***Case report***

**Acute Budd-Chiari Syndrome in a Young Patient: Diagnostic and Therapeutic Challenges**

**ABSTRACT:**

Budd-chiari syndrome is a condition in which hepatic venous outflow is obstructed. Budd-Chiari Syndrome is a rare but serious condition that can lead to significant liver damage if not diagnosed and treated early. Budd-Chiari syndrome (BCS) has a low incidence and prevalence; research indicates that there is about one case per million people annually.

To improve outcomes associated with this condition, prompt diagnosis and treatment are essential. This case report presents the clinical course of an 1-year-old male patient with no prior comorbidities presented with a chief complaints of right upper abdominal tightness which increased on intake of food, abdominal distension since 2 months which is insidious onset and gradually progressive and fever since 2 days. In this case, the patient’s clinical and physical examination showed altered echotexture of liver, caudate lobe hypertrophy with non-visualisation of hepatic venous flow and narrow calibre intrahepatic inferior venacava which was found to be positive for Budd-Chiari syndrome.

Anticoagulation is needed in some patients, especially those with underlying hematologic disorders as the cause of Budd-Chiari syndrome. Prothrombin time and activated partial thromboplastin time should be monitored once anticoagulation is started and should be maintained within the therapeutic range. Also monitor ALT,AST INR(international normalised ratio)levels throughout the therapy This case highlights the importance of a thorough clinical evaluation, early imaging, and appropriate management strategies to improve outcomes and prevent long-term complications.

**KEY WORDS:**

Budd-Chiari syndrome, thrombosis, anticoagulant therapy, hepatic vein

**INTRODUCTION**

A rare condition called Budd-Chiari syndrome (BCS) is characterized by a blockage of the hepatic venous outflow. From the hepatic venules to the confluence of the inferior vena cava (IVC) and the right atrium, the blockage can be either thrombotic or non-thrombotic.

BCS is a rare but significant syndrome because it can worsen a number of conditions, including hematologic or malignant diseases.[1]

In the global population, BCS affects 1 in 100,000 people. Acute symptoms such as hepatomegaly, ascites, and abdominal discomfort may be present in patients, as many more persistent symptoms associated with chronic portal hypertension.[2]

Blood clot in the hepatic veins are the most frequent cause of BCS. There are several causes for this including Protein C or S deficiency, Antithrombin III deficiency and conditions that are hypercoagulable (have a higher propensity to clot blood) Prior damage or damage to the blood vessels or liver, surgery that involves the abdomen or liver.[3]

The hepatic veins may get blocked by liver cancer or tumors. The hepatic veins may be compressed or invaded by tumors like hepatocellular carcinoma (HCC) or other malignancies that spread to the liver. Conditions such as inflammatory bowel disease (IBD) and systemic lupus erythematosus (SLE) might raise the risk of thrombosis and result in BCS.[4] Some persons have underdeveloped or deformed hepatic veins from birth, which can make them more vulnerable to BCS. BCS is diagnosed with the help of laboratory tests such as LFT, CBP, Coagulation profile and Imaging studies such as Ultrasound, CT or MRI.[5,6]

Treatment options for this syndrome include, Supportive Care like pain Management, Acetaminophen for mild pain, but narcotics avoided due to potential liver toxicity. Ascites Management: Diuretics (spironolactone and furosemide) and fluid restriction. Paracentesis for relief of severe ascites. Nutritional Support: A high-protein diet with adequate caloric intake.[7]

Anticoagulation is needed in some patients, especially those with underlying hematologic disorders as the cause of Budd-Chiari syndrome. Prothrombin time and activated partial thromboplastin time should be monitored once anticoagulation therapy is started and should be maintained within the therapeutic range. Also monitor ALT, AST, INR (international normalised ratio) levels throughout the therapy[8]

**CASE DESCRIPTION**

A16-year-old male patient with no prior comorbidities presented with a chief complaints of pain in the right upper quadrant and abdominal tightness which increased on intake of food, abdominal distension since 2 months which is insidious onset and gradually progressive and fever since 2 days. The patient was not associated with pedal edema, facial puffiness, yellowish discolouration of eyes and bleeding manifestations. patient has no history of diabetes mellitus, CAD, CVA, PTB. He was given a prescription of Apixaban 2.5 mg/bd for Budd-Chiari syndrome two months ago, and ten days later, he experienced vascular rash and impaired sensorium. Tab.Levetiracetam 500mg/bd and ointment soframycin was prescribed for 3 days.

There was nothing noteworthy about the patient's family history. None of his family relatives had a history of deep vein thrombosis, stroke, or liver disease.

On physical examination patient was conscious, cohorent and oriented, all the vitals were found to be normal. shifting dullness was positive. Patient has no addictions, bowel and bladder were regular.

On local examination, shape of the abdomen was found to be distended and 1mm size of multiple pecular hyperpigmented rashed were found all over the body. Dilated veins and scars were not found. laboratory findings were found that total leucocyte count is 6.30x103/mcL and platelet count was 132x103mcL which was mild thrombocytopenia no complaints of atypical cells or hemoparasites. Patients LFT reports showed an elevated AST (aminotransferace) 62 U/L, Total bilirubin 2.86 mg/dl and declined albumin levels(2.03 gm/dl), plasma Protein (4.71 mg/dl) were shown to be declined which indicates the patient was diagnosed as Chronic liver disease.

Superficial pulmonary artery Doppler test was done and results were shown altered echotexture of liver caudate lobe hypertrophy with nonvisualization of hepatic venous flow and narrow calibre of intrahepatic IVC (2.5mm) which is a sign for Budd-chiari syndrome. And Distal main portal vein, SMV and adjacent 1cm of splenic vein are thinned out - Secondary to Chronic thrombosis causing portal hypertension. Periportal collaterals seen. Proximal main portal vein and rest of the splenic vein is normal, Massive ascites, Moderate left pleural effusion and minimal right pleural effusion.

USG Abdomenresults shows there is an altered hepatic echotexture with mild surface irregularities and caudate lobe hypertrophy and non visualization of hepatic veins with IVC thrombosis (hepatic IVC): narrowed portal vein: moderate ascites with low level altered echoes.

Emergency CT abdomen showed Liver is normal in size, shape and shows diffuse altered attenuation with diffuse altered enhancement. Hepatic veins are not visualized / severely attenuated. Intra hepatic IVC calibre is severely reduced. And Intra hepatic portal is normal portal vein at head of pancreas, portal hepatis is replaced by multiple collaterals, few mesentery collaterals also seen. Proximal superior mesenteric vein is not visualized.

Ascitic fluid analysis shows colour: yellow; quantity :5ml and cell count was 80 cells.

Pathology reports showed that there is a deficiency of protein C (21%).

In order to stabilize the patient and manage their symptoms, the patient was admitted to the hospital. Initially high protein diet and low salt diet is adviced for the patient which can controls ascitic fluid accumulation. Then the patient underwent anticoagulant therapy with Apixaban 2.5mg twice per day. Inj.H.albumin 20% was administered as patient has hypoalbuminia and Tab.Ursodeoxycholic acid 300mg/od was given to enhance liver enzymes. Inj.Ceftriaxone 1gm/tid was given to treat underlying infection. By the end of the therapy patient condition was improved his ALT, AST levels were found to be normal.

**DISCUSSION**

A rare disorder called Budd-Chiari Syndrome (BCS) is brought on by blockage of the hepatic veins, which results in portal hypertension, ascites, and liver congestion[9]. External compression or primary thrombosis of the hepatic veins are the two possible reasons; thrombophilic diseases are frequently the underlying causes. This patient was diagnosed with BCS because of a protein C deficit (21%) that most likely contributed to the development of hepatic vein thrombosis.[10]

The patient's first symptoms, which included fever, distension, and tightness in the abdomen, were vague and gradually got worse over the course of two months. A vascular rash and abdominal distension with shifting dullness were among the physical findings that pointed to underlying liver pathology, which was verified by imaging and laboratory testing. BCS was supported by the results of the CT scan and Doppler ultrasonography, which revealed periportal and mesenteric collaterals, decreased caliber of the intrahepatic inferior vena cava (IVC), and non-visualization of the hepatic veins—all common indicators of portal hypertension. Chronic liver disease (CLD), which is frequently brought on by the long-term blockage of venous outflow in BCS, was indicated by the laboratory results, which included raised liver enzymes and bilirubin levels along with hypoalbuminemia.

The ascitic fluid study revealed an inflammatory condition, and the moderate thrombocytopenia could be related to splenomegaly and portal hypertension, both of which can sequester platelets. The patient's vascular rash and changed sensorium following Apixaban use further raise the risk of anticoagulant-related adverse events, which could include thrombotic or bleeding problems even in the face of anticoagulation treatment. These side effects, however, also call for close observation of anticoagulant medication and patient reaction.

In order to enhance hepatic venous outflow and stop more thrombotic episodes, anticoagulation therapy is usually used to treat BCS. An efficient direct oral anticoagulant (DOAC) that inhibits Factor Xa and can be used for long-term anticoagulation in individuals with thrombophilia, Apixaban 2.5 mg twice daily, was administered to this patient. Dietary changes (low salt, high protein diet) were part of the symptom management strategy to minimize fluid accumulation, a typical BCS consequence, and reduce ascites.

In addition, the patient was continuously watched for thromboembolic events, organ failure, and hemorrhage. Because of the patient's young age and the discovery of a protein C deficit, lifelong anticoagulation is probably required to control the risk of future thrombotic episodes and avoid recurrent thrombosis.

**CONCLUSION**

This case highlights how crucial it is to identify Budd-Chiari Syndrome in young individuals who exhibit liver impairment, ascites, and unexplained abdominal complaints. Results can be greatly enhanced by early diagnosis with imaging and laboratory testing, as well as by suitable anticoagulation and supportive care treatment. The condition of this patient further emphasizes the importance of underlying thrombophilia in the pathophysiology of BCS and the necessity of continuous anticoagulant treatment management and monitoring to avoid more problems. A multidisciplinary approach encompassing hepatology, hematology, and interventional radiology is essential for the best possible patient management because of the condition's complexity.

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