Episodic jaundice due to an intrahepatic biliary cystadenoma with biliary stricture masquerading as hydatid cyst

Introduction

Biliary cystadenomas are rare, benign but potentially malignant, multilocular, cystic neoplasms of the biliary ductal system. They usually arise in the liver (80-85%), accounting for less than 5% of cystic neoplasms of the liver.1 Less than 100 reports of intrahepatic biliary cystadenomas are identified in medical literature.2 In this case report, we present a patient with intrahepatic biliary cystadenoma (BCA) extending into the common bile duct (CBD), presenting as obstructive jaundice. Such occurrences are rare, and we review the imaging, diagnostic process and the surgical strategy.

Case Report

A 34-year-old lady was admitted with recurrent episodes of obstructive jaundice since 1 year. Liver function tests showed mild elevation of total bilirubin of 3.6 mg/dL; direct component: 2.1 mg/dL. Serum CA 19-9 was 35.9 U/mL. Echinococcal serology was equivocal. MRCP showed short segment stricture in CBD, 3.5 cm from the hilum with abrupt transition to normal caliber distally with moderate dilatation of intrahepatic ducts (Figure 1A). A well defined cystic lesion (4 x 4.5 cm) hypointense on T1 and hyperintense on T2 was seen in segment 4 with few thin internal septae, suspicious of hydatid cyst.

CECT revealed a suspicious intraluminal mid-CBD mass lesion, intrahepatic bile duct dilatation and segment 4 liver cyst (Figure 1B).

In view of CBD mass lesion associated with stricture, she was suspected to have a neoplastic mid CBD stricture along with segment 4 hydatid cyst of liver. Hence, she was planned for exploration for possible resection of the CBD lesion with adequate margins and clearance.

Figure 1: 1a) MRCP showing dilated proximal hepatic duct with abrupt cutoff (white arrow), 1b) CECT showing the segment 4 liver cyst (circle) & CBD mass lesion (arrow)
On exploration, upon transecting the CBD distally, a bile stained membranous mass protruded from the proximal dilated duct (Figure 2A & 2B). On further traction, it was found to be extending down from the segment 4 cyst. It was excised en toto along with the strictured segment of bile duct. Check choledochoscopy of the proximal biliary tree upto the second order division was normal. Upon probing the segment 4 cyst, it was seen communicating with the left main hepatic duct.

Frozen section of the excised CBD stricture was negative for malignancy and thought to be inflammatory due to internal rupture and leaking of hydatid fluid. Reconstruction was achieved by hepaticojejunostomy and the same roux-en-Y loop of jejunum was used to construct the adjacent cystojejunostomy, in view of the large biliary communication. She had an uneventful postoperative recovery.

Histology perplexed us as there was no evidence of hydatid cyst. Instead it showed cyst wall lined by a single layer of low cuboidal epithelium (Figure 3A) with the underlying stroma resembling ovarian stroma (Figure 3B) at places, reported as a multilocular biliary cystadenoma. So, we concluded that this biliary cystadenoma, originating in the segment 4 duct prolapsed into the bile duct and mimicked CBD mass and hydatid cyst. In view of the revised diagnosis, the patient has been advised completion surgery.

**Discussion**

BCA occurs most commonly in middle-aged females. Episodic jaundice has only been reported in five patients in English literature. Obstructive jaundice, although not always present, is the most frequent presenting symptom in patients with extrahepatic cystadenomas. On the contrary, in intrahepatic cystadenomas, biliary obstruction is rarely the chief presenting complaint.

Figure 2: 2A) Segment 4 cyst (a) & distended proximal common hepatic duct (b), 2B) CBD transection reveals membranous lesion proximally (c)

Figure 3: 3A) Cyst wall with low columnar epithelium; 3B) Stroma with ovarian type mesenchyme
There are only eight reported cases of intrahepatic BCA causing obstructive jaundice, either due to protruding polypoidal masses extending into a major duct or from intracystic haemorrhage or intraluminal mucin secretion by the tumour.\textsuperscript{5,7} The most likely cause of episodic jaundice in this patient would have been the protruberant cyst extending into CBD, causing ductal dilatation as reported by Taketomi et al with reactive inflammatory biliary stricture, not described before in literature.\textsuperscript{8}

BCA has two forms: the more common mucinous and the rare serous type. The former is further subdivided by the presence or absence of mesenchymal stroma. Mesenchymal stroma occurs exclusively in women.\textsuperscript{3}

Pre-operative diagnosis can be difficult, but helps to strategize surgery. Biliary cystadenoma has been mistaken for hydatid cyst earlier.\textsuperscript{9} On USG and CECT, BCAs appear as focal lesions with internal septae. Preoperative percutaneous biopsy has no additional value as it rarely produces a definitive diagnosis.\textsuperscript{6} Intraoperative choledochoscopy is useful to assess the ductal system, as exemplified by this case, helping to detect the origin of the intrahepatic component, which could be missed on preoperative imaging.\textsuperscript{9} Frozen sections are not very useful due to the variability in histology of cystadenomas and their inability to rule out cystadencarcinomas.\textsuperscript{11} Careful histopathologic evaluation of the resected specimen, therefore, constitutes the only safe diagnostic modality.\textsuperscript{6}

BCA has a high rate of recurrence and a potential for neoplastic transformation in approximately 10% of cases.\textsuperscript{12} Cystadenoma without mesenchymal stroma is known to be more aggressive, especially in men.\textsuperscript{3} In the past, treatment of BCA has included aspiration, marsupialization, internal drainage and partial excision. The main concerns of these methods are local recurrence, malignant transformation and misdiagnosis of cancer. The ideal treatment should be complete excision of the tumour which includes formal liver resection or wide local excision, as contemporary hepatic surgery has minimal morbidity and mortality.\textsuperscript{13} Follow-up is conducted best by performing abdominal US or CT scan at 6-month intervals for the first postoperative year and then annually.\textsuperscript{12}

Intrahepatic BCA can present with episodic surgical jaundice. Biliary cystadenoma should be suspected when radiologic imaging studies suggest a multilocular cystic hepatic lesion, especially in a woman.\textsuperscript{14} Histopathological examination establishes definitive diagnosis. To the best of our knowledge, this is the first case of BCA with an associated biliary stricture.
Pancreatic hydatid cyst masquerading as cystic neoplasm of pancreas

Introduction

Hydatid disease is a serious public health problem in endemic areas. Most common site of involvement is liver. However, extrahepatic sites such as lung, spleen, bone, kidneys and retroperitoneum have been described. Primary pancreatic lesions are very rare for hydatid disease with an incidence of less than 1%. The diagnosis is challenging and rarely made before surgery as the presenting symptoms and radiological findings may be similar to other commonly encountered cystic lesion of pancreas. We share our experience in a case of pancreatic hydatidosis, masquerading as a cystic neoplasm of the pancreas.

Case Report

A thirty years old lady presented with a history of lump abdomen for the last two months. This was associated with dull aching pain in the epigastric region. There was history of progressively increasing jaundice which was associated with itching and clay coloured stools. History of weight loss and anorexia was present. On examination, she was found to be deeply icteric. Her abdominal examination revealed a 10 x 8 cm globular lump in the epigastrium moving freely with respiration which was not continuous with liver dullness. The clinical diagnosis of surgical jaundice with a palpable gall bladder was made.

On investigation, her liver function tests revealed a total bilirubin of 13.8 mg/dl (normal- 0.8-1.0 mg/dl) with direct fraction of 11.0 mg/dl. Liver enzymes were mildly elevated (alanine transaminase-56 IU/L, aspartate transaminase-72 IU/L). The alkaline phosphatase was elevated to 2331 IU/L (normal 80-240 IU). Ultrasonography of the abdomen revealed mild hepatomegaly, distended gall bladder and dilated common bile duct. Magnetic resonance cholangio-pancreatography (MRCP) demonstrated a cystic swelling in close relation with the lower end of common bile duct with dilated common hepatic duct and intra hepatic biliary radical dilatation (Figure 1). The gall bladder was distended and MRCP suggested a diagnosis of type III choledochal cyst. On endoscopic ultrasound, it was diagnosed as a lesion arising from head of pancreas most probably a serous cystadenoma.

On laparotomy, there was an 8 x 6 cm cystic lesion arising from the head of pancreas which was compressing the distal common bile duct. Intra operatively a diagnosis of cystic neoplasm of head of pancreas (Figure 2) was made and a Whipple’s pancreaticoduodenectomy was done. Patient had an uneventful post operative recovery. On cut section of the specimen, the cyst had a thick wall (Figure 3) with clear fluid and no daughter cysts. The cyst was communicating with the main pancreatic duct and the common bile duct was separately...