Case Report

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Episodic Biliary Obstruction: Intrahepatic Biliary Cystadenoma

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Abstract

Biliary cystadenomas are rare, benign but potentially malignant, multilocular, cystic neoplasms of a biliary origin. They often present with non-specific symptoms. We present a rare case of an intrahepatic biliary cystadenoma causing luminal obstruction to the bile duct.

A 32-year-old female with obstructive jaundice was evaluated and diagnosed as having a cystic lesion in the liver. With a preoperative differential diagnosis of a hydatid cyst, she underwent laparotomy and enucleation of the lesion. Intraoperatively, the tumor was found to be extending into the bile duct, which was occluding the lumen. This was excised and a bilioenteric anastomosis was done. Her postoperative period was uneventful and she was anicteric on follow-up.

Biliary cystadenoma should be considered a differential diagnosis when radiologic imaging studies suggest a multilocular cystic hepatic lesion. Presence of jaundice in such patients should raise the suspicion of an intrabiliary component, the omission of which can lead to avoidable postoperative morbidity.

Key words: *Cystadenoma, jaundice, liver, mucinous*

Introduction

Biliary cystadenomas are rare, solitary, nearly always mucinous, multilocular, cystic tumors that can arise within the liver, extrahepatic bile ducts, or gallbladder [1]. The majority (80%) arise from the intrahepatic ducts and are usually asymptomatic [1,2]. Fewer than 100 reports of intrahepatic biliary cystadenomas have been in medical literature, of which only 8 have presented with obstructive jaundice [3-5]. In this case report, we present a patient with intrahepatic biliary cystadenoma (BCA) extending into the common bile duct, presenting as obstructive jaundice.

Case Report

A 32-year-old female presented with abdominal pain and recurrent episodes of jaundice for the past one year. Physical examination was unremarkable, apart from icterus. Her blood investigations were normal, apart from her liver function tests which showed mild elevation of total bilirubin of 3.6 mg/dL; direct component of 2.1 mg/dL. Her Serum CA 19-9 was 28.9 U/mL. Echinococcal serology was equivocal. MRI showed a well-defined cystic lesion (17.8 cm * 11.4 cm). Hypointensity on T1 and hyperintensity on T2 were seen in segments 5, 6, and 7 of the liver with few thin internal septae, suspicious of a hydatid cyst. There was no obvious intraluminal mass lesion in the bile duct observed on MRI (Figure 1). Therefore, she was planned for exploration and resection of the hydatid cyst. A non-anatomical liver resection of the lesion was done, and on examination of the distal cut end of the bile duct, there was a palpable mass extending onto the common hepatic duct. The bile duct was explored and a bile-stained membranous mass was noted within it. This was extricated from the bile duct, with a check choledochoscopy of the proximal biliary tree up to the second order division being done, which was found to be normal (Figure 2). Reconstruction was achieved by hepaticojejunostomy with a roux-en-Y loop of the jejunum. She had an uneventful postoperative period. Histopathology revealed the lesion to be a cyst lined by a single layer of low cuboidal epithelium, with the underlying stroma resembling ovarian stroma in places, reported as a multilocular biliary cystadenoma. The patient was well and anicteric on follow-up.

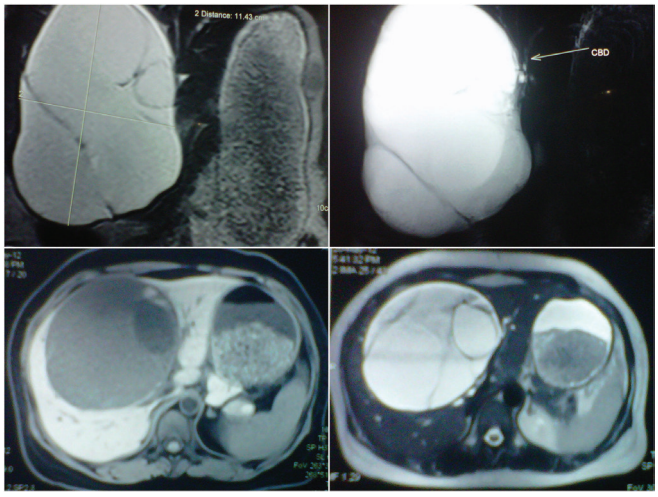

Figure 1. MRI of intrahepatic biliary cystadenoma.

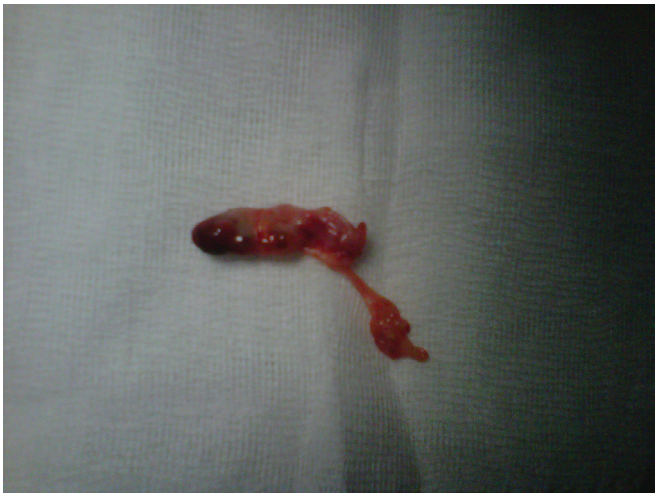

Figure 2. Intraluminal tumor cast extricated from bile duct.

Discussion

BCA occurs most commonly in middle-aged females [1,2]. Episodic jaundice has only been reported in eight patients in English literature [3]. Obstructive jaundice, although not always present, is the most frequent presenting symptom in patients with extrahepatic cystadenomas [2,6]. On the contrary, in intrahepatic cystadenomas, biliary obstruction is rarely the chief presenting complaint [2,6]. 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 hemorrhage or intraluminal mucin secretion by the tumor [3]. The most likely cause of episodic jaundice in this patient would have been the protuberant cyst extending into CBD, whereby causing ductal dilatation, as reported by Taketomi et al.[7] 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 [1,2,7-9]. Pre-operative diagnosis can be difficult, but helps to strategize surgery. Biliary cystadenoma is commonly mistaken for a hydatid cyst, especially in endemic areas like ours. On USG and CECT, BCAs appear as focal lesions with internal septae [1,2,3,10]. A preoperative percutaneous biopsy has no additional value, as it rarely produces a definitive diagnosis [1,2,3,10]. Intraoperative choledochoscopy is useful to assess the ductal system, as exemplified in our case [10]. Laboratory investigations may be helpful for

the differentiation between biliary cystadenoma and an infective cyst based on the presence of leukocytosis, positive amoebic and echinococcal serology [1,2,7,9]. Elevated cholestatic parameters are secondary to obstruction or compression of the biliary tree [1,2]. Elevated levels of CA 19-9 (serum and cyst fluid) have been suggested by some authors to distinguish cystadenoma from other cystic lesions [10]. Frozen sections are not very useful due to the variability in histology of cystadenomas and their inability to rule out cystadenocarcinomas [1,2,10]. Careful histopathologic evaluation of the resected specimen, therefore, constitutes the only safe diagnostic modality [1,3,10]. BCA has a high rate of recurrence and a potential for neoplastic transformation in approximately 10% of cases [1,7,10]. Cystadenoma without mesenchymal stroma is known to be more aggressive, especially in men. 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 tumor, which includes formal liver resection or wide local excision [1-3,10]. Follow-up is conducted best by performing abdominal US or a CT scan at 6-month intervals for the first postoperative year and then annually [3,11].

Conclusion

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 women. Histopathological examination establishes definitive diagnosis. Due to the reported malignancy potential, radical surgery such as wide local excision of the lesion or hepatic resection is recommended to minimize the risk of local recurrence. In all patients with intrahepatic biliary cystadenoma, the extrahepatic and intrahepatic biliary tracts should be thoroughly evaluated intraoperatively to ensure there are no residual tumor thrombi which could lead to undue postoperative morbidity.

Conflict of interest statement

The above doctors have no conflicts of interest or financial ties to disclose.

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