Case Report

Budd-Chiari syndrome due to giant hydatid cyst: a case report and brief literature review

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Abstract

Budd-Chiari syndrome is an uncommon disorder characterized by the thrombotic or non-thrombotic obstruction of hepatic venous outflow anywhere along the venous course from the hepatic venules to the junction of the inferior vena cava and the right atrium. The etiology of Budd-Chiari syndrome is classified as primary, attributable to intrinsic intraluminal thrombosis or the development of venous webs; or secondary, caused by intraluminal invasion by a parasite or malignant tumor or extraluminal compression by an abscess, solid tumor, or cyst, such as a hydatid cyst. In this study, we present a case of a giant hydatid cyst manifesting Budd-Chiari syndrome symptoms and signs by compressing the inferior vena cava and hepatic veins. In brief, the case demonstrates that hydatid disease should be considered in the differential diagnosis of Budd-Chiari Syndrome in areas such as Turkey, where hydatid disease is endemic.

Key words: giant hydatid cyst; liver; Budd-Chiari syndrome; external compression


Introduction

Hydatid disease is a parasitic infection caused by several species of the Echinococcus cestode. The most common form is E. granulosus, which produces cysts, primarily in the liver and lungs. Much less common is E. multilocularis, which produces an invasive tumor-like replacement of liver tissue. Hydatid disease is common in agricultural and pastoral societies and continues to be a serious public health problem in many countries, including Turkey [1,2]. The clinical signs and symptoms of hydatid disease vary from incidental findings to severe, life-threatening complications [1]. The most important factors in the development of these complications are the number, size and localization of cysts, and their relationship with adjacent organs and the hepatic vascular system. One of the most important complications of hydatid cyst disease of the liver is external compression of the liver and the surrounding vascular system [1,3]. The signs and symptoms of Budd-Chiari syndrome (BCS) may develop as a result of compression of the inferior vena cava (IVC), hepatic vein, and portal venous system by hydatid disease, portal hypertension, and thrombosis [1]. Similarly, compression of the biliary system may result in various degrees of signs and symptoms of obstructive jaundice.

BCS is defined as an obstruction of hepatic venous drainage due to various causes, leading to progressive liver damage and portal hypertension. Occlusion may occur at the level of the hepatic veins or the IVC at any point between the entrance to the hepatic veins and the right atrium [4]. The etiology of BCS can be classified as primary, attributable to intrinsic intraluminal thrombosis or the development of venous webs; or secondary, caused by intraluminal invasion by a parasite or malignant tumor or extraluminal compression by an abscess, solid tumor, or cyst, such as a hydatid cyst [5]. In this study, we present the case of a patient with a giant hydatid cyst manifesting with BCS symptoms and signs caused by compression of the IVC and hepatic veins.

Methodology

A 67-year-old woman had suffered from vague abdominal pain for four years and experienced malaise, fatigue, and anorexia for the previous two months. She had been referred to a private clinic three days previously with complaints of severe abdominal pain, high fever, and leg swelling. An abdominal examination revealed signs of severe peritonitis, and concave dullness on percussion was noted.
Laboratory examinations were as follows: white blood cell count: 17,300/mL (4.3–10.3); hemoglobin: 10 g/dL (13.6–17.2); platelets: 416,000/mL (156–373); albumin: 1.8 g/dL (3.4–4.8); total bilirubin: 3.55 mg/dL (0.2–1.2); direct bilirubin: 2.6 mg/dL (0–0.5); alkaline phosphatase: 302 U/L (40–150); blood urea nitrogen: 36 mg/dL (9.8–20.1); creatinine: 1.38 mg/dL (0.5–1.1); potassium: 6 mmol/L (3.5–5.1); sodium: 130 mmol/L (136–145); and INR: 1.8. All viral markers were negative.

A multi-detector computerized tomography (MDCT) scan revealed a cystic lesion (hydatid cyst) of approximately 20 × 16 × 18 cm with a lobulated contour, which was septate and completely filled segments 7 and 8 of the right lobe of the liver, extending to segments 4 to 6 and elevating the diaphragm. Furthermore, congestion was detected in segments 4 and 5, showing that venous drainage had worsened (Figure 1A). A surgical operation was planned, as the patient’s examination was compatible with severe acute peritonitis. After a laparotomy, approximately 5 L of cloudy ascites fluid were aspirated from the abdomen. Fluid analysis showed a leucocyte count of 1100/mL and a neutrophil ratio of 95%. An ascites culture was negative. The right lobe of the liver was severely atrophied and the left lobe was markedly hypertrophic. A wedge biopsy was taken from the left lobe due to the irregular appearance of the liver (Figure 2). Additionally, a cystic mass of about 20 × 20 cm with a thick wall completely covering the right lobe and invading the abdominal wall laterally and the diaphragm proximally was seen. After dissection, we determined that the cystic mass was compressing both the IVC and hepatic veins, thereby causing the BCS findings. After compresses soaked with 3% NaCl were placed around the cyst, a 10-mm trocar was inserted into the cyst, and the contents were aspirated. Then the anterior wall of the cyst was completely excised, after which all vesicular structures inside the cyst were evacuated from the abdomen. The operation ended after both biliary fistulas were closed primarily, and the cystic cavity was catheterized with a drainage catheter. Compression on the IVC and biliary ducts was completely relieved and the dilatation in the biliary ducts was markedly reduced on an MDCT scan taken on postoperative period (Figure 1B). After surgery, the patient was started on albendazole (10 mg/kg) for six weeks. The histopathological examination of the liver was compatible with stage II-III cirrhotic process and hydatid cyst.

**Figure 1.** Multidetector computed tomography images in the sagittal planes following contrast injection.

Sections taken during the preoperative period (A) show a 20 × 16 × 18 -cm cystic lesion with a lobulated contour and septations filling the right lobe of the liver, extending to segment 4 and elevating the diaphragm. The retrohepatic segments of the inferior vena cava (IVC), right hepatic vein, and middle hepatic vein could not be visualized.

Sections taken on postoperative day 3 (B) show a prominent retrohepatic segment of the IVC, indicating that IVC compression was relieved.
Discussion

Hydatid cysts are usually asymptomatic until diagnosed incidentally or various complications occur. Complications of hepatic hydatid disease occur in 5% to 40% of patients and include the formation of small cystobiliary fistulas, cyst rupture into the biliary tree, biliary compression, adjacent organ compression, cyst infection, spontaneous or traumatic intraperitoneal rupture, and, rarely, hepatic and perihepatic vascular tree compression [3,6].

The exact prevalence of BCS is unknown, but it has been estimated to affect 1/100,000 individuals in the general population worldwide, with a higher prevalence occurring in developing countries [5,7]. BCS affects individuals of all racial groups, usually during the third or fourth decade of life.

BCS can be classified as primary or secondary depending on the origin of the obstructive lesion. The primary type is caused by a hepatic venous outflow obstruction originating from an endoluminal venous lesion, such as the thrombus of a membrane. BCS is considered secondary when the obstruction of hepatic venous outflow results from the presence of material in the lumen not originating from the venous system, or from extrinsic compression [7,8]. Studies from different countries and continents have reported a large variety of BCS etiological factors. For example, Behcet's disease and hydatid disease, which are rare etiologies of BCS in many studies, are responsible for most BCS cases in Turkey. Most BCS etiological agents are identifiable in Western countries, whereas most cases in India and Japan are idiopathic. One of the most important causes of BCS in Western countries is the tendency for thrombosis, whereas the cause is caval webs in Eastern countries [9]. The table lists some of the diseases causing BCS.

The clinical presentation of BCS depends on the extent and rapidity of hepatic vein occlusion and on whether a venous collateral circulation has developed to decompress the liver sinusoids. According to the duration of disease, BCS can be classified as fulminant, acute, subacute, or chronic. Patients with the fulminant form present with hepatic encephalopathy within eight weeks after developing jaundice; however, this presentation is uncommon. The subacute form, which is most common, has a more insidious onset; ascites and hepatic necrosis may be minimal because the hepatic sinusoids have been decompressed by the portal and hepatic venous collateral circulation. The acute form presents with symptoms of short duration, intractable ascites, and hepatic necrosis with no venous collateral formation. The chronic form manifests as a complication of cirrhosis.

Abdominal pain, fever, ascites, leg edema, and hepatomegaly are present in almost all patients with BCS. Jaundice, gastrointestinal bleeding, and hepatic encephalopathy are less common [4,10]. However, the decompression of liver sinusoids by large intrahepatic and portosystemic collaterals has been described in asymptomatic patients (15% to 20%) with hepatic vein thrombosis.
The radiologic diagnosis of BCS in a patient with a hydatid cyst can be established using ultrasonography (US), color Doppler ultrasound (US), CT, and magnetic resonance imaging (MRI). Doppler US of the liver, with a sensitivity and specificity of 85% or more, is the technique of choice for the initial investigation when BCS is suspected [4,7]. CT scans may be recommended to visualize the vascular anatomy and assess the configuration of the liver when a transjugular intrahepatic portosystemic portosystemic shunt is considered. Unvisualized hepatic veins on CT are suggestive of disease, but false-positive or indeterminate results can occur in 50% of cases. MRI should be performed as a second-line imaging modality. MRI enables the visualization of hepatic vein thrombosis and the evaluation of the IVC, but it is more expensive than CT. Hepatic X-ray venography is the reference procedure to evaluate hepatic veins, the extent of thrombosis, and caval pressures. Inferior cavography should be performed to demonstrate stenosis or occlusion of the IVC [4,7].

Management of BCS should be planned depending on the etiology and the duration and severity of symptoms. Thrombolytic therapy should be the first choice in patients with acute thrombosis. Angioplasty with or without stenting is employed in cases of a web or stenosis in the IVC or focal hepatic venous stenosis [1]. A transjugular intrahepatic portosystemic shunt is the most convenient method for managing acute BCS, subacute BCS with a portocaval pressure gradient less than 10 mmHg, or BCS with an occluded IVC. The surgical placement of a shunt is a feasible approach in cases with a portocaval pressure gradient greater than 10 mmHg and in some cases of subacute BCS. However, the final treatment modality for fulminant BCS, cirrhotic cases, and cases responding inadequately to the surgical placement of a shunt is liver transplantation. All of these therapies have advantages and disadvantages. Thus management should be planned case by case.

A surgical approach should be used to eliminate compression because a venous outflow disorder due to external compression develops in almost all cystic Echinococcus cases causing BCS. Decompression by partial cystectomy normalizes venous drainage in cases lacking cirrhotic transformation. Radical surgical options, such as pericystectomy, should be avoided due to the high risks of hemorrhage and liver injury. Postoperative antihelmintic treatment duration may vary from three to eight weeks, depending on the surgery. The most frequently used agents are albendazole, mebendazole, and praziquantel.

Although rare in the Western world, hydatid disease should be considered in the differential diagnosis of BCS in areas such as Turkey, where hydatid disease is endemic. Decompression of cyst pressure is adequate treatment for early-stage cases.
References

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