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Hydatid Disease of the Liver: Spontaneous Rupture of the Portal Vein Into the Cyst Wall (A Rare Complication)

A 60-year-old man was admitted with a complaint of pain in the right upper quadrant of the abdomen together with itching. He had neither jaundice nor fever. He had not developed an anaphylactic reaction. On physical examination, maculopapular lesions and urticaria were observed. Eosinophilia was not defined. ELISA testing for hydatid disease was positive. The results of serologic tests for Echinococcus (indirect hemagglutination inhibition test, more than 1500) were abnormal. Ultrasound (US) (Lite-8) revealed a total of four cysts in segment 3 (approximately 4 cm), segment 4 (approximately 5 cm), segment 5 (approximately 3.5 cm) and segment 8 (approximately 8 cm). In segment 4, calcification of the cyst wall was observed radiologically and a pericystectomy was performed. Similar size cysts were found in Computerized Tomography (CT) (Figure 1). We based a Gharbi classification on the sonographic analysis of the morphology and structure of the hydatid cyst of the liver (8). According to this classification, the cysts were type III and type IV. In segment 3 and segment 8, a partial cystectomy was performed and temporary drainage of the perihepatic region was performed. In segment 5, a controlled cystostomy was carried out; however, upon entering the cystic lumen to administer scleroidal agent, abandoned bleeding started. There was a window between the cavity of the cyst and portal vein. When the Pringle maneuver was stopped for testing, abandoned bleeding and cystic vesicles were seen to come through the lumen. The patient had systemic hypotension due to bleeding during the operation. No anaphylactic shock was observed. A partial cystectomy was performed. The procedure involved removal of the hydatid debris and closure of the portal vein defect with a flap formed from the cyst wall. The flap was sutured with 5/0 polypropylene material and temporary drainage of the perihepatic region was performed. The patient was asked to come to a control session and physical examination. Liver tests, immunoserologic tests, CT, abdominal US and a splenoportogram were performed. The skin lesions of the patient recovered rapidly two months after the operation. Clinical signs, CT and US examination of the liver were normal. The results of an indirect hemagglutination inhibition test were normal in the first month after the operation. In the splenoportogram, the lumen of the portal vein was open. However, in the repaired portal vein segment, the contour of the vein was irregular (Figure 2).

Liver hydatidosis is a progressive, often recurrent (10 to 20% of all cases) disease (5, 9). In our ward, nearly 50 cases of hepatic hydatid disease are operated on. Of these, 6-8% comprise recurrent cases. In the present case, surgery was arranged for recurrence. The disease can involve any organ and mimic any pathologic condition.

Figure 1. CT scan showing the cysts.
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The skin lesion was interesting because it looked like nonspecific urticaria.

Symptoms arise either when the cyst has grown enough to cause pressure on adjacent organs or when a complication occurs (10). Rupture, secondary infection, and suppuration are the most common complications (2, 4). In our case, there was no secondary infection. Rupture of the liver cyst into adjacent structures such as the lung, biliary tree, and peritoneum is not uncommon and is usually associated with pain and fever (1). In rare cases, hydatid cysts can be opened to the portal vein and hepatic vein (6, 7). Angiography is very helpful.

Sclerosant injection and ligation depending on vascular complications, or anatomic and non-anatomic hepatic resection can be carried out (9, 11, 12). In the present case, the site of the opening into the portal vein was before branching. Surgical treatment other than venous wall repair (saphenous vein patch or synthetic patch) can be performed. However, we repaired the defect using a flap from the cystic wall an alternative method to the method described above. During the operation, the portocystic shunt was visually and clearly observed. However, it would have been better if we could have recognised the need for a preoperative splenoportogram or scintigraphy.

Diagnosis was made using clinical criteria, imaging techniques, skin tests and serology (1). In our case, the hepatic mass was palpable on physical examination. CT scans of the liver are helpful in establishing the diagnosis of echinococcal liver disease, as they often clearly delineate the calcified wall of the hydatid and daughter cysts within the hepatic cyst, and, in addition, the cyst can be localized in the liver by abdominal US (2). Other imaging methods such as angiography and scintigraphy are of little value in distinguishing echinococcal cysts from other space-occupying lesions of the liver (13). Eosinophilia is observed in only 5 to 15 per cent of infected individuals. In our case, eosinophilia was not observed. The diagnosis is confirmed serologically by the hydatid complement fixation and hydatid latex agglutination tests (2,10). The Casoni skin test is associated with a large number of false positive results and has been replaced by indirect hemagglutination and serum immunoelectrophoresis, which have a diagnostic accuracy of 85 to 90 per cent (4). The results of indirect hemagglutination testing were abnormal in our patient. The Elisa test was positive. The Casoni skin test and immunoelectrophoresis were not performed. A CT scan and splenoportogram were performed 4 months postoperatively. There was a pattern of serological response in the patient following the intervention and a significant decrease in the titer in the first 3 months.

In conclusion, penetration of a cyst into a vessel is a very rare complication (6, 7); however, spontaneous operative rupture of the portal vein into the cyst wall has been reported for the first time. In our case, the repair was able to be performed with a vein patch or a prosthetic graft to the defect of the portal vein. However, we observed that repair of the portal vein defect with the technique using a flap of cystic wall was also applicable. In addition, in suspected cases, a splenoportogram should be performed preoperatively to help determine the type of procedure to be used for repair.

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