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Case Report

Misdiagnosis of Budd Chiari syndrome, a case report from Afghanistan

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ABSTRACT

Introduction: Budd-Chiari syndrome is a rare disease characterized by hepatic venous flow obstruction. The obstruction may be thrombotic or non-thrombotic anywhere along the venous course from the hepatic venules to the inferior vena cava (IVC) junction to the right atrium. In clinical practice, cases can be misdiagnosed, particularly in regions where resources are limited, unless the clinician pays special attention to such diagnosis.
Case report and clinical discussion: Here, we would like to present a misdiagnosed case of Budd Chiari syndrome. This reported case is a case of 30 years old female patient complaining of dull abdominal pain and swelling. Initially, the patient consulted a local health facility where the patient was diagnosed with tuberculous peritonitis and subsequently treated with an anti-TB regimen empirically. Within a few days of taking medicine, she developed mild jaundice and lower limb edema. At this stage, the patient came to us, which after taking history, her physical examination unveiled mild jaundice, ascites, abdominal tenderness, and mild lower limb petting edema. The patient was recommended an abdominal CT scan with contrast, which revealed early enhancement and enlargement of the caudate lobe and non-opacification of hepatic veins with narrowing of the hepatic part of the inferior vena cava consistent with Budd-Chiari syndrome. The patient was started on warfarin and referred for a hepatic decongestive procedure. After four months of performing a transjugular portosystemic shunt, the patient came to us for follow-up. She had an excellent clinical improvement and was started on rivaroxaban 20 mg daily orally.
Conclusion: The main takeaway lesson of this particular case is to consider the differential diagnosis of ascites from an etiologic point of view and not to overemphasize a single etiology.

1. Introduction

First described in 1845, Budd Chiari syndrome is an uncommon and rare disease characterized by hepatic venous flow obstruction. The obstruction may be thrombotic or non-thrombotic anywhere along the venous course from the hepatic venules to the junction of the IVC to the right atrium [1,2]. The obstruction caused by thrombosis is called primary BCS, and obstruction caused by extrinsic compression is called secondary BCS [1,3]. Its prevalence and annual incidence are estimated to be 1 per million and 11 per million, respectively [4].

Here we would like to present the case of a young lady with the primary diagnosis of tuberculous peritonitis that was prescribed an anti TB regimen ex-juvantibusly. Subsequently, the patient developed jaundice within 1–2 days of starting the anti-TB regimen. At this stage, the patient came to us which after taking a thorough history, performing a physical examination, and an abdominal CT scan was diagnosed with Budd Chiari syndrome.

This case report has been reported in line with the SCARE Criteria [5].

2. Presentation of case

We report a case of 30 years old female patient who presented to the medical ward complaining of dull and ongoing abdominal pain and swelling for about three weeks. The issue first came to attention in a rural health facility where the case was misdiagnosed with TB peritonitis based on fundamental lab exams like the predominant lymphocyte count in peritoneal fluid and an elevated ESR. The patient was subsequently treated empirically with an anti TB regimen consisting of isoniazid, rifampicin, pyrazinamide, and ethambutol. After taking a few

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doses of the anti TB medicine, she also started complaining of loss of appetite, mild nausea-vomiting, mild jaundice, and mild lower limb edema, which developed later. The patient did not have a notable history of any disease. She was single, did not take contraceptive pills or hormone therapy, and did not smoke or drink alcohol. Moreover, the patient did not have any history of abdominal surgery, and all family members were healthy.

In general appearance, she looked restless and mildly icteric. Her vital signs were as follows: blood pressure 120/80 mmHg, heart rate equaled pulse rate and was 95 beats per minute, respiratory rate was 16 cycles per minute, and the temperature was 37° centigrade. The physical findings of the abdomen were as follows: in inspection, her abdominal contour was protuberant; in auscultation, normal bowel sounds were present; in palpation, she had generalized abdominal tenderness, especially at right upper quadrant; and in percussion, she had shifting dullness suggestive of ascites. In the local exam, mild jaundice was notable in her sclera, and mild +1 pitting edema was present in both lower limbs. Physical examination of other systems was unremarkable.

Her background hematologic laboratory results were as follows: hemoglobin level 11g/dl, leukocyte count 9000/μL(Neutrophil 69%, Lymphocyte 24%, Monocyte 4%, Eosinophil 2%, Basophil 1%) and platelet count 160000/μL. The patient's liver function test showed an elevated level of ALT (170 iu/L) and bilirubin (total 3 mg/dl, direct 1.1 mg/dl, and indirect 1.9 mg/dl), and a decreased level of serum albumin (2.3 g/dl). On further investigation, the viral hepatitis serologic tests were all negative. Moreover, the patient's INR was 1.9.

The ultrasound examination of the patient could not visualize hepatic veins and reported the features of chronic liver disease. The contrast-enhanced computed tomography revealed the early enhancement and enlargement of the caudate lobe with the inhomogeneous mottled appearance of the liver. It also showed the non-opacification of hepatic veins with narrowing of the hepatic part of the inferior vena cava. The above findings were associated with ascites, mild splenomegaly, and diffuse edema of the abdominal wall (Figs. 1-3).

Based on the clinical and imaging findings, she was diagnosed with Budd-Chiari syndrome. Subsequently, besides supportive therapy, including diuretics, the patient was started on low molecular weight heparin of 1mg/kg subcutaneously twice daily plus warfarin 2.5 mg daily. After three days of anticoagulant therapy, her subsequent INR was

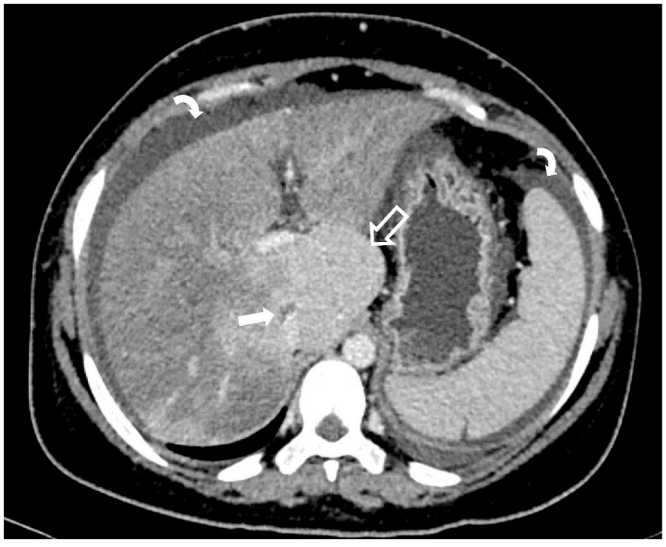

Fig. 1. Contrast-enhanced abdominal CT, axial section through the upper abdomen: Early enhancement and enlargement of the caudate lobe (open arrow) is noted with the inhomogeneous mottled appearance of the liver the so-called nutmeg liver. Mild splenomegaly is also appreciated. Marked narrowing of the hepatic part of the inferior vena cava is noted (white arrow).Free fluid is seen surrounding the liver and the spleen.

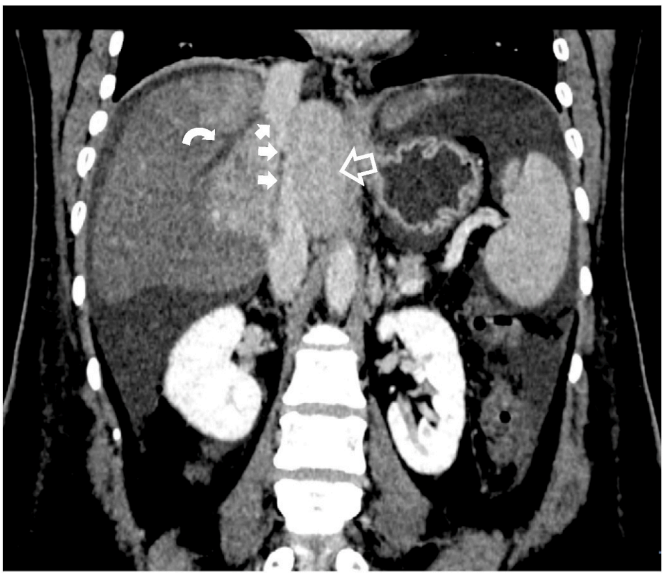

Fig. 2. Contrast-enhanced abdominal CT, coronal section through the upper abdomen: hypertrophied caudate lobe (open arrow) is well-enhanced compared to rest of the liver parenchyma. Marked narrowing of the hepatic part of the inferior vena cava is noted (white arrows). Non-opacification of the imaged right hepatic vein (curved arrow).

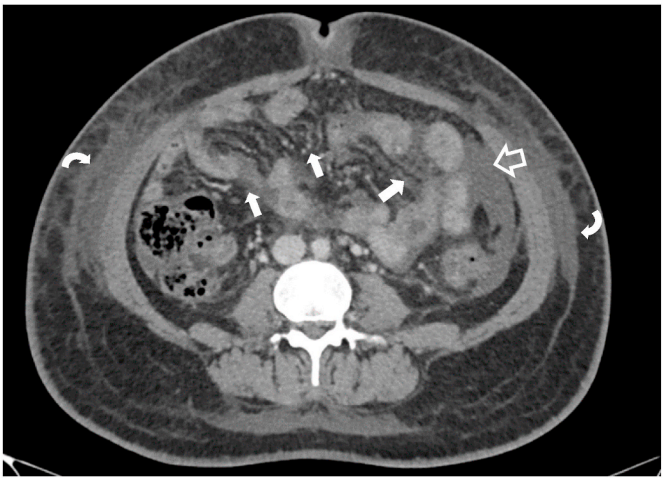

Fig. 3. Contrast-enhanced abdominal CT, axial section through the lower abdomen: Free fluid is seen in the peritoneal cavity (open arrow) with features or mesenteric congestion (white arrows) and diffuse edema of the abdominal wall (curved arrows).

2. The patient demonstrated some clinical improvement. Despite this, the patient underwent the hepatic decongestive procedure called transjugular portosystemic shunt (TIPS).

The patient came to us for a follow-up after four months. She had excellent clinical improvement and was started on rivaroxaban 20 mg daily. The follow-up lab exams were as follows: Total bilirubin 2.5 mg/dl (indirect bilirubin 1.7 mg/dl and direct albumin 0.8 mg/dl), ALT 42 IU/L, alkaline phosphatase 122 IU/L, serum albumin 2.7 g/dl, and INR 1.2. The Doppler ultrasound showed the shunt to be patent.

3. Discussion

Based on the relevant clinical history and physical examinations, our provisional diagnoses were TB peritonitis, drug-induced hepatitis, viral hepatitis complicated to liver cirrhosis, and hepatic vein thrombosis.

Because tuberculosis in all forms is a prevalent infectious disease in Asian countries [6], most extrapulmonary tuberculosis cases, especially in Afghanistan, are treated empirically based on clinical and basic laboratory findings due to economic constraints. That is why this case was misdiagnosed with TB peritonitis based on the clinical presentation of ascites and a predominant lymphocyte count in peritoneal fluid and subsequently treated with an anti-TB regimen. According to the patient's history, she received the medicine for 2–3 days which did not work well and instead increased her symptoms and developed jaundice. The patient came to us with this clinical picture, and we decided to admit her for further workup and investigation. By reviewing the literature, the time interval from the onset of anti-tuberculosis therapy till the detection of hepatotoxicity ranged between 7 and 90 days, and the average time was 24 days [7]. That is why we doubted the provisional diagnosis of drug-induced hepatitis. The viral hepatitis scenario has been ruled out by reporting negative results for virology serological tests. So, the hepatic vein thrombosis was strongly considered the culprit pathology, which was later confirmed by an abdominal CT scan.

Although the patient did not have other clinical manifestations characteristic of COVID 19, we suspected the SARS-CoV-2 virus as a possible etiology for the hepatic vein thrombosis. In the medical literature, there were accumulative reports of venous and arterial thrombosis including hepatic vein thrombosis due to SARS-CoV-2 virus infection [8,9]. However, SARS-CoV-2 virus infection could not be confirmed by PCR because when the patient came to us, we were facing the peak of the COVID 19 wave, and PCR was only available for confirming severe cases of COVID 19. The patient could not afford to perform extra lab exams to clarify the possible other underlying etiologies.

This article underlines the importance of differential diagnosis of ascites accompanied by jaundice and the appropriate usage of costly imaging in resource-limited regions where patients do not have access to health insurance.

4. Conclusion

The main takeaway lesson of this particular case report is to consider the differential diagnosis of ascites from an etiologic point of view and to bear in mind Budd Chiari syndrome in such cases, especially when ascites is associated with jaundice and abdominal pain. In addition, it is necessary to check the liver function tests before any hepatotoxic drug prescription to avoid any future misconceptions.

Ethical approval

This case report was approved by the ethical committee of Kabul University of Medical Science.

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Author contribution

Fardeen Baray contributed to study design, paper drafting and revision, and the final approval of the paper to be submitted.

Mohammad Behroz Noori contributed to study design, paper drafting and revision, and the final approval of the paper to be submitted.

Mohammad Maroof Aram contributed to study design, paper revision, and final approval of the paper to be submitted.

Hidayatullah Hamidi contributed to study design, paper revision, providing images with the captions, and final approval of the paper to be submitted.

Provenance and peer review

Not commissioned, externally peer-reviewed.

Patient 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 upon request.

Research registration

N/A.

Guarantor

Fardeen Baray, MD.

Declaration of competing interest

The authors have declared no conflicts of interest.

Acknowledgment

Nothing to declare.

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