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Budd-Chiari Syndrome: A Rare Complication of Hepatic Hydatid Cyst

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Abstract **Case Report**

Budd-Chiari syndrome is a rare disease characterized by a defect in hepatic venous drainage. It can be primary, occurring due to venous thrombosis, or secondary, caused by compression or invasion of the hepatic drainage veins. Symptoms can be acute, chronic, or even asymptomatic. Diagnosis is based on imaging tests that show hepatic vein obstruction, collateral circulation, and morphological changes in the liver. Treatment aims to identify and treat the underlying disease.We report a case of a patient with Budd-Chiari syndrome secondary to a liver hydatid cyst to highlight the radiological aspects of this pathology.

Keywords: Budd-Chiari Syndrome, hepatic hydatid cyst, hepatic venous drainage.

INTRODUCTION

Budd-Chiari Syndrome is a rare disease characterized by a defect in hepatic venous drainage, which can be partial or complete. This entity can be primary or secondary and presents with different symptoms. In this article, we report a case of a patient with secondary Budd-Chiari Syndrome complicating a hepatic hydatid cyst.

CASE PRESENTATION

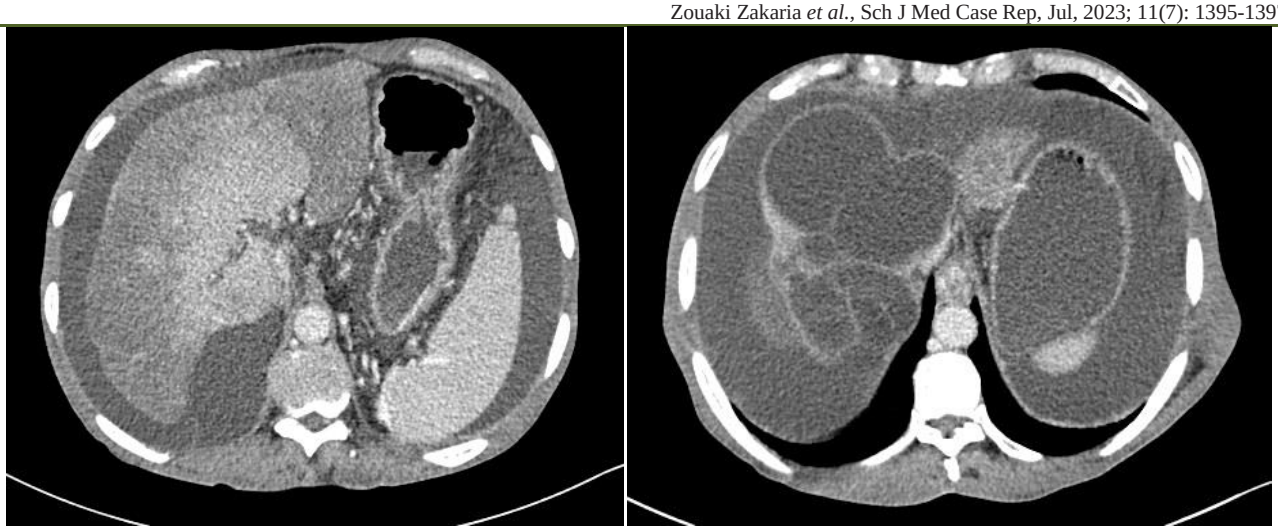
The patient was a 75-year-old man from a rural area with a history of contact with dogs. He presented with two episodes of medium-volume hematemesis without melena, associated with large-volume ascites and chronic stabbing pain in the right hypochondrium of moderate intensity, with intermittent evolution over six months, and intermittent cholestatic jaundice. The patient had no transit disorder but presented with asthenia, unquantified weight loss, and a feeling of fever.

Clinical examination revealed cutaneous-mucous pallor with stable hemodynamic status, signs of hepatic insufficiency and portal hypertension, as well as sensitivity in the right hypochondrium without Murphy's sign or palpable mass.

The test results showed iron-deficiency anemia at 10.2 g/dl, thrombocytopenia at 90,000/mm3, white blood cells at 4490/mm3 with normal renal function, and a prothrombin level of 60%. Moderate cytolysis and cholestasis were also detected.

Esophagogastroduodenoscopy showed grade III esophageal varices with type 1 gastric varices with red signs, which required hemostatic elastic band ligation without incident.

Abdominal ultrasound and abdominal computed tomography revealed a large multivesicular hydatid cyst of the hepatic dome (Figure 1) compressing the inferior vena cava and the suprahepatic veins, as well as a dysmorphic liver with nodular contours, with a lack of peripheral liver enhancement after contrast injection compared to the center. This was associated with abundant ascites.



a **b**

Figure 1: CT scan after portal contrast injection. Large multivesicular hydatid cyst of the hepatic dome compressing the suprahepatic veins and inferior vena cava (a) with a dysmorphic liver with nodular contours, peripheral atrophic liver enhancement defect compared to the center, and abundant ascites (b)

Hydatid serology was positive, while viral serologies for hepatitis B and C were negative.

Unfortunately, the patient could not undergo surgery for the hydatid cyst, and the outcome was unfavorable. He died from complications related to hepatic insufficiency.

DISCUSSION

Budd-Chiari syndrome is a rare disease [1] characterized by obstruction of hepatic venous return, which can occur at various levels ranging from hepatic venules to the terminal portion of the inferior vena cava [2]. This obstruction can be primary in the case of thrombosis, or secondary when it is due to extrinsic compression or invasion. In our case, Budd-Chiari syndrome is secondary to hepatic venous compression caused by a liver hydatid cyst, which is a parasitic disease resulting from the intrahepatic development of Echinococcusgranulosus.

The clinical manifestations of Budd-Chiari syndrome are variable. In acute forms, there is right hypochondrium pain, hepatomegaly, and ascites. In chronic forms, abdominal pain is more moderate and associated with ascites. In some cases, the disease can be asymptomatic and discovered incidentally [3].

Imaging plays a crucial role in the diagnosis of Budd-Chiari syndrome. Hepatic venous obstruction is the main visible sign on Doppler ultrasound in the majority of patients, whether it is due to thrombosis of the main hepatic veins or the inferior vena cava in primary forms, or to compression or invasion of these veins in secondary forms. This obstruction leads to the development of collateral circulation between a permeable portion of the hepatic vein and a normal vein [4]. These diversion routes can be intrahepatic or extrahepatic, and their transverse arrangement, unlike the radial aspect of usual hepatic vascularization, is very characteristic [5]. The visualization of these supplementary routes is considered a sensitive sign [6]. Hepatic venous insufficiency leads to perfusion abnormalities and morphological changes in the liver. In acute forms, hepatomegaly and less enhanced hepatic parenchyma are observed due to congestion. In chronic forms, the classical appearance is that of "mosaic" enhancement after CT or MRI injection, reflecting sinusoidal distension. Dysmorphism will appear and is characterized by atrophy of poorly drained territories, often peripheral, in favor of central, better-drained territories. Thus, segment I, which benefits from independent venous drainage directly connected to the inferior vena cava, hypertrophies very significantly in nearly half of the cases, and its drainage veins, usually virtual, are often dilated, with a diameter greater than 3 mm being in favor of the diagnosis [4]. Portal pressure is increased and leads to portal hypertension with splenomegaly, ascites, and esophagogastric diversion routes. Nodules develop in chronic forms of the syndrome. They correspond to regeneration nodules, a consequence of the decrease in portal vascularization and compensatory hyperarterialization.

Phlebography is reserved for patients for whom endovascular treatment is indicated. It allows the detection of occlusion or stenosis of one or more hepatic veins, as well as visualization of a collateral venous network. It also allows visualization of the state of the inferior vena cava and measurement of pressure [5].

Liver biopsy is indicated only if the diagnosis is not established by imaging or if thrombosis is localized to small hepatic veins [7].

Without treatment, symptomatic forms of Budd-Chiari syndrome have an unfavorable outcome. Treatment is based on the recognition and treatment of the underlying disease, prevention of venous thrombosis, and restoration of venous drainage.

CONCLUSION

Budd-Chiari syndrome is a rare disease that can complicate liver hydatid cysts, especially when they are large in size and located at the hepatic dome. Studying the relationship between the cyst and the hepatic veins and inferior vena cava using imaging techniques allows for an early diagnosis of the syndrome even at an asymptomatic stage, thereby improving the prognosis.

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