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Case Report

A Case Report on Direct Intrahepatic Portosystemic Shunt in Post-Traumatic Acute Budd-Chiari Syndrome: A Rare and Life Threatening Complication

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ABSTRACT

Budd-Chiari Syndrome (BCS) is a rare disease with an annual incidence of 0.1 to 10 per million people caused by the impaired venous outflow from the liver, primarily through the hepatic veins and inferior vena cava. BCS can be classified based on its etiology (primary or secondary), clinical course (acute, chronic, acute or chronic lesion), and morphology. Hypercoagulable conditions, myeloproliferative diseases, anatomical variability of the inferior vena cava, and environmental conditions are all etiological factors. Survival rates in treated patients range from 42 to 100%, depending on the etiology and presence of risk factors such as Child-Pugh score parameters, sodium and creatinine plasma levels, and treatment choice. Without treatment, 90% of patients die within three years, primarily from complications of liver cirrhosis. The recommended therapeutic approach for BCS is based on a stepwise algorithm that includes medical attention, interventional therapies to reinstate vessel stability (angioplasty, stenting, and local thrombolysis), Trans Jugular Porto Systemic shunt (TJPS) placement, and orthotropic liver transplantation as a last resort rescue treatment.

Keywords: Budd-chiari syndrome; Hepatic vein; Crohn's disease; Liver; Post traumatic BCS; Thrombosis

ABBREVIATIONS

ALB: Albumin; ALP: Alkaline Phosphatase; BCS: Budd Chiari Syndrome; CECT: Contrast Enhanced CT scan; CT: Computed Tomography; DIPS: Direct Intrahepatic Portosystemic Shunt; ECG: Electrocardiogram; INR: International Normalised Ratio; IVC: Inferior Vena Cava; IVUS: Intravascular Ultrasound; JVP: Jugular Venous Pulse; MRI: Magnetic Resonance Imaging; PT: Prothrombin Time; SGOT: Serum Glutamic Oxaloacetic Transaminase; SGPT: Serum Glutamate Pyruvate Transaminase; TC: Total Count; TIPS: Trans Jugular Porto Systemic shunt; TP: Total Protein; USG: Ultrasound Sonography; 2D ECHO: 2D Echocardiogram

INTRODUCTION

Budd-Chiari Syndrome (BCS) is a rare and potentially fatal disorder marked by blockage of the hepatocellular outflow tract at any level in between the junction of the inferior vena cava and the right atrium and the small hepatic veins. Geographic differences have a significant impact on the prevalence of BCS. While BCS is more common in certain Asian countries, such as Nepal, it is considered a rare disorder in Western countries. A myeloproliferative disorder is the most common underlying prothrombotic risk factor, but it is now recognized that nearly half of patients have numerous underlying prothrombotic risk factors. BCS has a wide range of clinical expressions, making it a

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possible differential diagnosis for many acute or chronic liver diseases [1]. Regardless of the cause, increased hepatic sinusoidal pressure and portal hypertension quickly follow, likely to result in venous overcrowding and ischemic damage to nearby sinusoidal hepatocytes. If therapeutic approaches or the innovation of a venous collateral framework do not relieve hepatic sinusoidal pressure, nodular regeneration, fibrosis, and eventually cirrhosis occurs [2].

A single hepatic vein obstruction is usually silent, but when two hepatic veins become occluded, venous congestion occurs, stretching the liver capsule. This can be excruciatingly painful. The sinusoids dilate, and interstitial fluid filtration occurs, resulting in liver congestion. When the filtrated fluid exceeds the lymphatic system's capacity to drain it, it begins to pass through liver capsule, resulting in ascites [3]. Portal venous hypertension and reduced blood flow to the hepatocytes via the portal vein are caused by thrombosis and venous flow interruption. As a result, the centrilobular hepatocytes suffer hypoxic injury. Acute severe cases can result in liver failure, while chronic cases can result in ascites and hepatomegaly with maintained liver function. Fibrosis develops over time, eventually leading to cirrhosis [4]. The caudate lobe is the most commonly affected lobe in Budd-Chiari syndrome because its blood is directly shunted into the inferior vena cava. As a result, when the hepatic veins become occluded, the caudate lobes hypertrophy [5].

The clinical manifestations of BCS are diverse, varying from acute liver failure to entirely asymptomatic patients. The classic triad of abdominal pain, ascites, and hepatomegaly is common among patients, with abdominal pain presenting in 61% of cases, ascites in 83%, and hepatomegaly in 67%. Fever, pedal edema, and truncal hepatic veins are other clinical features. Esophageal bleeding (5%) and hepatic encephalopathy are less common clinical expressions [6].

Depending on the type of the hepatic venous outlet blockage, BCS is further classified as primary or secondary. Secondary BCS occurs when flow is hindered by compression or intrusion of a lesion outside of the hepatic venous outlet path; examples include malignant and cystic extrinsic blockage. When flow is obstructed by an endoluminal aberration, it is categorized as primary BCS. Thrombosis seems to be the most prevalent trigger of primary BCS, though there are geographical differences, with idiopathic membranous obstructions being more common in Asia [7].

There is no single test that can be used to confirm the diagnosis of Budd-Chiari syndrome. The treatment is made on classic clinical symptoms and conditions that predispose to thrombosis, such as cancer. The initial test of option is Doppler ultrasonography, which normally helps support the diagnosis. Computed Tomography (CT) or Magnetic Resonance Imaging (MRI) may be useful if it is unavailable, ambiguous, or cannot be performed. Venography may be useful if these tests fail to establish a diagnosis but suspicion remains high.

Budd-Chiari syndrome treatment focuses on relieving the obstruction, preventing clot progression, restricting progressive liver injury, and avoiding or managing complications. The standard treatment is anticoagulants. They begin with low-molecular-weight heparin and afterwards progress to warfarin, which is taken for the rest of their lives. For warfarin patients, PT/INR should be supervised and kept within the therapeutic range. Other treatment options include thrombolysis and stenting, as well as the placement of a Trans jugular intrahepatic portosystemic shunt in acute forms of BCS that do not respond to other treatments. If all other treatments fail, surgical decompression may be considered [8]. When all other treatments fail or a patient develops decompensated cirrhosis, liver transplantation may be an option. According to published studies, the ten-year overall survival ranges from 69 to 84% [9].

CASE REPRESENTATION

A 41 year old male patient was admitted with abdominal pain for three months, abdominal distention for two months and yellowish discoloration of eyes for 3 weeks. He is a known case of hypothyroidism. The patient also has a history of blunt trauma to the abdomen prior to the onset of these complaints.

On examination it was found that deep icterus present, no pedal edema and no varicosities. JVP was observed in patient. Hepatojugular reflex was absent for the patient. Abdomen was distended with shifting dullness during abdominal examination. Dilated veins present over the anterior abdominal wall and lateral parts of flanks, flow below upwards with no back veins. Also noted firm non-tender hepatomegaly with no bruit.

Lab investigations

Blood investigation was performed and result was obtained as follows.

- Hb-12.9 gm%
- Platelet-1.0 L cells/mm³
- TC-7000 cells/mm³
- Total Bilirubin-7 mg/dL
- Direct Bilirubin-2.7 mg/dL
- SGOT-1257 units/liter of serum
- SGPT-1259 units/liter of serum
- ALP-125 IU/L
- TP-6.2 g/dL
- ALB-2.6 g/dL
- PT/INR-34.6/2.59
- Urea-19 mmol/L
- Creatinine-1.2 mg/dL
- Na-135 mEq/L
- K-3.7 mEq/L

1. Ascitic fluid- High SAAG High Protein
2. ECG was within normal limits.
3. 2 DECHO was within normal limits.

Ultrasound Sonography Test (USG) of abdomen was performed and result obtained as follows (Figure 1).

- **Liver:** Mild coarse, surface regular, poor portal vein flow present, periportal collaterals present.
- **Hepatic veins:** Echogenic particles with extension into Intravenous Cholangiogram (IVC).

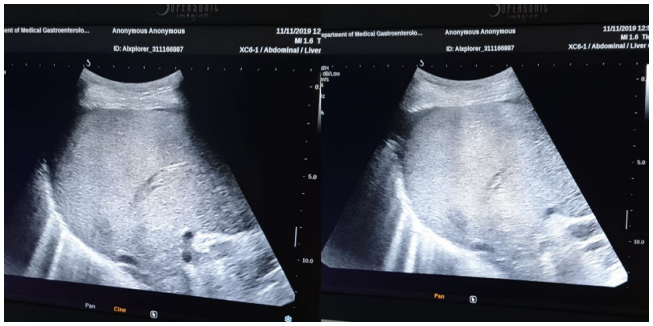

Figure 1: Ultrasound images for abdomen and liver.

CECT abdomen was done and result obtained as follows.

- Hepatomegaly with mild caudate lobe hypertrophy
- Complete thrombosis of the hepatic vein S/O Budd-Chiari syndrome
- Moderate to gross ascites

Etiological workup

- Autoimmune workup was negative
- Thrombophilia workup was negative

Diagnosis

From the above subjective and objective data diagnosis was made as follows.

- Hepatic Venous outflow tract obstruction.
- Acute Budd-Chiari syndrome mixed type predominantly involving hepatic veins.
- Ascites
- Jaundice
- Etiology-Post traumatic

Treatment

TIPS are an alternative interventional method of treating refractory Budd-Chiari Syndrome (BCS) if conservative medical management has failed. TIPS may not always be technically effective due to hepatic vein thrombosis and the inability to catheterize the hepatic veins. In these cases, Direct Intrahepatic Portosystemic Shunt (DIPS) with portal vein entry from the IVC has been a viable alternative that may alleviate portal hypertension in these patients. DIPS typically involve use of transabdominal ultrasound to reach the portal vein (Figure 2).

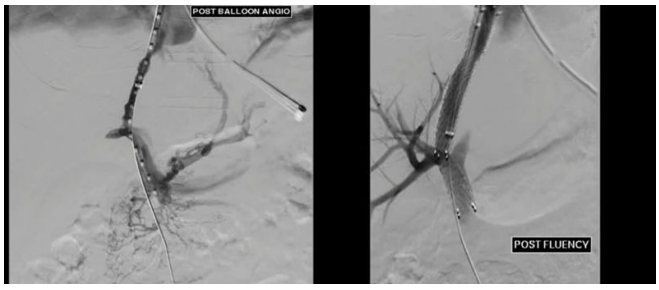

Figure 2: Marker pigtail venogram of portal vein showing the portal vein with the intraparenchymal tract. Post stent deployment showing good flow across the stent graft.

RESULTS AND DISCUSSION

IBD has been much more common over the last four decades, with around 5 million people in North America and Europe suffering [10].

BCS is a rare and potentially life-threatening condition associated with hepatic venous outflow obstruction. In the West, thrombosis is the main reason for hepatic outflow obstruction. It is currently clear that most patients have multiple underlying prothrombotic conditions, underscoring the importance of a thorough workup. Some prothrombotic factors foster a predilection for occurring in a certain area of the hepatic venous outflow tract (e.g. pure hepatic involvement versus IVC involvement.) However, when choosing a treatment strategy among a multidisciplinary team of Hepatologist, interventional radiologists and transplant surgeons, other imaging such as CT and MRI is most often necessary. All patients should be started on anticoagulation given the presence of underlying prothrombotic conditions. Still, anticoagulation is sufficient in only 18% of patients in halting the progression of liver disease, often necessitating more-invasive treatment strategies. When imaging reveals a short focal and segmental stenosis within the hepatic outflow tract, recanalization with angioplasty and stenting seems reasonable. However, most patients will have a more diffuse affliction within the hepatic outflow tract. For these patients TIPS is a suitable solution.

DIPS are the creation of a parenchymal tract between the IVC and the right branch of the portal vein to guide Intravascular Ultrasound (IVUS). However, one disadvantage of the procedure is that it necessitates the use of specialised equipment (IVUS), which is costly. Several interventionists revised this technique by percutaneously inserting a needle into a portal venous branch and then directly into the IVC using transabdominal ultrasound guidance. DIPS was chosen as a feasible choice for patients with acute and hyperacute BCS in some cases and small series.

CONCLUSION

There is an urgent need to educate patients, particularly those who have risk factors, about the likelihood of this condition. Basic education about symptoms associated with liver irregularities should be provided. If patients begin to experience symptoms, they should be recommended to seek professional advice. More importantly, they should be encouraged to follow up with their clinicians on a regular basis.

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CONFLICT OF INTEREST

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- Data collection- Eldhose Elias George, Anjali Shaju
- Data analysis and interpretation- Eldhose Elias George, Anjali Shaju, Dr Bony George
- Drafting the article- Eldhose Elias George, Anjali Shaju
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