



## Case Report

## Case of Budd-Chiari syndrome, an enigma as abdominal tuberculosis in a tubercular endemic country: A case report

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## ABSTRACT

**Introduction and importance:** Budd-Chiari syndrome is an uncommon disease due to obstruction of hepatic venous outflow. Clinical manifestations range from asymptomatic cases to those requiring liver transplants. The study highlights the importance of diagnosing a case of Budd-Chiari syndrome which has been suspected with abdominal tuberculosis where anti-tubercular drugs may themselves damage the liver.

**Case presentation:** Herein we report a case of 18 years old female presenting with upper abdominal pain along with recurrent abdominal distention, jaundice, and deranged liver function. Also, adenosine deaminase level was raised in both pleural and peritoneal fluids, hence, anti-tubercular treatment was started but could not be continued as she developed adverse reactions to these drugs. CT scan later revealed features suggestive of Budd-Chiari syndrome. Initially, she was managed with balloon angioplasty, but her condition worsened ultimately requiring a liver transplant.

**Clinical discussion:** Budd Chiari syndrome can present with subtle presentation and since abdominal tuberculosis is very non-specific, the two conditions can be very confusing, particularly in the tubercular endemic region. Detailed clinical assessment along with proper investigations and imaging should be performed for early recognition as both conditions are associated with high morbidity and mortality if not treated timely.

**Conclusion:** The necessity of careful investigation and consideration of Budd-Chiari syndrome as an important cause of ascites with jaundice and deranged liver function in TB endemic regions along with early anticipation of liver transplant is necessary, as in this case.

## 1. Introduction

Budd-Chiari syndrome (BCS) is an infrequent disease condition due to obstruction of the hepatic venous outflow tract, categorized as intrinsic factors such as thrombosis or phlebitis and extrinsic compression by cysts, and tumors, or abscess [1]. The etiology differs with the regions in the world, with the west in favor of prothrombotic conditions such as factor V Leiden mutation, prothrombin G20210a mutation, protein C/S deficiency, antithrombin III deficiency, myeloproliferative disorders, myelofibrosis, Behcet's disease, antiphospholipid syndrome, oral contraceptives, etc. Whereas membranous obstruction of the vena cava (MOVC) or primary inferior vena cava (IVC) thrombosis in the eastern region [2–4]. Classical BCS presents with symptoms of

abdominal pain, ascites, and hepatomegaly [5]. Imaging modality includes ultrasonography, computed tomography, magnetic resonance imaging, and venography [5]. The management of BCS includes treatment of underlying cause along with anticoagulants, subsequently thrombolysis if clots are present, and diuretics as medical therapy; if persistent then further treatment options include angioplasty or stenting of the vein and/or transjugular intrahepatic portosystemic shunt (TIPS) [6–9]. Liver transplantation is the ultimate treatment in case of fulminant acute liver failure and when the conventional treatment fails [8–10].

Here we present to you a case of an 18-year-old female with acute BCS, who was initially started on empiric anti-tubercular drugs due to suspected tuberculosis. The study highlights the importance of

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diagnosing a case of Budd-Chiari syndrome which has been suspected with abdominal tuberculosis where anti-tubercular drugs may themselves damage the liver.

This case report has been reported as per SCARE 2020 criteria [11].

## 2. Case presentation

18-years-old female, non-smoker, non-alcoholic, and with no history of chronic illness presented with complaints of pain over epigastric and right hypochondrium for 10 days. The non-radiating pain developed acutely, on and off affecting her daily activities. Also, she noticed gradually progressive abdominal distention. Her bowel and bladder habits are normal.

On examination, she was averagely built with stable vital signs. Further, icterus was noted in bilateral conjunctiva along with bilateral lower limb pitting edema extending up to knees. On abdominal examination, it was distended with a centrally placed inverted umbilicus. Tenderness was noted prominently over the right hypochondriac region. Shifting dullness could be elicited. Abdominal organs could not be palpated. A bowel sound was heard. No abnormality was detected on examination of the respiratory, cardiovascular, and central nervous systems.

Investigations revealed normal blood cells count. Her PT was 79.10 with an INR of 5.65 and aPTT of 93.2 seconds. The total protein level was 8.60 with a serum albumin level of 3.70. As for the liver function test, her total bilirubin level was 3mg/dl and her unconjugated bilirubin level was 1.90mg/dl. The renal function test was normal. Ascitic fluid was serosanguinous with SAAG of 1.6 and adenosine deaminase (ADA) level of 4.8U/L. Pleural fluid was transudative with an ADA level of 8. Serum LDH level was 285U/L and CA-125 level was 528U/ml. Serology testing for hepatitis A, Hepatitis B and C, syphilis, and EBV was non-reactive. Ultrasonography (USG) of the abdomen and pelvis revealed thickened gall bladder wall with moderate fluid in the peritoneal cavity. It also revealed bilateral pleural effusion, more on the right side.

The patient was admitted to the medical ward and symptomatic management for pain and abdominal distention was done along with antibiotics. An initial CT scan of the abdomen done on March 16, 2020, showed ascites, mesenteric and aortocaval lymphadenopathy with bilateral pleural effusion as shown in Fig. 1. Colonoscopy was done a week later which showed benign sigmoid polyp with normal rest of the colon. As ascitic and pleural fluid tapping both revealed raised ADA levels and being a tuberculosis endemic country, anti-tubercular therapy (ATT) was started but was stopped after 2 weeks as she developed ATT-

induced hepatitis. She had repeated hospitalizations with multiple USG-guided ascitic fluid tapping done for symptomatic relief. Repeat CT scan done on April 27, 2020 which could not visualize the hepatic veins with a mottled enhancement of liver suggesting towards Budd-Chiari Syndrome as shown in Fig. 2. Thus, from clinical features, investigations, and radiological viewpoint, a final diagnosis of Chronic Budd-Chiari disease with chronic liver disease with decompensation was made.

Subsequently, hepatic, portal, and inferior vena cava doppler USG was done which showed normal color flow and spectral pattern in the hepatic artery, the biphasic flow pattern in the hepatic vein, and normal color flow and spectral pattern along with biphasic flow pattern in inferior vena cava. There was coarse echotexture of the liver and hypochoic lesion in segment VIII of the liver along with the mild perihepatic collection. She was even transferred to ICU as she developed increased drowsiness. On April 30, 2020, she underwent MHV (Middle hepatic vein) balloon angioplasty which significantly improved her ascites. Again, on May 6, 2020 transcatheter thrombolysis with streptokinase was performed. Repeat venography on May 8, 2020 showed leaks of contrast into the peritoneum which was then coil-embolized. The patient was started on warfarin but was discontinued due to high INR.

Even though initially she was asymptomatic, her condition deteriorated subsequently. Her Child-Pugh score was 10 and her MELD score was 18 hence, was advised for liver transplantation for unresolving ascites, spontaneous bacterial peritonitis, and hepatic encephalopathy. She had undergone a Living Donor Liver Transplant (LDLT) of the right lobe with a modified middle hepatic vein graft on July 3, 2020 in India. Findings of diseased liver included: enlarged congested liver, ascites (10L), atretic, thrombotic hepatic veins, and conventional portal vein anatomy. Postoperatively, she was shifted to ICU. On the second post-operative day, her liver enzymes were raised with narrowing and biphasic IVC flow on Doppler suggestive of impaired outflow. Therefore, IVC stenting was done which was uneventful. Following the procedure, she was started on anticoagulation. Follow-up Doppler showed improved liver outflow with an improvement in liver enzymes. Subsequently, she was shifted to the ward and after a series of normal liver enzymes, she was discharged with stable vitals.

After hospital discharge, she is under immunosuppression, antibiotics, antifungals, multivitamins, Insulin, steroid, aspirin (for 3 months), and nicoumalone alternate days to maintain an INR of 2–3. Her post-transplant hepatic Doppler done 6 months following the transplant showed normal hepatic veins and portal vein with normal flow. Currently, she is doing well and under tacrolimus, mycophenolate, and nicoumalone only. She is also advised for blood workup (LFT, PT/INR, Tacrolimus level) 2 monthly and follow up 6 monthly with all



Fig. 1. Axial CT scan of the abdomen showing the presence of ascites and pleural effusion.



Fig. 2. Axial CT scan of abdomen showing mottled appearance of liver.

investigations (Chest Xray, Blood profile, Doppler, etc.).

### 3. Discussion

Budd-Chiari syndrome (BCS) being a rare disease itself, it is also associated with high comorbidity and even mortality if not treated timely [12,13]. BCS only consist of 1.3% of the total patient undergoing a liver transplant [7]. Most common associations with BCS in our eastern region were found to be in alcoholics with poor nutrition and bacterial infection [2]. Nepal is one of the countries with a high burden of tuberculosis (TB), where the incidence is more than 100 cases per 100,000 population [14]. Due to the high TB prevalence, abdominal tuberculosis is one of the common differentials with consideration of cirrhosis, congestive heart failure, infectious hepatitis, neoplasm, etc. With the above-mentioned presentation of the case along with elevated ADA levels in the analysis of pleural and ascitic fluids, no clinical and laboratory evidence of other disease conditions lead this case's provisional diagnosis to be highly likely to be abdominal tuberculosis. The symptoms of abdominal TB are very non-specific [15] and also in an early stage of BCS. Acute BCS might present with only subtle changes in imaging in the early stage of disease, for it lacks the development of the collateral [16]. Detailed history, clinical assessment, and multi-modality imaging with ultrasonography followed by computed tomography or magnetic resonance imaging and venography along with liver biopsy in case of uncertainty with high suspicion should be considered even in a country like ours with high TB endemicity, to rule out BCS. This can help in early recognition and prompt management of the condition reducing the morbidity and complication of the disease. A similar case was reported by Fardeen Baray et al. [17] in Afghanistan where the importance of inclusion of BCS as a differential in patients presented with ascites and jaundice even in resource-limited settings.

Regarding the step-wise approach to the management of BCS, we would like to point out the need to consider liver transplant early in the course of disease who present with severe liver function derangements such as ours with elevated bilirubin and elevated prothrombin time [18, 19].

### 4. Conclusion

Budd-Chiari syndrome should be considered as a differential among patients from endemic TB regions presenting with ascites, jaundice, and deranged liver function. Extensive investigations with imaging and even biopsy for prompt recognition with early management of the condition are required. A liver transplant should be considered in patients with severe liver dysfunction.

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None.

### Authors contributions

Author 1: Concept of study, case data collection, revising, and editing the manuscript, Author 2: Concept of study, literature review, revising, and editing the rough draft into final manuscript, Author 3: Contributed in writing the manuscript draft, revising, and editing the manuscript, Author 4: Case data collection, literature review, revising and editing the manuscript, Author 5: Concept of study, literature review, revising and editing the manuscript, Author 6: Literature review, revising and editing the manuscript, Author 7: Literature review, revising and editing the manuscript, All authors were involved in manuscript drafting and revising, and approved the final version.

### Registration of research studies

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### Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

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### Declaration of competing interest

None.

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### Appendix A. Supplementary data

Supplementary data to this article can be found online at <https://doi.org/10.1016/j.amsu.2022.104607>.

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