



A CASE OF BUDD CHIARI SYNDROME IN ASSOCIATION WITH MASSIVE ASCITES WITH RADIOLOGICAL EVIDENCE.

Radiodiagnosis

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ABSTRACT

Budd-Chiari syndrome (BCS) is a rare condition induced by thrombotic or nonthrombotic obstruction of the hepatic venous outflow which is characterized by hepatomegaly, ascites, and abdominal pain. We report a 10-year-old male child diagnosed as a case of BUDD CHIARI disease associated with massive ascites. The clinical and laboratory findings were compatible with diagnosis. On imaging, diagnosis of BUDD CHIARI with associated ascites Budd-Chiari syndrome was diagnosed which was hallmarked by marked attenuation of veins, caudate lobe enlargement.

KEYWORDS

Budd Chiari, Ascites, imaging.

INTRODUCTION

Budd-Chiari syndrome (BCS) is a heterogeneous group of conditions characterized by complete or partial obstruction of hepatic venous outflow.¹⁻³ There is an increase in hepatic sinusoidal pressure secondary to hepatic venous outflow obstruction. This results in portal hypertension and liver congestion which can result in hypoxia and hepatocyte dysfunction.^{2,3}

Budd-Chiari syndrome (BCS) is a rare condition induced by thrombotic or nonthrombotic obstruction of the hepatic venous outflow which is characterized by hepatomegaly, ascites, and abdominal pain. 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.

The BCS is diagnosed by radiological imaging. 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.

CASE REPORT

A 10 year- old boy admitted in MMIMSR, MMDU, Mullana Ambala, India, with complaints of pain abdomen from 7 days associated with multiple episodes of vomiting, complaint of fever spikes, and complaint of unable to pass stools from past 2 days with a significant past history of treatment Chronic liver disease from last 5 years but the records were not available. On abdominal examination, abdomen was distended and liver was palpable 6 cm below the right costal margin, fluid thrill was present. No signs of Hepatic encephalopathy was present.

Ascitic Tap was done 500 ml of straw coloured fluid was collected and sent for the following investigations.

TLC OF FLUID	55cells/cumm
DLC OF FLUID	43%Polymorphs, 57%Lymphocytes
Glucose	170mg/dl
Protein	1.4gm/dl
ADA	9.7U/L

On admission, Complete hemogram (hemoglobin-7.4gm% microcytic hypochromic picture was seen), Liver function test (deranged SGOT -159 U/L, SGPT -102U/L). Renal function test (normal), urine routine (normal), Pro thrombin time(20.2 seconds), INR(2.26 seconds) and activated partial thromboplastin time (48.7

seconds), Viral markers sent came out to be negative and on ophthalmology opinion no KF rings were seen.

Doppler Ultrasonography of abdomen was done giving impression of non visualised fibrosed hepatic vein with multiple venovenous collaterals with wildly coarsened echotexture of liver, suggesting of BUDD CHIARI with associated ascites.

Following this CECT Abdomen was done giving impression of

- Hepatomegaly with heterogeneous enhancement pattern with non visualised of the hepatic veins and compression of the hepatic vein.
- Massive ascites
- Splenomegaly

Suggesting BUDD CHIARI SYNDROME.

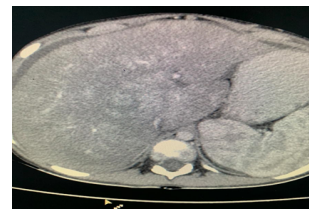


Image 1- Venous Phase of Triphasic CT showing Marked Attenuation of veins in liver. Liver showing non enhancing areas with enlargement of caudate nucleus.

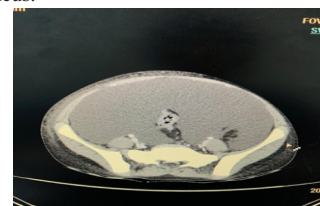


Image 2- Massive Ascites In the peritoneal cavity.

Following this During the course of hospitalization opinion was taken with Deptt of Gastroenterology, Patient was managed with oral WARFRIN, I/V ceftriaxone, oral lactulose and conservative treatment. Patient was diagnosed as a case of Budd Chiari in association with Massive Ascites, discharged on warfarin and kept on follow up weekly.

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. Hoffman HD et al⁵ 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 hematomas⁶.

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.

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.

CONCLUSION

Budd-Chiari syndrome is a 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 Radiological evidence can confirmed the diagnosis like this patient. This case report highlights a rare case of BCS in a child.

CONFLICT OF INTEREST: None.

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