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Budd Chiari Syndrome- A Rare Case Report

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Abstract

The Budd–Chiari syndrome (BCS) is a rare life threatening and clinically challenging disorder defined by obstruction of hepatic venous outflow anywhere from the small hepatic veins to the suprahepatic inferior vena cava. It is considered as a manifestation of underlying prothrombotic risk factors of which myeloproliferative disorders are the most common ones. We describe a 41year old Male who presented with bilateral lower limb edema and abdominal distension with visible veins over the anterior abdomen which on further evaluation was diagnosed as Budd-chiari syndrome. Budd Chiari syndrome requires a prompt and accurate diagnosis with an aggressive treatment which depends on the cause, clinical presentation and anatomical location of the problem.

Keywords: *Budd-chiari syndrome , prothrombotic state , Thrombophlebitis , Myelofibrosis.*

Introduction

Budd-chiari syndrome is a rare condition characterized by obstruction of hepatic venous outflow tract with an occurrence in 1 in 100,000 of the population worldwide[1,2].The term Budd-chiari was coined after the work of George budd and Hans Chiari who described 3 cases of hepatic vein thrombosis in 1845 & pathological description of obliterating endophlebitis in hepatic veins in 1899 respectively[1] Budd-chiari syndrome is classified into primary[1,3] when the obstruction is related to a primary venous problem and is considered secondary when it is related to an extrinsic cause like compression due to abscess, tumor & cyst[2,4,5,6]. Clinical presentation of Budd-chiari syndrome may vary from completely asymptomatic condition to fulminant liver failure. Hence it is important for an early diagnosis so as to provide an appropriate management at the right time.

Case Report

41 year old male patient presented with complaints of bilateral lower limb swelling since 4 years associated with abdominal distension and visible veins over the anterior abdomen since 8 months. There was exacerbation of lower limb swelling for the past 8 months & abdominal

distension for the past 6 months. He had noticed occurrence of veins over the left side of abdomen which is insidious in onset and gradually progressive to involve the entire abdomen. There was no history of vomiting, hematemesis, constipation, malena. There was no history of any altered sensorium, breathlessness, chestpain or palpitations. Patient had no comorbidities.

General Examination of the patient was normal & systemic examination of cardiovascular, neurological and respiratory systems showed no abnormalities. Abdomen examination revealed dilated veins over the anterior abdomen involving para umbulical region and bilateral flanks. The abdomen was distended with the umbilicus in the midline and all quadrants moving with respiration

The liver was palpable below the right costal margin and blood flow in the dilated veins is from below- upwards .The liver span was 12cms and shifting dullness present with no fluid thrill.

Routine investigations were done which were normal .Ultra-sound abdomen showed liver size of 15.5cm with coarse parenchymal echotexture and dilated hepatic veins, the intra hepatic portal vein appears to be reduced in caliber and inferior vena cava was dilated with no occlusion. Portal venous Doppler showed Hepatic veins and inferior vena cava appears dilated and Intra-hepatic portal vein appears reduced in caliber and increased periportal echogenicity which may represent periportal Edema or fibrosis or congestive heart failure. 2D echo was done which is normal. CECT- ABDOMEN was done which showed mild luminal narrowing in the supra hepatic Inferior vena cava at the confluence of hepatic veins with small spec of calcification within and multiple dilated tortuous collaterals in anterior abdominal wall (features of chronic Inferior vena cava thrombosis). Protein C and S, serum-homocysteine levels were done which are within normal limits. ANA and JAK-2 mutation were negative. So patient was diagnosed to have Budd-chiari syndrome.

Discussion

The most common causes for Budd-Chiari syndrome include inherited and acquired coaguable states. The inherited causes include V leiden mutation, protein C and S deficiency, anti thrombin 3 deficiency and prothrombin G20210A mutation which result in hepatic vein thrombosis and further pericipitating to Budd-Chiari syndrome. The acquired causes include myeloproliferative disorders like polycythemia vera, paroxysmal nocturnal hemoglobinuria, essential thrombocytosis and myelofibrosis[4,7,18]. Other conditions which have been reported as risk factors for the development of Budd-Chiari syndrome include anti-phospholipid syndrome, hypereosinophilic syndrome, behcet disease and ulcerative colitis[4].The pathophysiology of Budd Chiari syndrome includes occlusion of hepatic veins leading to venous outflow compromise in the liver which ultimately results in increase in sinusoidal and portal pressure leading to hepatic congestion and ascites. Hepatocytes undergo ischemic damage that eventually leads into non inflammatory centrilobular cell necrosis. If the damage is massive the patient will present with fulminant form of Budd Chiari syndrome. The patient with Budd Chiari syndrome can present as fulminant, acute, subacute and chronic state.[9] Fulminant type develops in a few days and patient presents with severe hepatic failure with elevation of enzymes, hyperbilurubinemia, encephalopathy and coagulopathy[10]. The liver is massively enlarged and painful with ascites[8,11,12]. The acute variant develops within 1 month and is characterized by acites, abdominal pain hepatomegaly, elevation of liver enzymes, renal failure and coagulopathy[11]. The subacute form is the most common type and usually insidious in onset and asymptomatic with no ascites.[8] The chronic type of Budd Chiari syndrome is characterized by development of portal hypertension with ascites. The hepatic enzymes can be normal or minimally elevated.[11] The general clinical manifestions encountered in Budd Chiari syndrome include abdominal pain and

distension, ascites, liver failure, lower limb edema gastrointestinal bleeding and encephalopathy.[4,9] Clinicallly Budd Chiari syndrome should be suspected in patients who present with any of the following : fulminant liver failure with acute onset ascites and hepatomegaly, massive ascites with preserved liver function, unexplained chronic liver disease and an associated thrombogenic disorder[1]. Imaging studies like conventional ultrasound, Doppler ultrasound, CT, MRI and catheter venography are helpful in the diagnosis of Budd Chiari syndrome. As in our case Ultra-sound abdomen showed liver size of 15.5cm with coarse parenchymal echotexture and dilated hepatic veins, the intra hepatic portal vein appears to be reduced in caliber and inferior vena cava was dilated with no occlusion. Portal venous Doppler showed Hepatic veins and inferior vena cava appears dilated and Intra-hepatic portal vein appears reduced in caliber and increased periportal echogenicity which may represent periportal Edema or fibrosis or congestive heart failure. 2D echo was done which is normal. CECT- ABDOMEN was done which showed mild luminal narrowing in the supra hepatic Inferior vena cava at the confluence of hepatic veins with small spec of calcification within and multiple dilated tortuous collaterals in anterior abdominal wall (features of chronic Inferior vena cava thrombosis).

Medical Management of Budd Chiari syndrome involves treating the underlying cause: Portal hypertension, ascites, anticoagulation and thrombolysis.[1,9,13]. Surgical management involves membrane resection, IVC reconstruction, portosystemic shunts, portoarterial shunts and liver transplantation[14-18] Endovascular management includes balloon angioplasty, stunt placement, catheter directed thrombolysis and Trans jugular intrahepatic portosystemic shunt. Our patient was managed with IVC – angioplasty with stenting & was done. Clinically patients symptoms of abdominal distension and bilateral lower limb edema decreased and abdominal veins disappeared .Patient was started on inj - Fragmin (Dalteparin) 5000iu s/c od &Tab - Clopilet 75 mg od. Patient was discharged with Tablet. Dabigatran 110mg p/o bd.

Conclusion

Budd Chiari syndrome is an unusual clinical condition which requires an accurate and prompt diagnosis and an aggressive treatment. Probably patients of Budd Chiari syndrome are best managed in a tertiary care hospital where liver transplantion could be done in necessary situations.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient has given his consent for his images and other clinical information to be reported in the journal. The patient understands that his names and initials will not be published and due efforts will be made to conceal his identity, but anonymity cannot be guaranteed.

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