Missed Diagnosis

Carolí’s disease misdiagnosed as hydatid liver cysts

M. Akoglu and B.R. Davidson

Department of Gastrointestinal Surgery, Turkey Advanced Specialization Hospital, Ankara, Turkey and Academic Department of Surgery, Royal Free Hospital and School of Medicine, London, UK.

Summary: A 27 year old woman who presented with upper abdominal pain was found on ultrasonography to have multiple liver cysts consistent with hydatid disease. Three years previously she had undergone evacuation of multiple infected liver cysts thought to be due to hydatid disease. Computed tomographic scanning supported the diagnosis of hydatid disease affecting the right lobe of the liver. At laparotomy the right lobe contained multiple cysts which were removed by right lobectomy. Histology revealed congenital dilatation of the intrahepatic bile ducts with fibrosis (Carolí’s disease) but no evidence of hydatid disease.

Introduction

In countries where hydatid disease is endemic, ultrasonography is the most commonly used investigation in patients suspected of having hydatid liver cysts. Only when the possibility of complications is present, such as patients presenting with jaundice or cholangitis, would computed tomographic (CT) scanning or cholangiography normally be considered. Although this policy is satisfactory for the majority of patients it will result in occasional misdiagnoses. A case is presented in which a pre-operative diagnosis of hydatid liver disease based on the patient's history, clinical findings and both ultrasound and CT scanning was found at subsequent laparotomy and histological examination to be congenital dilatation of the intrahepatic bile ducts (Carolí’s disease).

Case report

A 27 year old female school teacher presented to the out-patient clinic of the Turkey Advanced Specialization Hospital with an exacerbation of right upper quadrant pain which had been present for many years.

Three years previously she had been investigated and treated at another centre for a similar upper abdominal pain. She had been found to have a palpable hepatomegaly and ultrasonography showed the presence of multiple cysts in the right lobe of the liver suggestive of hydatid disease, although hydatid serology (IgG, IgM, indirect haemagglutination and Casoni skin tests) had been negative. A laparotomy was performed with evacuation of infected liver cysts and biopsy of the cyst wall. The biopsy of the cyst wall showed non-specific infective granulation material.

Following evacuation of the liver cysts her right upper quadrant pain continued. One year later she again was found to have hepatomegaly and ultrasonography showed multiple cysts in the right lobe of her liver. She continued to be followed up at the out-patient clinic over the subsequent 3 years where her general condition was found to be satisfactory although her abdominal pain and hepatomegaly persisted. Ultrasonography, repeated on several occasions, showed no evidence of resolution of her cysts, some of which contained sludge (Figure 1).

Clinical examination on arrival at the Advanced Specialization Hospital revealed a well looking woman with a marked hepatomegaly.

Abdominal ultrasound examination showed the presence of multiple cysts, some containing sludge, within the right lobe of the liver. The appearances were suggestive of recurrent hydatid disease. An abdominal CT scan was carried out which supported the ultrasound findings, the whole of the right lobe being occupied by multiple cysts and the left lobe being hypertrophied in compensation (Figure 2). No cysts were present in the other abdominal organs. Blood tests on admission, including full blood count, liver function tests and clotting screen, were normal.

A further laparotomy was carried out at which the right lobe of the liver was found to be enlarged by multiple cysts, the left lobe appearing normal. A right hepatectomy was performed. On the evening following surgery a further laparotomy was required for post-operative haemorrhage. Her post-oper-
thin or thick walled, containing daughter cysts or a floating lamina, are infected or have ruptured into the biliary tree. In the present case the ultrasound examination carried out prior to the initial surgery showed multiple large cysts with hypoechoic shadows. These findings, suggestive of a thick walled cyst, were felt to represent mature hydatid cysts. Although infection of a hydatid cyst produces thickening of the cyst wall the ultrasound findings are entirely non-specific. Similarly the initial biopsy report of non-specific infected granulation tissue failed to clarify the diagnosis.

To corroborate the findings of ultrasonography, hydatid serology was carried out with measurement of serum immunoglobulins, the indirect haemagglutination and the Casoni skin test. Although these were negative this does not preclude hydatid disease, standard serological tests being falsely negative in 10–20% of cases. Similarly, false positive serology may occur particularly in patients with malignancies or liver disease. Newer serological tests for hydatid disease are both more sensitive and more specific and may, therefore, have provided reassurance that hydatid disease was not present.

CT scanning did not provide any additional information on the extent of liver disease in the present case although it did exclude the presence of cysts within other organs. This is a finding in common with other studies comparing ultrasonography with CT scanning for assessing hydatid disease where CT scanning has been found to add little information to that of ultrasonography.

Caroli’s disease is a congenital saccular dilatation of the intrahepatic bile ducts. It is rare but most often presents with cholangitis, predisposed to by the formation of stones within the abnormal ducts. Occasionally, as in the present case, the changes of bile duct ectasia and hepatic fibrosis may be confined to a segment or lobe of the liver. The presentation and findings in the present case are unusual in view of the lack of cholangitis, absence of intrahepatic stones, the localization to the right lobe and the persistence of fluid-filled cavities. In retrospect the initial operation is likely to have been the drainage of a liver abscess, a common complication of Caroli’s disease, rather than the evacuation of an infected hydatid cyst.

The diagnosis of Caroli’s disease was suspected at the second operation due to the fibrous texture of the liver. Liver resection is generally contraindicated in this disease due to the diffuse and extensive nature of the pathological changes. Drainage of the biliary tree by the formation of a biliary-enteric anastomosis may prevent recurrent cholangitis and can be fashioned to allow subsequent percutaneous procedures through a Roux-en Y-loop. Resection is, however, an accepted plan of management when the disease process is local-

Discussion

Many studies have suggested that ultrasonography is a reliable method for the diagnosis of hydatid liver cysts and that an accurate evaluation may be made as to whether the cysts are single or multiple,
ized to a single segment or lobe of the liver as occurred in the present case.\textsuperscript{12} Resection of the affected lobe of the liver is also controversial in the management of hydatid disease, the majority of cysts being adequately treated by evacuation or local excision along with tube drainage or omentoplasty.\textsuperscript{13} When extensive disease is present and resection is technically feasible, however, resection may be considered in centres where this is associated with an acceptably low morbidity and mortality.\textsuperscript{14}

This case has demonstrated that the common practice of using ultrasonography alone for the diagnosis of liver hydatid disease may lead to misdiagnosis. This may be avoided by combining imaging with accurate serological testing.\textsuperscript{15}

References


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