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Case report

Budd-Chiari Syndrome: What is the Cause?

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Introduction

Budd-Chiari syndrome (BCS) is a rare condition. It results from occlusion of hepatic venous outflow. Abdominal pain, ascites, and liver enlargement are classic triad symptoms in BCS. Myeloproliferative disorders, thrombophilias, malignancy, and infections are common etiological factors for BCS.[1]It can be due to thrombotic or nonthrombotic obstruction of hepatic venous outflow. It was first described by Budd in 1845 and subsequently by Chiari in 1899.[2,3] Site of hepatic outflow obstruction in BCS is variable. It is frequently noted at level of inferior vena cava (IVC) and hepatic veins. Untreated patients with BCS have poor prognosis. Death occurs due to progressive liver failure in three months to three years after the diagnosis is made.[4] With shunt procedures and liver transplantation, 5 year survival is up to 87%.[5,6,7] Here we describe a patient with BCS, who was diagnosed at our Medical Unit. Etiology of BCS could not be made. Patient was managed conservatively as he deferred interventional radiological procedure Diagnosis of BCS was made on CT scan and subsequent Doppler ultrasonography

Case Report

A 24 years old young man presented with progressive abdominal distension and shortness of breath for 3 months. Before onset of these symptoms he had been treated for upper abdominal pain and fever that lasted for 10 days. He also complained of dry cough, anorexia, fatigue, and undocumented weight loss. At admission he was pale,had tachycardia (pulse rate 106/minute), and tachypnea (respiratory rate 30/minutes). Pedal edema up to ankles and lymphadenopathy (bilateral cervical and axillary) were noted. Axillary lymph nodes were of 2-3 cm in size and were located in central and apical area. Cervical lymph nodes were 1.5x1cm in size, and located in anterior cervical and supraclavicular location. Lymph nodes were soft to firm, discrete, and mobile. Most were well circumscribed with few matted nodes as well. Skin overlying lymph nodes was normal. Abdominal examination showed tender hepatomegaly (span 16 cm) and moderate ascites. Bilateral mild pleural effusions were noted on chest examination. Rest of clinical examination was unremarkable. All routine investigations were within normal limits (Table 1)Pleural and ascitic fluid examination were suggestive of transudate (Table 2) Chest radiograph showed bilateral pleural effusion (Figure 1). Ultrasonography showed hepatomegaly, ascites, abdominal lymphadenopathy, bilateral pleural effusion. Echocardiography was unremarkable .CT scans of chest, abdomen, and pelvis were done that showed inhomogeneous enlarged, mottled liver with delayed post contrast enhancement of liver periphery and around hepatic veins. The caudate lobe was enlarged (caudate lobe hypertrophy) and showed increased contrast enhancement compared with the remainder of the liver. Hepatic veins were not visualized (thrombosed). Marked abdomino-pelvic ascites, splenomegaly, collateral formation in anterolateral chest wall, bilateral cervical, mediastinal, axillary and abdominal lymphadenopathy were also noted (Figure 2).Doppler ultrasound also showed similar findings. Upper gastrointestinal endoscopy showed grade esophageal varices.Biopsy of axillary and mediastinal lymph node showed chronic non-specific lymphadenitis. Bone marrow examination revealed reactive changes. Pleural biopsy showed chronic inflammatory changes. No definite granuloma or feature suggestive of malignancy were noted. Diagnosis of idiopathic Budd-Chiari was made. Patient was treated with broad spectrum antibiotics, analgesics, diuretics, and injectable steroids, in addition to anticoagulation. Anti tuberculous therapy was also initiated. He deferred interventional radiology shunt procedure despite counseling and got discharged on request. No remarkable improvement was noted on follow up after 1 month.Despite repetitive telephonic requests he has not come for follow up.

Table I. Hematological and biochemical investigations

Investigation	Result	Investigation	Result
Hemoglobin	8.6 gm%	Total bilirubin	0.6 mg%
MCV	61.4 fL	Alanine transferase (ALT)	40iu/L
Peripheral film	Unremarkable	Alkaline phosphatase	109iu/L
WBCs	$8.3\times10^9/mm^3$	Sreum Albumin	3.2 gm/dl
Platelets	$369\times10^9/mm^3$	Prothrombin time (PT)	WNL*
ESR	20 mm at 1 hour	Activated partial thromboplastin time (APTT)	WNL*
Urea	24 mg%	Serum Potassium	3.8mm/L
Creatinine	0.9 mg%	Serum Sodium	138mm/L
Urine R/E	Unremarkable	Serum Lactic dehydrogenase (LDH)	558 iu/L
ANA	Negative	Hepatitis B, C and HIV serology	Negative
Anti-phospholipid antibody	Negative	Protein C and S	Normal
RA factor	Negative	Serum Ferritin	26.73 ng/mL
Blood and bone marrow cultures	No growth	Brucella serology	Negative

*WNL; within normal limits

Table 2. Ascitic and pleural fluid analysis

Ascitic fluid		Pleural fluid	
Test	Result	Test	Result
RBC count	1550cells/µl	RBC count	1050cells/µl
Total leukocyte count	80cells/µl	Total leukocyte count	130 cells/µl
Polymorphs	15%	Polymorphs %	10%
Lymphocytes	85%	Lymphocytes %	90%
Proteins	0.9 g%	Proteins	1.3 g%
Adenosine deaminase (ADA)	12 u/l	Adenosine deaminase(ADA)	10 u/l

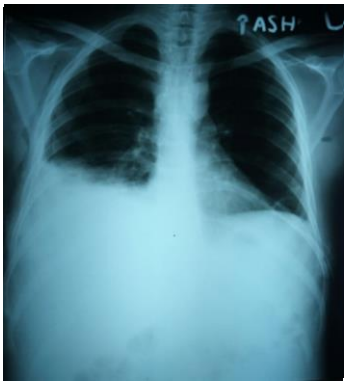

Figure 1. Chest X-Ray showing bilateral pleural effusion

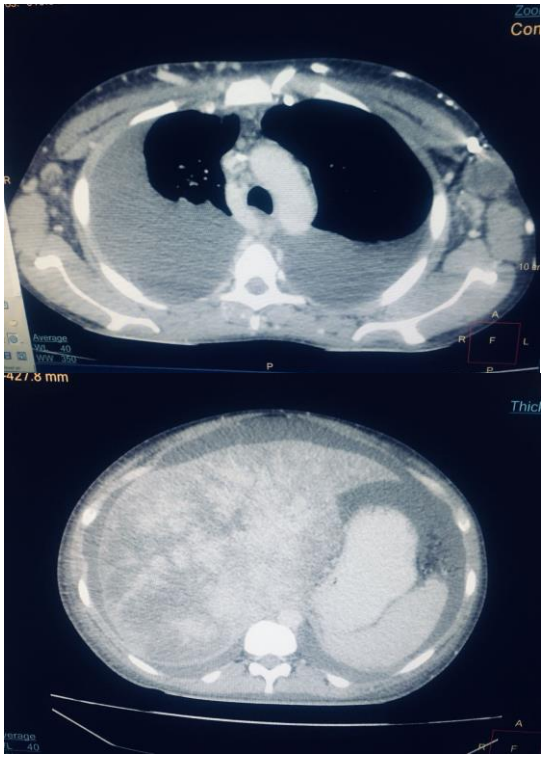

Figure 2: CT Scan showing mottled liver (arrow) and caudate lobe hypertrophy, ascites, and bilateral pleural effusion

Discussion

Gender and age wise differences have been noted in BCS patients belonging to Asian and non-Asian countries. Asian BCS patients are predominantly male and have 45 years median age. Female predominance and thirties to forties year age group involvement in non-Asian BCS patients has been noted.[8]Our patient was young and male. BCS can be classified based on

site of venous obstruction and clinically. Hepatic veins, and inferior venae cavae alone or combined may be obstructed in BCS due to thrombosis, external compression or webs. Four clinical types of BCS are; 1) acute, 2) subacute, 3) chronic, and 4) fulminant.[9,10]In acute type that occurs within weeks, patients have painful enlargement of liver. Ascites and upper gastrointestinal bleeding due to development of varices may occur. Moderate derangement of liver enzymes is also noted. In subacute type, patients present with progressive ascites that may be accompanied by swelling of feet developing in months. These patients have minimal derangement of liver enzyme. Our patient had sub acute type of BCS. Patients with chronic BCS have features of decompensated cirrhosis. Patients with fulminant type have jaundice, coagulopathy and marked derangement of liver enzymes. Hepatic encephalopathy develops in these patients within 2 months of jaundice onset.

Ascitic fluid protein concenteration in BCS patients is generally >2 g% and gradient of serum to ascites albumin is <1.1.[11]Values may be different in acute type of BCS. Ultrasongraphy has 85-90% sensitivity and specificity for diagnosing BCS when performed by triplex method.[12] CT scan, magnetic resonance imaging, hepatic venography, and liver biopsy are further helpful for making diagnosis. Interestingly we were thinking of tuberculosis and neoplastic in our patient as at presentation he had lymphadenopathy, ascites, and pleural effusion. His initial ultrasound also showed hepatomegaly, ascites, and abdominal lymphadenopathy. Situation changed when we noted that ascites and pleural fluid analysis were transudative. CT scan findings pointed to the diagnosis of BCS.

Etiological list of BCS is exhaustive. Most patients with BCS have underlying thrombotic disorders. In up to 33% patients etiological diagnosis cannot be made and the condition is termed idiopathic. Myeloproliferative disorders, malignancy, infections, pregnancy, and contraceptive use cause 90% of BCS.[13] Our effort in this regard remained fruitless, although we started the patient on anti-tuberculous therapy empirically.

Treatment of BCS patients can be divided in to four types; 1) treatment of the etiology, 2) conservative, 3) surgical, and 4) interventional radiological. Conservative treatment includes; managing ascites and portal hypertension, anticoagulant therapy, and use of thrombolytic agents according to clinical scenario. Surgical options include various shunt procedures and liver transplantation. Transjugular intrahepatic portosystemic shunt (TIPS), angioplasty, and stenting are interventional radiological options.

Prognosis in BCS is variable. Patients with unknown etiology die in 3-36 months after diagnosis is made. Three year survival in these patients is 10%.[4]38-87%, five year survival has been noted in patients who are treated with various shunting procedures. After liver transplant five year survival is 70%.[5,6] Outcome in our patient is expectedly poor as etiological diagnosis was not made and surgical or interventional radiological options were deferred by the patient.

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