Portal Hypertension as a Rare Complication of Hydatid Cyst: Diagnosis and Management

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Abstract

Hydatid cyst of the liver is a significant health problem in endemic areas. Echinococcus granulosus has a better prognosis than E. alveolaris. Portal hypertension (pre-hepatic, hepatic, post-hepatic) is a very rare complication of hydatid cyst of liver. A literature search revealed only eleven publications on the subject. The mechanism depends on the cyst type and cyst location; with alveolar disease causing hepatic (cirrhosis) type of portal hypertension in the largest number of cases. Unilocular disease can also cause pre- and post-hepatic portal hypertension by localized obstruction, and is more amenable for surgical therapy. Computed tomography can identify certain specific signs of the post-hepatic type. A high index of suspicion should be reserved for patients of liver hydatidosis who develop gradual-onset ascites.

PLACE OF STUDY/SOURCE

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INTRODUCTION

Hydatid cyst of the liver is a significant health problem in endemic areas. In India, the disease has a high prevalence, particularly in the farming community. Echinococcus granulosus, which causes a unilocular type of cyst, has better a prognosis than E. alveolaris, which causes a multilocular infiltrating type. Symptoms can vary form non-specific mild abdominal pain to severe anaphylaxis due to rupture into the peritoneum. Most symptoms can be attributed to compression or displacement of surrounding structures, or, to ingrowth into hollow viscera. [1] Portal hypertension (PHT) is a very rare complication of hydatid cyst of liver. On a search of PubMed™ and MedLine™, using the keywords 'portal hypetension' and 'hydatid cyst', we could identify only six publications in the English language [2-7], and five in other languages [8-12] dealing with PHT due to liver hydatidosis (LH). In a recent retrospective analysis of 35 patients of liver hydatidosis treated surgically over a period of 20 years, Avgerinos et al., [3] identified PHT as a complication in only one case. The mechanism of PHT can be pre-hepatic, hepatic, or, post-hepatic (Budd-Chiari syndrome). The presentation and management of PHT due to LH, according to the causative mechanism, is reviewed below.

PRE-HEPATIC

There are 3 reports where the authors have identified a pre-hepatic mechanism causing PHT in LH. [4,5,11] El Fortia et al., reported a 60-year-old woman with perihilar splenic varices without other signs of PHT. [4] Ultrasound alone was useful in identifying compression at the splenic hilum by a calcified hydatid cyst, and, splenectomy was performed to resolve the PHT. In the other two cases, the obstruction was at the hepatic hilum causing portal cavernomatosis. [5,11] One of these required shunt surgery [5], while the other [11] was detected to have multiple other intrahepatic cysts on CT and MR imaging. The latter case was planned for liver transplantation due to liver failure, but died awaiting surgery following a variceal bleed.

From these cases, it may be ascertained that pre-hepatic PHT is usually caused by a unilocular hydatid cyst. CT scan would be useful for delineating the obstruction. Treatment in most cases would be surgical removal of the obstructing cyst, or, porto-systemic shunt surgery.

HEPATIC

Cirrhosis of the liver with portal fibrosis can occur in advanced cases of LH. This mechanism has been reported in 10 cases by 3 authors. [7,10,13] In most of these patients, the offending organism was Echinococcus alveolaris. This is not

surprising, as infiltrative growth of E. alveolaris into the bile ducts is known to cause cirrhotic changes in the liver. These patients are likely to present with ascites, and other signs of liver cell failure. The disease is progressive with a poor prognosis. Treatment is supportive, as radical surgery is mostly unsuccessful. [6,13] Liver transplantation for advanced alveolar echinococcosis (EA) was performed for 17 patients by Bresson-Hadni and colleagues. [13] Fifteen of these patients had PHT, which in 7 was of the Budd-Chiari type, and in the rest was hepatic. There was a higher recurrence rate and lower survival in patients with EA than in patients with livers transplanted for other indications. Thus, liver transplantation cannot be routinely recommended for all patients with EA.

POST-HEPATIC

There are twelve instances of post-hepatic PHT, or the Budd-Chiari syndrome due to LH described in literature. [2,6,8,9,13] Of these, nine patients had EA. In alveolar disease, the post-hepatic mechanism often co-exists with cirrhosis, and the treatment and prognosis for such patients is similar to those described by Bresson-Hadni [13] (vide supra). In five patients [2,6,8,9], there was localized obstruction to the hepatic veins or the inferior vena cava. This focal obstruction was caused by E. granulosus in three, and E. alveolaris in two patients. In all five patients, the liver functions were normal. CT was effective in diagnosis by identifying compression at the caval site, presence of collaterals, non-visualization of hepatic veins, and, ascites. [6,8,9] Because of the unusual location of the disease, different strategies have been adopted by different authors. Combined resection of the liver and vena cava was performed by Mekeel et al., [2], though this has been debated by some as being too radical for benign disease. [14] A novel therapy has been successfully attempted by Vogel and colleagues in the form of percutaneous stenting of the hepatic veins. [6] The authors recommend this approach in selected patients of posterior localized alveolar disease where liver function is normal. They believe that resection for such cases is often only palliative with short-lasting benefit, thereby not justifying the associated morbidity.

We feel that due to the rarity of the condition, no recommendation can be made, and the treatment needs to be individualized. Operative resection seems to be an attractive option more for unilocular disease. The patients with LH and chronic Budd-Chiari syndrome would be very difficult to identify in the clinical setting. Ascites may be the only symptom, and liver functions may be normal. A high index of suspicion is required, along with careful interpretation of the subtle CT findings. Direct measurement of portal pressure by invasive techniques is not practical in the clinical setting.

When the ascites is acute in onset, the first suspicion must be of cyst rupture, and urgent steps must be taken to provide definitive treatment and prevent anaphylaxis. A single case of acute Budd-Chiari syndrome due to unilocular cyst has been reported. [8]

CONCLUSIONS

Review of previous experiences of this rare manifestation of LH leads us to the following conclusions:

PHT in LH is rare, and can occur by all three mechanisms. The exact mechanism in a particular case depends on the cyst type (unilocular or multilocular), and cyst location. Identification of the mechanism helps in deciding therapy.

A patient of LH with ascites or gastrointestinal bleed must be suspected to have PHT, but only after satisfactorily ruling out cyst rupture as the cause of ascites.

CT scan is useful in diagnosis, and specific CT signs have been identified. Doppler ultrasound may also be helpful for follow-up of treated patients.

EA is more commonly responsible for PHT, by causing either liver cirrhosis or the Budd-Chiari syndrome. A few cases of E. granulosus causing localized pre- or post-hepatic PHT have been reported.

For localized disease, operative resection is a feasible option, especially with E. granulosus. For cirrhotic patients, treatment needs to be individualized, and liver transplantation may help a few selected patients.

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