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CASE REPORT

Budd-Chiari Syndrome: An Unexpected Diagnosis in a Clinical Practice in Somalia

Síndrome de Budd-Chiari: Un diagnóstico inesperado en la práctica clínica en Somalia

Ahmed Muhammad Bashir[1] , Mohamed Abdulkadir Hassan-Kadle[2,3] , Ahmed Adam Osman[4] , Ahmed Adam Mohamud[5]

1. Department of Internal Medicine, Mogadishu Somalia-Turkiye Training and Research Hospital, Mogadishu, Somalia.
2. Center for Digestive and Liver Disease, SomGastro Clinic, Mogadishu, Somalia.
3. College of Medicine & Health Science, Abrar University, Mogadishu, Somalia.
4. Department of Radiology, Mogadishu Somalia-Turkiye Training and Research Hospital, Mogadishu, Somalia.
5. Faculty of Medicine and Surgery, Salaam University, Mogadishu, Somalia

Corresponding author
Ahmed Muhammad Bashir
E-mail: ambashir@hotmail.com

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Abstract

Although hepatic vascular diseases are relatively uncommon in clinical practice and require accurate diagnosis and treatment, Budd-Chiari syndrome (BCS) is a group of disorders described by hepatic venous outflow obstruction, which can be thrombotic or non-thrombotic. It occurs at any level of hepatic veins or hepatic portion of the inferior vena cava (IVC) to the right atrium as defined by the European Association for the Study of the Liver (EASL) guidelines. We report a 55-year-old male patient with a history of diabetes mellitus who developed a Budd-Chiari Syndrome that has the classical triad of BCS (Abdominal pain, Ascites and hepatomegaly). The investigations showed an alteration of liver function tests and imaging characteristics of sub-acute Budd-Chiari syndrome. We could identity the causal factors of BCS in this case. Although many cases have been reported in the literature, none is from Somalia or east African countries. The diagnosis could be reached with abdominal ultrasound, but because of limited clinical experience in diagnosis imaging, many cases may be misdiagnosed. We recommend our physician and radiologist to keep this diagnosis in their mind and diagnose, so the patients could be managed properly.

Key words: Budd-Chiari Syndrome, Abdominal Pain, Ascites, Hepatomegaly.

Resumen

Aunque las enfermedades vasculares hepáticas son relativamente infrecuentes en la práctica clínica y requieren un diagnóstico y tratamiento precisos, el síndrome de Budd-Chiari (SCB) es un grupo de trastornos descritos por la obstrucción del flujo de salida venoso hepático, que puede ser trombótico o no trombótico. Se produce a cualquier nivel de las venas hepáticas o de la porción hepática de la vena cava inferior (VCI) hasta la aurícula derecha, según la definición de las directrices de la Asociación Europea para el Estudio del Hígado (EASL). Informamos de un paciente varón de 55 años con antecedentes de diabetes mellitus que desarrolló un síndrome de Budd-Chiari que presenta la tríada clásica del SCB (dolor abdominal, ascitis y hepatomegalia). Las investigaciones mostraron una alteración de las pruebas de función hepática y características de imagen del síndrome de Budd-Chiari subagudo. Pudimos identificar los factores causales del SCB en este caso. Aunque se han descrito muchos casos en la literatura, ninguno procede de Somalia o de países del este de África. Se pudo llegar al diagnóstico con la ecografía abdominal, pero debido a la limitada experiencia clínica en el diagnóstico por imagen, muchos casos pueden ser diagnosticados erróneamente. Recomendamos a nuestros médicos y radiólogos que tengan presente este diagnóstico y lo diagnostiquen, para que los pacientes puedan ser tratados adecuadamente.

Palabras clave: Síndrome de Budd-Chiari, Dolor abdominal, Ascitis, Hepatomegalia.

Introduction

A British internist George Budd first described Budd-Chiari Syndrome (BCS) in 1845, and reported three cases of hepatic vein thrombosis due to abscess-induced phlebitis, and then an Austrian pathologist Hans Chiari expanded it by presenting 13 cases and described the first pathological features of liver with "obliterating endophlebitis" of the hepatic veins[1].

BCS is an uncommon and rare vascular disorder characterised by hepatic venous outflow obstruction, which can be either thrombotic or non-thrombotic and it occurs at any level hepatic veins or hepatic portion of the inferior vena cava (IVC) to the right atrium as defined EASL guidelines[2]. This excludes the absence of right sided heart failure, pericarditis, or sinusoidal obstruction

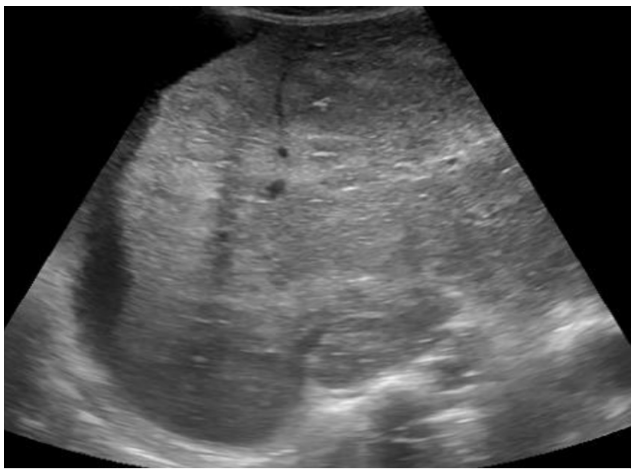
syndrome[3]. The BCS can lead to acute liver failure, liver cirrhosis, and even hepatocellular carcinoma[4].

The BCS can be classified according to aetiological factors into primary and secondary. Primary BCS is a flow obstruction due to an endoluminal lesion or primary venous process, such as thrombosis or phlebitis. This primary BCS obstruction can range from the small hepatic veins to the orifice of the IVC into the right atrium. The most common causative factors of BCS that have been identified are haematologic or prothrombotic state that is approximately estimated around 75% of patients, so haematological disorders that are associated to BCS include; polycythaemia Vera, essential thrombocythemia and myelofibrosis. Prothrombotic conditions that associated to BCS include; paroxysmal nocturnal haemoglobinuria, antiphospholipid syndrome and inherited deficiencies of protein C, protein S, and antithrombin III. Secondary BCS is when flow is obstructed due to compression or invasion of lesion outside the hepatic venous outflow tract such as benign or malignant diseases (e.g. abscesses, hepatocellular carcinomas, and renal cell carcinomas, or secondary to cardiac or pericardial diseases) are more linked to BCS while less common causes that associated with BCS are inflammatory bowel disease, aspergillosis and Behcet's syndrome[4,5].

BCS is diagnosed clinically and confirmed by investigation. Abnormal liver function tests are a trademark, however they can also be normal. Serum aminotransferase levels may be five times normal in acute and fulminant BCS. Radiological imaging confirms a BCS diagnosis. Hepatomegaly, limited liver vein visibility, and compressed IVC are all shown on abdominal ultrasonography. Intrahepatic collaterals, splenomegaly, and ascites indicate chronic BCS[5].

The BCS management focuses on controlling of portal hypertension and ascites with systemic anticoagulation to prevent progressive extension of the venous thrombosis[6].

Figure 1A: Liver Ultrasonography revealed enlarged liver with heterogeneous parenchyma and no definite focal lesion. Hepatic veins were not visualized. Marked ascites noted perihepatic area.



We present a case of BCS in an adult male patient who came to internal medicine clinic with abdominal pain and distension.

Case

A 55-year-old male patient from Mogadishu, Somalia, presented to us with a complaint of a two-month history of gradually progressive abdominal distension. He is a known case of diabetes mellitus and takes metformin 850mg tablet twice a day and Gliclazide 60mg tablet once per day. The abdominal distension is associated with nausea and loss of appetite. Complete blood count revealed haemoglobin 15g/dL, leukocytes 8.3x1000mm^3, and platelets 429×1000/mm^3. Liver biochemistry showed a total bilirubin of 1.65mg/dL, direct bilirubin 0.89mg/dl, aspartate transaminase 76 IU/L, alanine transaminase 83 IU/L, alkaline phosphatase 249 IU/L, gamma glutamyl transferase 381 IU/L and albumin 5 g/DL. The International normalised ratio (INR) was 1.25. Ascitic tap showed Serum Ascites Albumin Gradient (SAAG) >1.1.

Liver sonography revealed enlarged liver with heterogeneous parenchyma and no definite focal lesion. Hepatic veins were not visualized. Marked ascites were noted perihepatic area (**Figure 1A**). Further imaging with abdominal CT without (**Figure 1B**) and with contrast (**Figure 2**) Showed a Mottled appearance of peripheral hypodensity in the liver parenchyma. Hepatic veins do not opacify. Mildly enlarged Caudate lobes with normal enhancement. Portal veins opacity normally (**Figure 3**). Upper gastrointestinal endoscopy showed small oesophageal and gastric varices (**Figure 4**). Therefore, based on the clinical and imaging finding, the patient was diagnosed with subacute form Budd-Chiari syndrome. Treatment with diuretics and anticoagulant was initiated.

Figure 1B: Unenhanced Axial CT scan shows slightly heterogeneous liver parenchymal appearance, enlarged caudate lobe with Massive Ascites.

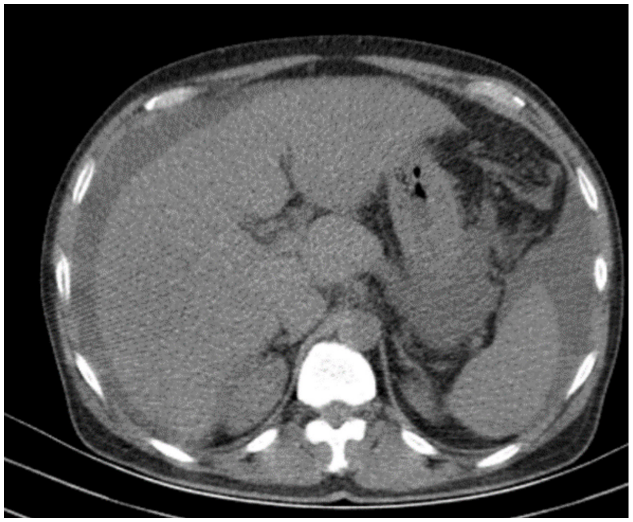


Figure 2: Contrast-enhanced Axial CT scan Shows Mottled appearance peripheral hypodensity in the liver parenchyma(yellow arrow). Hepatic veins do not opacity (blue arrow). Mildly enlarged caudate lobes with normal enhancement (red arrow). Portal veins opacity normally (thick black arrow head).

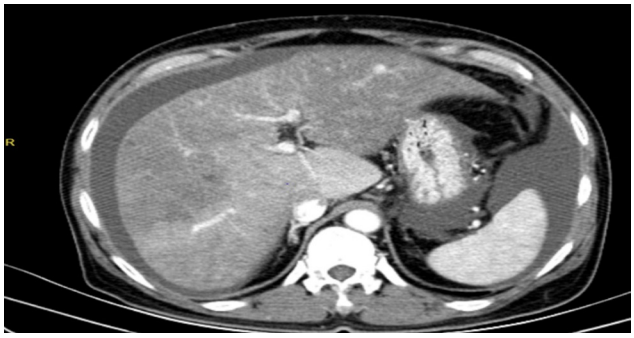


Figure 3: Contrast-enhanced Axial and Coronal CT scans showed no enhancement branch of hepatic veins draining İVC (Red arrow) corresponding to occlusion due to thrombosis (Black arrow).

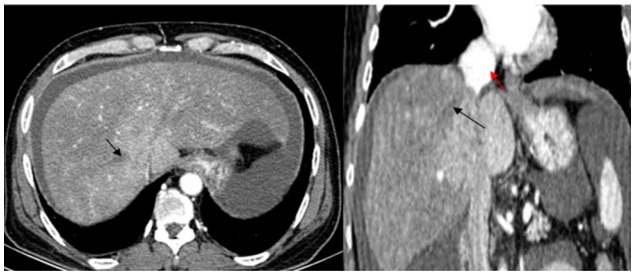
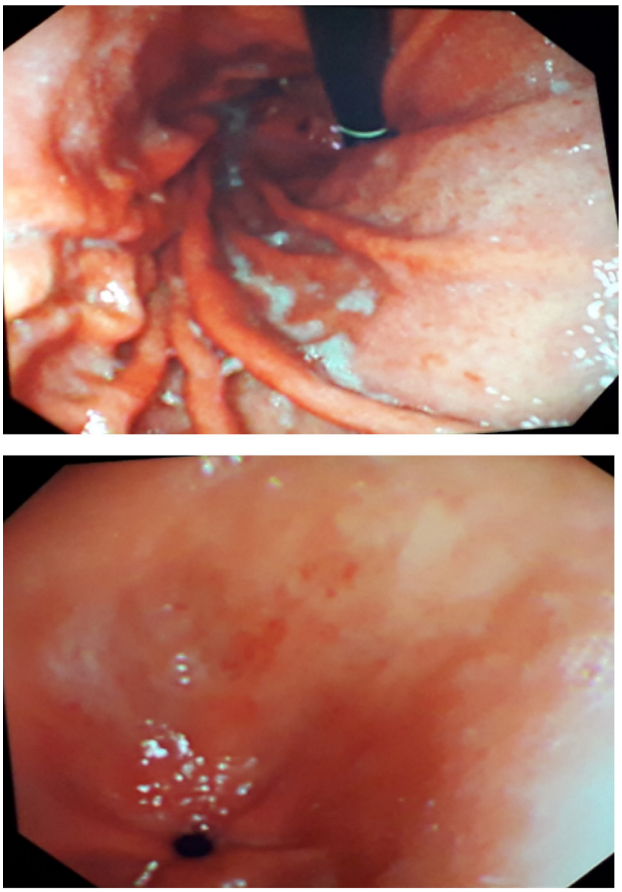
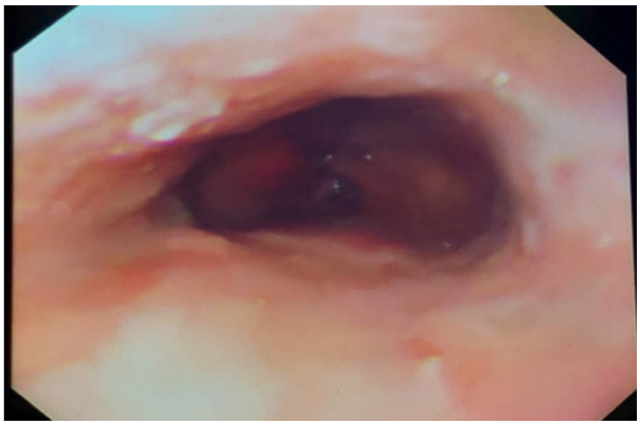
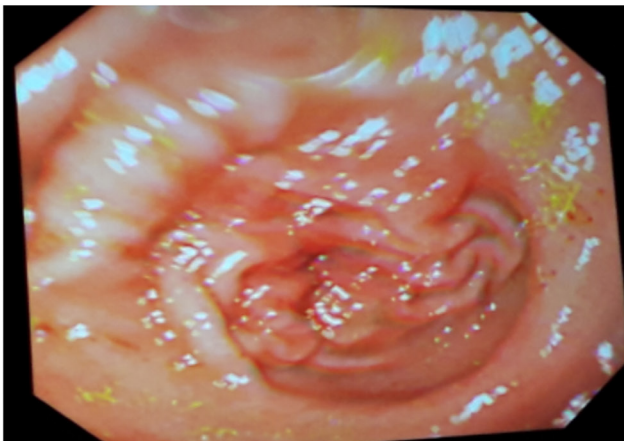


Figure 4: Upper gastrointestinal endoscopy showed small oesophageal and gastric varices.







Discussion

Budd-Chiari syndrome is a rare disease of the liver, with its nature is heterogenous that characterised by occlusion of hepatic venous[1]. Reporting such as this case in Somalia is difficult in our context due to misdiagnosis or a low index of suspicion. BCS is usually diagnosed as clinically and radiologically. We report the first case in the country that had abdominal pain, ascites and with the presence of hepatomegaly with imaging studies revealed the presence hepatic venous thrombosis so the diagnosis was most suggestive BCS.

Clinically, the BCS most often appears with a triad features that are abdominal pain, Hepatomegaly and ascites, while laboratory analysis of BCS are included liver function test such transaminase, bilirubin and alkaline phosphatase, Gama glutamyl transferase, albumin, coagulation test and platelet count. The ultrasound is first useful choice investigation while the Computer tomography and magnetic resonance imaging studies are the second choice of the investigation, so the combination of these imaging modalities, clinical information and laboratory

should suffice to diagnosis of BCS in the clinical practices[6]. Portal veins opacity normally with exclusion of the presence of a tumour, abscess or a cyst that originated outside the veins that may result of obstruction to the hepatic outflow via compression or invasion which is secondary BCS and the finding of gastrointestinal endoscopy that showed small oesophageal varices in this would suggest that our patient had a primary type BCS of a subacute form.

The Treatment of BCS can be divided into medical, radiological and surgical procedures; the goals of treatment are to prevent thrombus propagation, restore the flow in clogged veins, decongest the liver, and treat and prevent complications related to fluid retention, malnutrition and portal hypertension[6].

Conclusion

BCS is a rare disorder in our clinical practice that requires accurate diagnosis and immediate therapy. Diagnosing such a rare case requires high suspicion index and highly skilled radiologists, especially in low resource countries like Somalia, where communicable disease dominates. Early detection of symptoms helps in early diagnosis and management of the disease.

Ethical Approval

In our institution, Ethical approval is waived from case reports.

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.

Provenance and peer review

Not commissioned, externally peer-reviewed

Conflict of Interest

Authors declare no conflict of interest.

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None

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