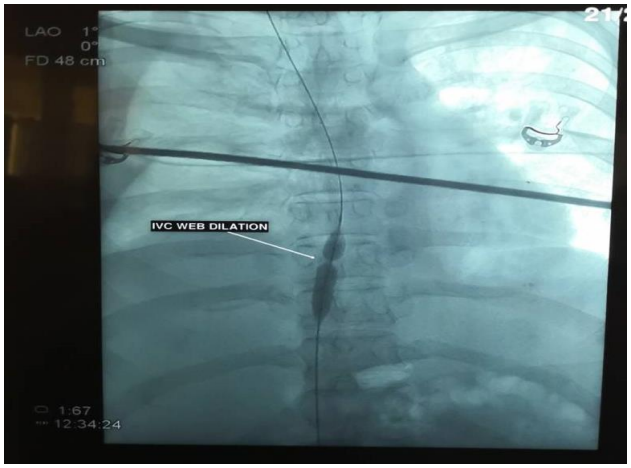

Figure 3: IVC web.

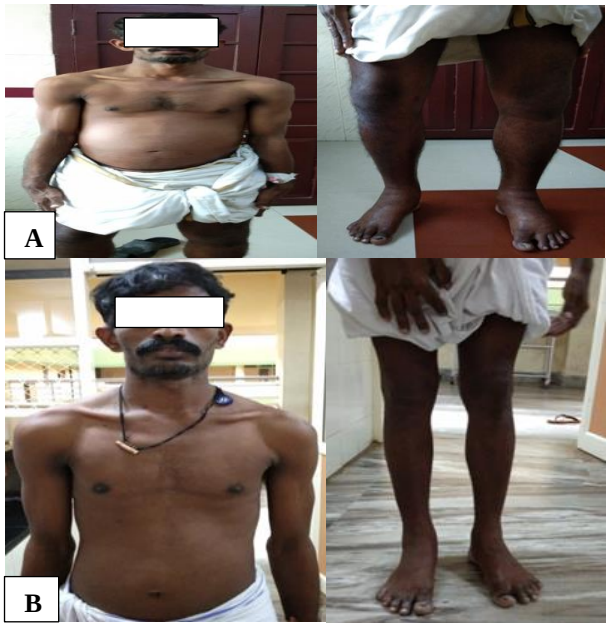

Figure 4: (A) Pre-procedure; and (B) post-procedure.

DISCUSSION

BCS is characterized by the incomplete or complete obstruction of hepatic venous outflow. Hepatic veins are the only conduits of blood from liver to IVC and heart. HVOO leads to increased hepatic venous pressure, resulting in venous collateral formation. In addition, increased hepatic venous pressure results in elevated sinusoidal pressure, portal hypertension and portal vein thrombosis. This cascade of events ultimately results in hepatocyte dysfunction leading onto liver failure.

Aetiology of BCS is divided into primary and secondary. Primary BCS can be due to intrinsic intraluminal thrombosis or webs. Majority of BCS up to 75% cases are due to hypercoagulable states like protein C, protein S, deficiency, factor V laden mutation, anti- phospholipid antibody syndrome, paroxysmal nocturnal haemoglobinuria, oral contraceptive use, pregnancy, SLE. Other conditions include myeloproliferative disorders like polycythaemia rubra vera, essential thrombocytosis, myelofibrosis which are also associated with thrombosis.[2] Membranous obstruction or IVC web also known as obliterative hepatocavopathy mostly due to post thrombotic sequelae.[3] Web histopathology has revealed intima replaced with fibrous laminar structure, organized thrombi, recanalization and calcifications. This fact can corelate well clinically with late onset of disease in some patients.

BCS presentation can range from an asymptomatic state to fulminant hepatic failure or cirrhosis depending on how acutely the obstruction developed. Treatment varies according to onset (acute verses chronic) and severity (fulminant hepatic failure vers decompensated cirrhosis or asymptomatic). Acute BCS is fatal if untreated. Aggressive interventions like thrombolysis or stenting which allow recanalization and helps to relieve hepatic congestion are used when the disease is acute (that is within 4 weeks of onset and in the absence of cirrhosis). Other options include conservative management with diuretic therapy and paracentesis or surgical portosystemic shunt. However, these options are palliative at best. Portosystemic shunting is associated with significant operative mortality and morbidity but improves long term survival in comparison with conservative management.[7] Although liver transplantation has been done in patients with the BCS techniques like angioplasty and stent placement to relieve the HVOO may obviate the need for more invasive and risky procedures.[4,5]

CONCLUSION

This case demonstrates one of the rare causes of BCS that is inferior vena caval web. Knowledge about the existence of such a remediable lesion is important, and its recognition and early intervention can help prevent the complications like hepatic failure and cirrhosis. Endovascular intervention which include balloon dilation and stent placement has become the first line therapy for primary BCS due to its minimal invasiveness and efficacy.

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