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CASE REPORT

A Case of Idiopathic Hepatic Vena Cava- Budd Chiari Syndrome with Diagnostic Venography

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Abstract

Budd-Chiari Syndrome (BCS) is a potentially life threatening rare hepatic vascular disease that may present as acute or chronic liver failure. It has several varieties depending on etiology and level of obstruction of hepatic blood outflow. In our geographical area BCS presents differently than western countries. Idiopathic hepatic vena cava- Budd Chiari Syndrome (HCV-BCS) is more common in this area where as prothrombotic conditions are important etiological factors for classical BCS elsewhere. We present a male peasant presenting with subtle symptoms of liver disease and eventually diagnosed as having idiopathic HVC-BCS. Invasive venography was used to identify and to detect extent of obstruction.

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Introduction

Budd-Chiari Syndrome (BCS) is one of the rare hepatic vascular disorder with an estimated prevalence at one case per 100 000 individuals.[1] This hepatic venous pathology was originally described as a vascular disorder that encompasses an array of symptoms resulting from obstruction of hepatic blood outflow to the right atrium at the level of the hepatic veins or hepatic portion of the inferior vena cava.[2] According to etiology BCS can be classified in to primary and secondary. It is considered primary when hepatic outflow tract obstruction is due to an endoluminal venous lesion regardless of the cause or level of obstruction (i.e., thrombus or web).[3, 4] BCS is of secondary cause when the hepatic venous outflow tract originates from a lesion outside the venous system (tumor, abscess, cysts)".[5] BCS secondary to malignant lesions, especially hepatocellular carcinoma, is not considered as primary BCS. Classical BCS (Fig-1) presents with acute abdomen, ascites and enlarged liver. Hepatic Vena Cava Budd-Chiari syndrome (HVC- BCS) (Fig-2) presents with chronic abdominal pain and venous distension over

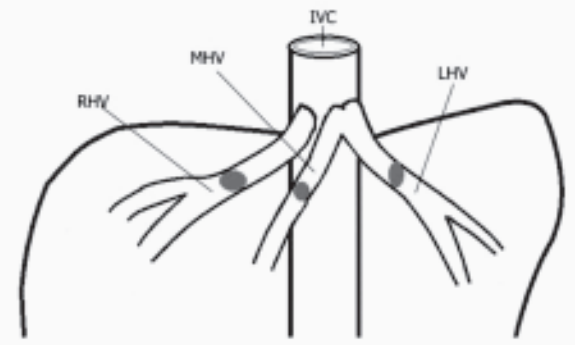

Fig-1: *Classical BCS. RHV= right hepatic vein, MHV= middle hepatic vein, LHV= left hepatic vein, IVC= inferior vena cava. Level of obstruction within the hepatic veins.*

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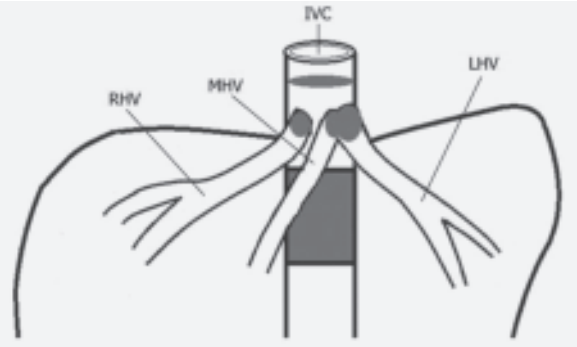

Fig-2: *Hepatic vena cava- BCS: RHV= right hepatic vein, MHV= middle hepatic vein, LHV= left hepatic vein, IVC= inferior vena cava. Level of obstruction IVC with hepatic veins.*

abdominal wall. Classical BCS is common in women with pure hepatic vein obstruction (49%-74%). HVC-BCS is more common in men with obstruction often located in both the vena cava and hepatic vein (14%-84%).[6] Pure IVC or combined IVC/HV obstruction is common is asian countries[7], whereas pure HV obstruction is frequent in Western countries. Besides at least 75% of patients with primary BCS have one or more underlying prothrombotic conditions.[8, 9] As mentioned earlier BCS is a rare disorder and only a few cases are reported from our country. We present a case report of primary HVC-BCS without any underlying prothrombotic conditions which makes it a great appeal to report. We hope it will help the physician to understand the clinical scenario with whom these patients may present, the underlying involvement of venous system and venography findings of such a case.

Case Report:

A 35-year-old male peasant from rural area presented with right sided upper abdominal pain, abdominal distension and swelling of lower limbs for 3 months. He was reasonably well before this medical condition, then he developed mild to moderate dull aching right upper abdominal pain which had no radiation, not associated with food intake. There was no aggravating or relieving factor. He added that he had noticed gradual distension of his abdomen for the same duration. He also complained about nausea with occasional vomiting along with the abdominal pain and distension. Vomitus was mixed up with altered blood for two occasions. Patient also added several episodes of black tarry stool for last 1 month. There was no other bleeding manifestation. He had also become anorexic as day passed. His condition deteriorated as he noticed heaviness of his lower limbs.

On query he had no dyspnea, chest pain , cough, palpitation, previous hospitalization, blood transfusion, jaundice in the past. He was normotensive and non diabetic. He consulted with local physician and was on oral paracetamole, oral frusemide and spirolactone. Prior to this he did not take any significant medication. None of his relatives had similar illness. Even none of his family member was suffering from diabetes, hypertension or heart disease.

Patients was heavy smoker with 15 pack-year smoking history in 20 years but he denied alcohol consumption. He belongs to lower socioeconomic class and lives in a muddy house. He drinks from tube well which is free of arsenic.

On examination he had ill looking face with abdominal distension, lower limbs swelling and visible engorged over the abdominal wall. He was moderately anemic, non-icteric. Pitting edema of lower limbs was present. Vital signs were within normal range. There was no clubbing, leukonychia, koilonychias, flapping tremor, palmer erythema. Jugular vein was non-distended. There was also no lymphadenopathy or goiter.

Abdominal examination revealed distended abdomen with inverted but transverse umbilical slit, with both flanks full. Dilated and tortuous visible superficial abdominal veins visible and they were arranged in parallel fashion & filled only from downside to upward (Fig 3). Lower margin of liver was 2 cm from costal margin along the right mid-

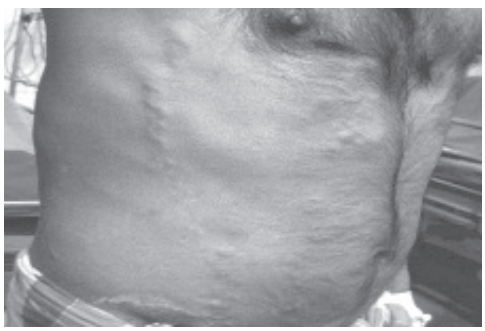

Fig-3: *Abdominal wall varices.*

clavicular line and it was tender, farm in consistency with regular margin & smooth surface. There was ascites as evident by positive shifting dullness. Spleen and kidney were not palpable or ballotable. Bowel sound was present.

Cardiovascular, respiratory system and neurological examinations revealed normal findings.

A complete blood count disclosed hemoglobin 5.5 g/dl (normal: 12-14 g/dl) with total platelet count 85 x 10^9 /L (normal: 150 – 400 x 10^9 /L) but white cell count within normal range. Peripheral blood film showed microcytic hypochromic anemia. Hemoglobin electrophoresis revealed 98% hemoglobin A and 2 % hemoglobin A2. Fasting blood sugar was normal.

Liver function test disclosed serum bilirubin 2 mg/dl (normal <3 mg/dl), serum alanine transaminase, aspartate transaminase were minimally elevated. Serum alkaline phosphatase was 195 u/L (normal: 30-150 u/L) whereas serum albumin was low (23 g/L; normal: 35-50 g/L). Prothombin time was revealed to be 15.3 seconds with international normalization ration (INR) 1.28 (control 12 seconds). Viral markers were investigated and revealed negative anti- hepatitis C antibody and hepatitis B virus surface antigen were negative.

Renal function test disclosed initial serum creatinine 2.4 mg/dl with subsequent return to baseline. Serum urea and serum electrolytes were within normal range.

Ultrasonography of whole abdomen showed hepatomegaly with coarse parenchyma, enlarged coarse spleen, dilated portal vein and ascites. A doppler study of portal vein, inferior vena cava (IVC) and hepatic veins revealed moderate portal hypertension, thrombus within the lumen of IVC, portalization of IVC & hepatic veins.

Subsequently c-ANCA, p-ANCA and antinuclear antibody (ANA) were done and found to be negative as was non reactive VDRL. A thrombophilia screen was unrevealing as were bleeding time and clotting time.

So patient was managed conservatively with anticoagulant & diuretics; and invasive hepatic venography by right femoral vein puncture was planned to confirm the diagnosis as well as to see the extent of obstruction in IVC. (Fig 4 & 5)

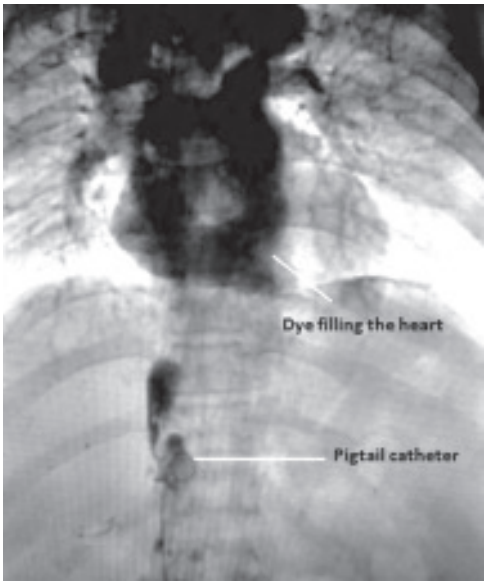

Fig-4: *Inferior venacavography.*

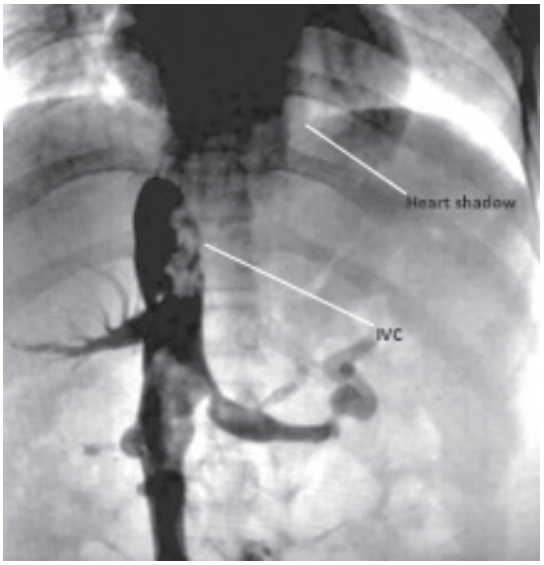

Fig-5: *Inferior vanacavography showing obstruction between IVC and heart shadow.*

Discussion

We presented a case of Budd-Chiari Syndrome where IVC is involved and no prothombotic etiology can be found. An underlying thrombotic condition can be detected in more than 80% of patients with isolated BCS.[10] Budd-Chiari Syndrome was first described by Budd[11] in 1845 and later by Hans Chiari in 1899. Initially they described the clinical

scenario as a rare vascular disorder resulting from obstruction of hepatic blood outflow at the level of hepatic venins or hepatic portion of the IVC.[2] With the advancement of diagnostic and therapeutic techniques providers have expanded upon these initial characterization. It is challenging to identify the precise location of the obstruction which clinically and prognostically significant. In 2003, Valla[1] proposed clinical manifestation depends on level of obstruction.

In 1998, Okuda et al[12] stated that primary hepatic venous thrombosis (Classical BCS) and thrombosis at the level of IVC were two separate syndromes. Classical BCS appears to be more common in western population and usually has a known etiology (Fig-5).[5] On the contrary HVC-BCS (Fig-6) appears to be more common in East Asian patient population and is more often idiopathic or due to membranous obstruction; and it commonly presents with a chronic onset of less severe symptoms. The clinical scenario of our case is consistent with the above mentioned findings. Interestingly, the location, size and chronicity is also clinically important as it directs therapeutic approach for patient management.[13]

A diagnosis of BCS should be considered in all patients presenting with acute or chronic liver disease especially when common causes for liver disease have ruled out. Imaging test plays an important role in early diagnosis of BCS and assessment of location of obstruction.[14-16] Routine ultrasonography of whole abdomen, Doppler ultrasound, MRI & CT scan are commonly done investigations to see the patency of hepatic venous system. Doppler Ultrasound has over 85% sensitivity and should be first choice of imaging investigation.[17,18]

Though majority of population with primary BCS have prothrombotic conditions, patients presenting with HCV involvement frequently have negative thrombophilia scan. This finding is again true for our case. It was said that measurement of protein C, protein S or antithrombin concentration should be regularly performed in BCS patients and their first degree relatives.[4] But recent data suggest that same recommendation may not be appropriate for HVC-BCS patients.[5]

55%-76% of reported populations of classical BCS are female where as HVC-BCS is more common in men (51%-66%) and more likely to present with an IVC obstruction with or without involvement of hepatic vein (69%-100%).[5] Our male patient had both hepatic vein and IVC involvement.

According to Shin N et al (2016), classical BCS usually presents within 6 months & 60-85% of the patients have acute presentation. But the definition of chronic vs. acute were not explicitly delineated. In comparison, HVC-BCS typically presents with chronic symptoms with an average duration of symptoms prior to diagnosis ranging from 44-96 months.[19] Spleenomegaly, abdominal wall varices, lower extremity varices and discoloration are more commonly associated with HVC-BCS.[20] Out patient had abdominal wall varices and lower limb edema. (Fig-3)

Data from Budd-Chiari Syndrome review by Shin N et al also supports the possibility of two different types of BCS with separate etiologies: classical BCS and HVC-BCS. Classical BCS patients have increased thrombophilic risk factors than patients with HVC-BCS, where as idiopathic hepatic venous outflow obstruction is more common. On the contrary MTHFR C677T mutations are commonly found in HVC-BCS in comparison to classical BCS.[5]

Historically it has been speculated that there might be some association between standard of living and HVC-BCS. A recent prospective study from western India found no association between socioeconomic status and location of hepatic venous outflow obstruction. Although a correlation between living in mud houses and IVC membranous obstruction was observed.[21] Our patient is also from a low socioeconomic status and has been living in mud house.

Non-invasive imaging tests are at most causes sufficient to diagnose BCS. However if they are inadequate, invasive venography and liver biopsy should be further considered. Venography is useful for accurate assessment of extension and location of outflow obstruction and measurement of Hepatic Venous pressure.[22] In our case plain ultrasound & Doppler ultrasound cannot locate the level of obstruction definitely, so patient had undergone inferior venacavography with right femoral vein approach under fluoroscopic guidance which showed an obstruction at IVC. (Fig-4 & 5)

Conclusion:

In conclusion, Budd-Chiari Syndrome is a life threatening condition that should be kept in mind while investigating any patient with acute or chronic liver disease without any obvious etiology. In contrast to thrombophilic conditions which is more common in western, in our geographic area a different mechanism of hepatic venous outflow obstruction may play the role. A thorough investigation to find out obscure thrombophilic pathology may not be a routine measure for patients presenting with BCS in our locality. Also it may relieve some economic burden these poor country people may weigh by the disease itself.

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