

Hydatid Cyst of Liver: A Rare Cause of Secondary Budd-Chiari Syndrome

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ABSTRACT

Hydatid cyst of the liver, causing compression of the inferior vena cava (IVC) and hepatic veins is a rare cause of secondary Budd-Chiari syndrome (BCS). As the hydatid disease is endemic in India, it is a rare but treatable cause of BCS. The early diagnosis and timely intervention can prevent hepatic complications leading to BCS in the affected patients.

Keywords: Budd-Chiari syndrome, Hydatid disease, Magnetic resonance imaging.

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INTRODUCTION

Hydatid cyst of the liver, causing compression of the inferior vena cava (IVC) and hepatic veins is a rare cause of secondary Budd-Chiari syndrome (BCS). India has been documented as a highly endemic area of hydatid disease.¹ Due to varied presentation of this disease in patients it is rarely considered in the differential diagnosis of diseases causing BCS and portal hypertension.^{2,3}

This case report highlights the rare complication of hydatid cyst of liver leading to secondary BCS diagnosed on imaging.

CASE REPORT

A 35-year-old female hailing from a rural area was referred to our tertiary center with abdominal pain and gradually increasing lump in the right hypochondrium for the last 7 years. Examination of the head, neck, chest and cardiovascular systems were normal. On abdominal examination there was minimal tenderness in the right upper quadrant on palpation hepatomegaly with firm nodular liver was palpable 5 cm below xiphisternum. Spleen was palpable 4 cm below left costal margin. There was no pedal edema or ascites. Her laboratory investigations revealed hemoglobin 10.3 g/dl; total leukocyte count $8.834 \times 10^9/l$; differential count: Neutrophils 62%, lymphocytes 32%, monocytes 3% and eosinophils 3%; platelets $210 \times 10^9/l$. Serum bilirubin 3.0 mg/dl, serum aspartate aminotransaminase 56 IU/l, serum alanine aminotransaminase

80 IU/l, alkaline phosphatase 475 IU/l and prothrombin time 13 sec. Upper GI endoscopy was normal and varices were not seen.

Contrast enhanced computed tomography (CECT) abdomen had been done prior to referral and it demonstrated a large, partially exophytic mass with foci of calcifications in liver which was reported as a malignant hepatic mass. The patient was referred to us for magnetic resonance imaging (MRI) and MR venography. There was a $17 \times 6.3 \times 5.7$ cm exophytic hepatic mass lesion appearing hypointense on T1 weighed images and hyperintense on T2 weighed images with a T2 hypointense rim and showing no significant contrast enhancement. It was extending into the subphrenic space and gastro-hepatic ligament. The mass was causing compression of the intrahepatic IVC and the hepatic veins were not visualized. The mass was also causing mild segmental dilatation of segment 2 and 3 biliary radicles. The hemiazygos and azygos venous systems were dilated with dilated splenoportal axis. Multiple intrahepatic collaterals were also seen (Fig. 1). The findings were suggestive of calcified, complex hydatid cyst causing secondary BCS. Hydatid serology was positive. Biphase CECT abdomen was repeated for presurgical evaluation. It showed a large hydatid cyst with peripheral calcification causing compression of the hepatic veins with mild splenomegaly and prominent hemiazygos and azygos venous system

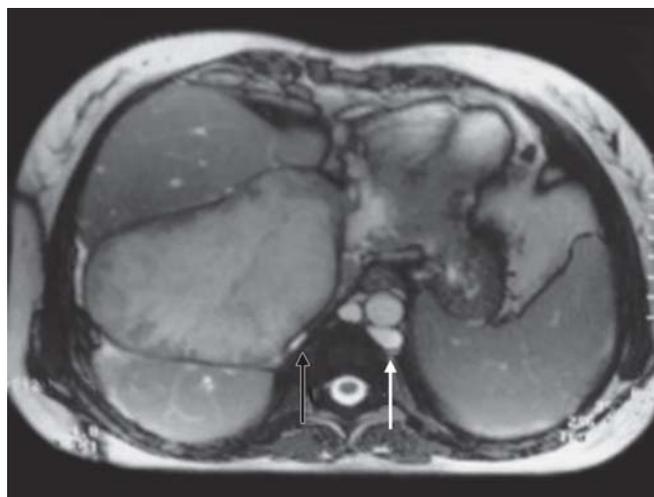


Fig. 1: Axial TRUF1 sequence demonstrating the compression of the intrahepatic IVC (black arrow) by the mass. Note the dilated hemiazygos and azygos venous system (white arrow)

(Fig. 2). Few dilated collaterals were also seen in the gastrohepatic ligament and at splenic hilum.

On color Doppler, a complex calcified echogenic mass was seen causing marked luminal compression of the intrahepatic IVC with nonvisualized hepatic veins. Few intrahepatic collaterals were also seen.

FNAC of the patient was done from the mass which proved it to be hydatid cyst. The patient was taken up for surgery and the cyst was resected along the planes of the hepatic veins and IVC with all precautions to prevent spillage of the hydatid fluid. Postoperatively, the patient recovered well and a repeat color Doppler after 3 months showed normal flow in the intrahepatic IVC and hepatic veins with resolution of the intrahepatic collaterals.

DISCUSSION

Obstruction of hepatic venous outflow is the characteristic feature of BCS. The obstruction has to involve at least two of the hepatic veins to produce BCS. The most common cause of BCS in Asia is membranous obstruction. Hydatid disease of the liver is a rare cause of BCS and is primarily encountered where the disease is endemic and has been diagnosed as the second most frequent cause after membranous obstruction of BCS in an endemic areas.^{4,5} In India, hydatid cyst causing BCS was first described by khuroo MS.⁶

Hydatid cyst of liver causing BCS is usually rare because the cyst has to be large in size and be in a specific position so as to compress the IVC and hepatic veins. It has to cause extrinsic compression of at least two suprahepatic veins and affect two or more hepatic segments of the dome.⁷ The lesions can vary from few millimeters to large (15-20 cm) size. As the cyst grows in size it causes

compression of the IVC/hepatic veins leading to mechanical mass effect as well as inflammatory effect due to cyst contents leading to venous thrombosis. As the disease progresses signs of portal hypertension due to hepatic veins compression can occur.

Ultrasound can demonstrate the cyst, its position, internal contents, calcifications and relation with the vessels along with other signs of BCS like enlargement of the caudate lobe, ascites, splenomegaly and narrowing, nonvisualization or thrombosis of the hepatic veins. On color Doppler, absent or monophasic flow in the hepatic veins, inferior vena cava, or both and intrahepatic collaterals may be seen.⁸ CECT abdomen provides better assessment of hepatic parenchyma, hepatic veins and IVC along with the relationship of the cyst with the vascular structures. Signs suggestive of BCS include: abnormal hepatic perfusion, caudate lobe hypertrophy, thrombus formation in the inferior vena cava or the hepatic veins, intrahepatic collaterals, regenerative nodules or calcifications.⁹ MRI is the modality of choice for depicting the hepatic veins, IVC and the relationship with the cyst. It clearly shows thrombosis of hepatic veins, inferior vena cava or both, along with other features of BCS like mosaic attenuation of the liver, caudate lobe hypertrophy and intrahepatic collaterals.¹⁰

Surgery is usually required for secondary BCS due to extrinsic compression of the hepatic veins by hydatid cysts but has increased risk of increased mortality and morbidity. Nowadays venoplasty and stenting can be done instead of cyst excision¹¹ with good results.

BCS is a common condition in our country and our main emphasis should be in its early diagnosis and detection, so as to prevent complications like portal hypertension and subsequent liver damage. We would like to emphasize considering hydatid disease as a rare but treatable cause of secondary BCS in endemic areas as in India.

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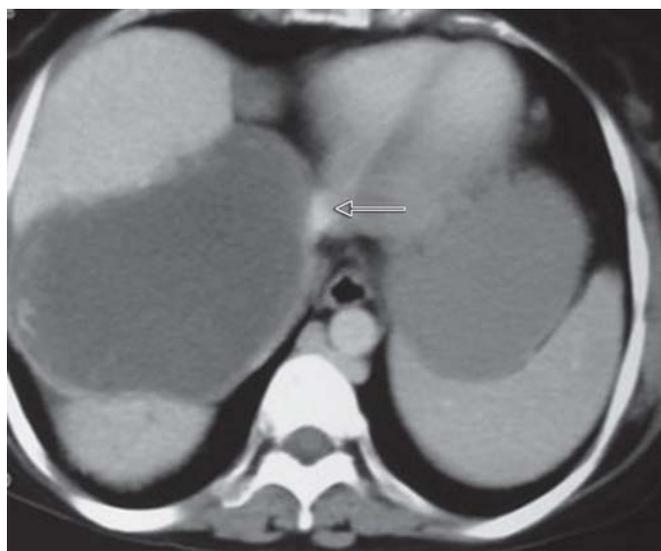


Fig. 2: CECT abdomen shows a large calcified hydatid cyst causing compression of the IVC (white arrow)

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