

A Rare Case of Budd Chiari Syndrome in a Child

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Abstract

A 7-year-old male child presented with the complaints of tense abdominal distension and swelling over feet since 1 month. The patient had repeated episodes of similar complaints since last two years with partial or complete relief after taking various forms of allopathic therapy. On imaging, Budd-Chiari syndrome was diagnosed which was hallmarked by occluded Inferior vena cava (IVC), caudate lobe enlargement and heterogeneous liver enhancement. Initially thrombolysis done leading to complete remission for the patient for 15 days, later on again the patient developed similar complaints for which balloon venoplasty and hepatic vein stenting was done.

Key Words: Ascites, Hepatic venous obstruction, Venography.

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1- INTRODUCTION

Budd-Chiari syndrome (BCS) is a rare condition induced by thrombotic or non-thrombotic obstruction of the hepatic venous outflow which is characterized by hepatomegaly, ascites, and abdominal pain. The classical BCS were first described by Budd in 1845 (1, 2), and later by Hans Chiari in 1899 (2, 3). The primary BCS is defined as hepatic outflow obstruction regardless of the cause or level of obstruction. The obstruction can range from the small hepatic veins (HV) to the orifice of the inferior vena cava (IVC) into the right atrium. Secondary BCS is defined as a hepatic venous outflow obstruction due to compression or invasion by extra vascular lesions, including benign or malignant diseases such as abscesses, hepatocellular carcinomas, and renal cell carcinomas, or secondary to cardiac or pericardial diseases.

Obstruction of large or small-caliber veins leads to hepatic congestion as blood flows into, but not out of, the liver. Microvascular ischemia due to congestion causes hepatocellular injury. Portal hypertension and liver insufficiency result. The BCS is diagnosed by radiological imaging. More than half of the cases of classic BCS occur between the ages of 20 and 39 years (1-3). Because of the rarity of this lesion in the pediatric age group, a good clinical suspicion is required along with radiological investigations to confirm the diagnosis.

2- CASE REPORT

A 7- year- old boy child admitted in AVBR Hospital, Sawangi (M), Wardha, India, with swelling over the abdomen since 1 month, gradual in onset and began from the abdomen and progressed to the legs. The patient also gave history of decreased frequency of urination. Six months earlier, during the first admission of the patient, Doppler ultrasonography of HBS showed thrombus extending from the

IVC into the walls of right atrium with no evidence of intrahepatic venous obstruction. The patient was taken up for thrombolysis after which, the patient was started on anticoagulants. A follow-up IVC Doppler showed 70% reestablishment of flow. Patient improved symptomatically. The patient was again brought with similar complaints after 15 days. Patient was taken up for IVC venography during the visit which revealed narrowing of the entire IVC along with involvement of hepatic vein could be due to fibrosis.

IVC venoplasty along with hepatic vein venoplasty was done and the patient again showed marked improvement discharged. The patient again presented after 15 days with similar complains with Doppler suggestive of blockage in the IVC for which, IVC balloon venoplasty was done. After this procedure, patient was discharged on anticoagulants as usual but the abdominal distention did not resolve completely. Hence after a few days the patient came back so, this time child was taken up for IVC balloon venoplasty with hepatic vein stenting and continued with anticoagulation therapy. After the procedure, a review color Doppler was done which showed good flow across the hepatic vein stent and the patient was showing steady signs of improvement.

3- DISCUSSION

Budd-Chiari syndrome (BCS) is very rare in children as compared to adults and most authors mentioned an incidence of about 1 case per million people per year in the general population of the world. Dilawari et al. (4), found that 177 cases of this syndrome, only 5% were found to be below 12 years of age (2). Hoffman HD et al. (5) reported the largest pediatric series of 9 cases with a membranous obstruction of the IVC. Most of the cases of classic BCS occur between the ages of 20 and 39 years. The primary BCS when related to a primarily venous disease (thrombosis or

phlebitis), and secondary BCS when related to compression or invasion by a lesion outside the veins such as tumors, abscesses, intrahepatic cysts and hematomas (6). The main pathophysiological event is venous flow obstruction between hepatic venules and the suprahepatic segment of the IVC. Silent BCS means occlusion of a single hepatic vein whereas overt BCS means the occlusion of at least 2 hepatic veins. The obstruction of the HV may lead to venous congestion of the liver which causes hepatomegaly. The chronic changes after blockage includes Centrilobular fibrosis which may be seen within weeks, and periportal nodular regenerative, progressive fibrosis and cirrhosis which may not appear for months.

The classic triad of BCS is abdominal pain, ascites, and hepatomegaly. Physical examination may reveal the jaundice, ascites, hepatomegaly, splenomegaly, ankle edema and prominence of collateral veins. Regardless of the cause, patients with develop post sinusoidal portal hypertension, which leads to complications similar to those observed in patients with cirrhosis. However, the systemic hemodynamic effects differ than those that are observed classically in patients with cirrhosis. Harmanci et al. (7), revealed long term follow up study of 62 patients with primary BCS.

They found an acquired cause of BCS in 64.5% cases, whereas in 9.7%, no identifiable cause was detected. A diagnosis of Budd-Chiari syndrome should be considered in any patient who presents with acute or chronic liver disease, as the clinical manifestations are extremely diverse. The most common cause in adults is thrombosis, which is secondary to an underlying myeloproliferative disease and in children; the most common cause is a membranous obstruction of the inferior vena cava (8-9).

Liver function tests include Aminotransferases and alkaline phosphatase can be normal or increased. Serum albumin, serum bilirubin and prothrombin level can be normal or abnormal, and in some cases are markedly increased. Ascitic fluid examination provides useful clues to the diagnosis, including high protein concentrations (>3g/dl), and serum ascites-albumin concentration gradient >1.1g/dl are suggestive of BCS. Serum creatinine level can be elevated, usually due to prerenal dysfunction.

Doppler ultrasound, venography and liver biopsy have been very helpful in its diagnosis.

Doppler ultrasonography findings can be specific for HV obstruction which include large HV with absent flow signal or reversed, or turbulent flow, and an absent or flat HV waveform without fluttering. CT scan will show a pattern of patches with increased enhancement in the central portion of the liver and decreased enhancement in the peripheral region due to portal backflow in acute BCS whereas liver atrophy with an enlarged caudate lobe and multiple intrahepatic and extra hepatic collateral veins seen in subacute and chronic BCS.

Hepatic venography may show the following changes: complete occlusion of hepatic vein, may or may not be associated with a stenosis of the intrahepatic IVC, spider web appearance and intrahepatic collateral veins. Hepatic venography showed narrow occluded veins in our case. Inferior vena cavography should be performed to demonstrate stenosis or occlusion of the IVC. Digital subtraction angiography (DSA) is the gold standard for evaluation of the IVC and hepatic veins. A liver biopsy with the presence of necrosis, suggests a need for porto-systemic shunts, whereas an extensive fibrosis favours a liver transplant (10-12). The treatment can be medical therapy,

radiological procedures and surgical with anticoagulation, especially in case of underlying hematological disorder as the cause of the BCS. Long term anticoagulation is often needed. Common surgical shunts are intrahepatic portosystemic shunt (TIPS), and side-to-side portacaval shunt. Liver transplantation is the preferred option where shunting is not possible or in fulminant cases.

4- CONCLUSION

Budd-Chiari syndrome is a rare clinical entity characterized by hepatic venous outflow obstruction. Commonly it is misdiagnosed as a case of only chronic liver disease in children but a good venography can confirmed the diagnosis like this patient. This case report highlights a rare case of BCS in a child.

5- CONFLICT OF INTEREST: None.

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