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Case report

Hepatobiliary cystadenoma: diagnostic uncertainty

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Background

Hepatobiliary cystadenoma is a rare tumour that can be clinically 'silent' and only discovered as an incidental finding on ultrasonography (US). It can also be symptomatic with abdominal pain and jaundice or develop internal bleeding and (exceptionally) malignant degeneration. Therefore the treatment of choice is liver resection.

Case outline

A 77-year-old woman was admitted with mild jaundice and right hypochondrial pain. For 7 years she had been known to have a silent liver lesion, always considered to be a simple cyst on US. US and CT scan revealed a multi-septate mass involving segments IV, V and VIII of the liver, with thick walls, no calcifications and no contrast enhancement. US-guided aspiration showed the presence of old blood-stained material. The patient was operated with a suspected diagnosis of bleeding into a simple cyst. A Lin fenestration was performed with wide excision of the anterior wall of the cyst. Pathological examination demonstrated a mucinous hepatobiliary cystadenoma. The postoperative course was uneventful, and follow-up at 2 years confirmed no recurrence.

Discussion

Imaging will normally help to distinguish the occasional hepatobiliary cystadenoma from the common simple cyst. Otherwise, when a complication occurs, preoperative differentiation may become impossible, and requires histological examination of the cyst after surgical removal.

Keywords

hepatobiliary cystadenoma, biliary cyst, fenestration

Introduction

Hepatobiliary cystadenoma is a rare entity, although an increased incidence has recently been noted with the widespread use of ultrasonography (US) and computed tomography (CT) [1]. The diagnosis is based on certain specific features, which permit cystadenoma to be distinguished from the much more frequent simple cyst. The standard management is always operative because bleeding, infection, rupture and malignant degeneration may complicate the cystadenoma, causing difficulties in differential diagnosis. We present a case of complicated hepatobiliary cystadenoma.

Case report

A 77-year-old woman, a cardiopath with hypertension, was admitted as a surgical emergency with right hypochondrial abdominal pain and mild jaundice. She has been known for 7 years to have a giant hepatic cyst (12 × 6 cm), in the right liver that had always been asymptomatic and had not changed on serial US investigations over the years. On admission laboratory investigations showed cholestasis (total bilirubin 142.8 μmol/l ↔ (8.40 mg/dl); ↔ alkaline ↔ phosphatase 3402 IU/L, normal 98–279 IU/L). Also serum tumour markers were elevated (CEA 213 ng/ml, normal <5 ng/ml; CA 19.9 4960 U/ml, normal <37 U/ml). Other laboratory tests were within normal limits. US showed a large hypoechoic lesion (16 × 8 cm), occupying segments IV, V and VIII of the liver, with thick walls and a partially solid content. The right intrahepatic biliary ducts were dilated. On CT scan, the cyst appeared partially septate, compressing both the right biliary tract and the intrahepatic vena cava (Figure 1).

US-guided aspiration yielded normal hepatocytes and 1 L of old blood-stained material. The tumour marker levels of cystic fluid were normal (CEA and CA 19-9). After a temporary symptomatic relief for 2 days, almost complete re-filling of the cyst was noted on a repeat scan. In view of the clinical history and preoperative investigations, a diagnosis was made of haemorrhage into a large

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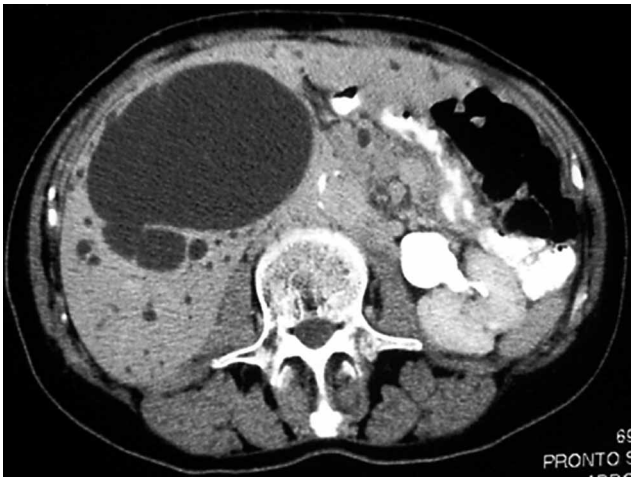

Figure 1. *CT scan showing hypodense mass with incomplete septations.*

simple cyst. As percutaneous aspiration only produced temporary relief of symptoms, operation was performed via a right subcostal incision. An enormous hepatic cyst was seen on the surface of the liver extending to the gallbladder and covered superiorly by a thin layer of parenchyma (Figure 2). Following cholecystectomy, 600 ml of bloody fluid were aspirated from the cyst. An extensive fenestration was performed at the lower part of the mass and also on the hepatic surface. Multiple biopsies of the internal wall of the residual cyst were obtained. Pathological examination showed a mucinous hepatobiliary cystadenoma, made up of a single layer of mucus-secreting columnar epithelium, supported by a basement membrane without evidence of cellular atypia or stromal invasion.

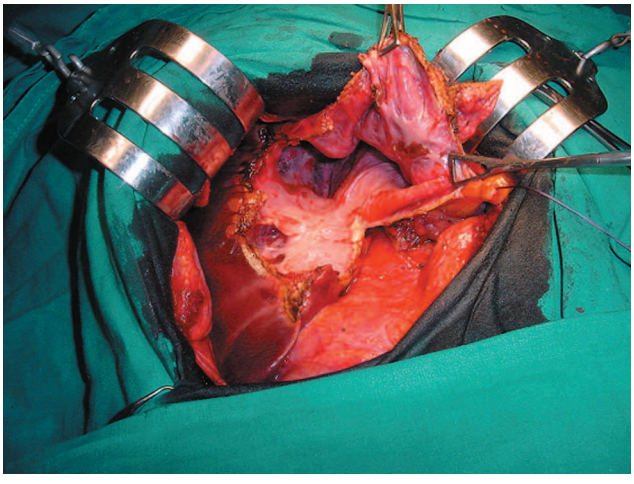

Figure 2. *Appearance of the giant septate cyst during the Lin procedure.*

The postoperative course was uneventful, with prompt resolution of jaundice and pain and normalisation of the serum tumour markers. In view of the age of the patient, the morphological features of the lesion (voluminous and central), and the histopathological absence of malignancy, we avoided definitive cyst resection. Two years later, the patient is in good health, and US confirms complete decompression of the biliary system with no recurrence.

Discussion

Since Keen reported the first biliary cystadenoma resection in 1892, fewer than 200 cases have been described [2]. Although the aetiology is unknown, many theories have been proposed, suggesting that these tumours derive from ectopic remains of primitive foregut, or result from obstruction of congenital aberrant bile ducts [3]. Biliary cystadenoma is usually encountered in women (96%), with a median age of 50 years.

The tumour is typically intrahepatic (80%), generally affects the right side of the liver, and varies in size from 2.5 to 30 cm [4]. Most patients remain asymptomatic for many years unless a complication occurs. Reported complications (5%) have included internal bleeding, infection, rupture and malignant transformation. In these cases the cyst may enlarge causing jaundice (30% for the intrahepatic form, 85% for the extrahepatic form), abdominal swelling or pain and fever if an abscess occurs [5].

Uncomplicated cystadenoma is often diagnosed incidentally on US and CT scans that show a thick external wall, internal septa, communication with the bile ducts, focal nodularity, calcification and sometimes polypoid projections [1, 4]. These features in association with increasing levels of CEA and CA 19-9 in blood or cyst fluid help to distinguish cystadenoma from the more frequent simple cyst [5, 6]. While surgical treatment for simple cyst is only needed for complications and Lin fenestration at laparoscopy or laparotomy is the best procedure, for hepatobiliary cystadenoma, complete resection is mandatory whenever possible, because partial excision is associated with a 50% local recurrence rate and the risk of malignant transformation over time [7–9]. Cystadenocarcinoma is an extremely rare tumour, with fewer than 40 case reports [10]. Differentiation from a benign tumour is based on laboratory and cytological

findings but sometimes may be impossible before operation. There is some evidence that elevation of CA 19-9 may be an important marker for these malignant tumours, yet transient elevation has been also reported in complicated benign cysts, as in our case [6].

When a non-parasitic cyst is complicated by bleeding or abscess formation, it enlarges to cause jaundice, as in the present case. Differentiation between cystadenoma and simple cyst then becomes very difficult before operation, because both lesions are likely to have a thick wall, internal septa, blood-stained fluid and increased levels of tumour markers [1, 4]. In our case, the cyst was complicated by bleeding, leading at first to expansion of the mass, with consequent pain and cholestasis, and secondly to the formation of intracystic septa and parietal thickening. The history and clinical presentation suggested a bleeding into a simple cyst.

As the cyst was very large and centrally placed with no suspicion of malignancy and as the patient was elderly and frail, complete resection was considered too aggressive. Lin fenestration was undertaken to provide wide unroofing of the cyst, to obtain a certain diagnosis and relieve pain, with a low morbidity rate and an acceptable chance of recurrence. Intra-operative frozen section would not have changed our surgical approach, first because this procedure has a low specificity, and second because the functional reserve of the non-resected liver (<30%), would have been insufficient to support a wide resection even in the presence of malignant change [5].

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