CASE REPORT PEER REVIEWED | OPEN ACCESS

Biliary cystadenoma presenting as hilar intraluminal cyst causing episodic biliary obstruction

Saravanan J., Jeswanth Sathyanesan, Ravichandran Palaniyappan

ABSTRACT

Introduction: Biliary cystadenomas are rare benign cystic lesions of biliary origin with potential of malignancy. Mostly occurring in women (90%) with non-specific symptoms. Case Report: We presented a very rare case of biliary cystadenoma in hilar region causing episodic biliary obstruction. A 60-year-old female with episodic obstructive jaundice was evaluated and diagnosed as having cystic lesion near the hilum of liver. With differential diagnosis of biliary cystadenoma or hydatid cyst, she underwent laparotomy. Intraoperatively the cystic lesion was found in the segment IVb with communication into the hilar region as pedunculated polypoid lesion, which was occluding the lumen causing biliary obstruction. The cystic lesion in segment IVb was excised and the communicating polyp with the hilum was removed along with T tube drainage of common bile duct. Her postoperative period was uneventful and T Tube was removed during the follow up. She was symptom free and anicteric during the follow up period. Conclusion: Biliary cystadenoma should be suspected when radiologic imaging studies suggest a multilocular cystic hepatic lesion, especially in middle aged women.

Keywords: Biliary cystadenoma, Hydatid cyst, Liver cyst, Obstructive jaundice

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Received: 31 December 2015
Accepted: 05 February 2016
Published: 08 March 2016

How to cite this article

Saravanan J, Sathyanesan J, Palaniyappan R. Biliary cystadenoma presenting as hilar intraluminal cyst causing episodic biliary obstruction. Int J Hepatobiliary Pancreat Dis 2016;6:10–13.

Article ID: 100047IJHPDSJ2016

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doi:10.5348/ijhpd-2016-47-CR-3

INTRODUCTION

Biliary cystadenomas are rare benign cystic lesion of liver, that can arise within the liver, extrahepatic bile duct, or gallbladder [1, 2]. Although these lesions are believed to arise from the biliary tree, the exact mechanism of their formation is unknown. To date, less than 200 cases have been reported in medical literature, of which only

nine have presented with obstructive jaundice till date [2–5]. In this rare case report, we present a patient with intrahepatic biliary cystadenoma communicating to hepatic hilum causing luminal obstruction.

CASE REPORT

A 60-year-old female was presented with abdominal pain with episodes of jaundice for six months. Apart from jaundice, physical examination was unremarkable. Laboratory investigations showed cholestatic liver profile with normal white blood cell count and inflammatory marker levels. Ultrasound scan (USS) of the abdomen revealed large cystic lesion in seg 4 of liver with multiple septations communicating with hepatic duct and there was a floating debris with smooth narrowing distal to CD-CHD junction. Her Serum CA 19-9 was 14.9 U/mL. Echinococcal serology was equivocal. MRI scan with MRCP scan abdomen revealed mixed signal intensity lesion with cystic lesion situated in 4b segment of liver showing calcification and septations extending up to porta hepatis with dilated IHBR and EHBR (Figure 1). Possibility of biliary cystadenoma/hydatid cyst with probable rupture into bile duct considered. She underwent an endoscopic retrograde cholangiopancreatography (ERCP) with sphincterotomy. Cholangiogram showed filling defect at hilum. Following the procedure, the patient was well without any symptoms. For definitive management she was taken up for surgery. thick walled cystic lesion of size 6*5 cm attached to inferior surface of seg 4b of liver was excised completely. The cyst had a communication with the bile duct at the hilum. The polypoid lesion was extricated from bile duct (Figure 2) and the communicating rent was sutured with 3-0 vicryl along with T-Tube drainage of common bile duct. The T-tube was removed six weeks later, after confirming absence of a bile leak. During follow-up she was anicteric and symptom free.

DISCUSSION

Biliary cystadenomas are potentially malignant neoplasms occurring predominantly in middle-aged women [1]. 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 [2–6]. Till date only nine reported cases of biliary cystadenomas of intrahepatic origin causing obstructive jaundice, either due to intraluminal mucin secretion by the tumor or from intracystic hemorrhage or by protruding polypoid masses extending into a major duct has reported. In this case, it was due to protruding polypoid mass from the segment IVb cystic lesion. The most recent WHO classification defined the hepatic mucinous cystic neoplasms as a cyst-forming epithelial neoplasm, usually showing no communication with the bile ducts, composed of cuboidal to columnar, variably mucin-producing epithelium, and associated with ovarian-type subepithelial stroma [7]. Commonly reported symptoms include vague abdominal pain (up to 90%) and nonspecific mass symptoms (>30%) [8]. The diagnosis of biliary cystadenoma require a high level of suspicion. Diagnosis of biliary cystadenomas is often possible on an ultrasonography, CECT scan, and MRI scan ofabdomen [9]. On ultrasound, cystic nature of the lesion with multiple loculi, septations, and internal echoes, with papillary projections is typical. The cyst is well-demarcated and thick-walled, globular or ovoid with a non-calcified wall. Doppler study may show the vascular flow within the lesion. On a CT scan, the tumor appears as low-density areas with focal enhancement after contrast administration. The septa and the mural nodules may be visible. An irregular thickness of the cyst wall, presence of mural nodules, or papillary projections indicates the possibility of a malignancy [7–9]. Hypervascularity of mural nodules on CT scan also suggests malignancy [10]. MRI scan is able to characterize the nature of fluid within

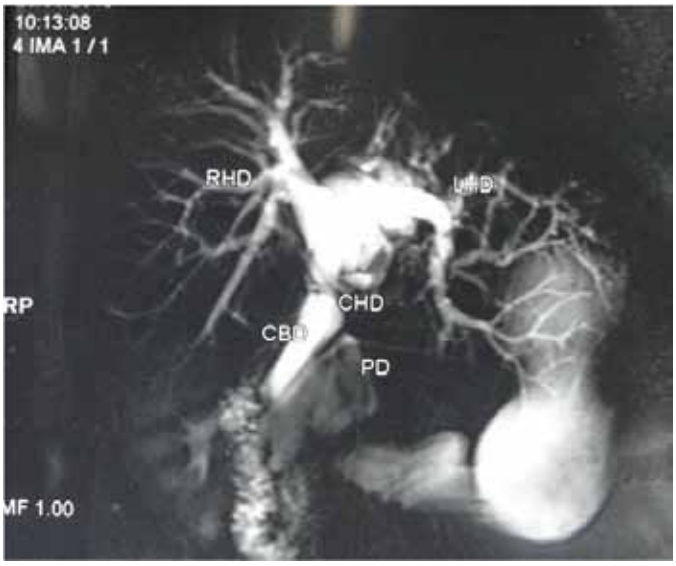

Figure 1: Magnetic resonance imaging scan of intrahepatic biliary cystadenoma.

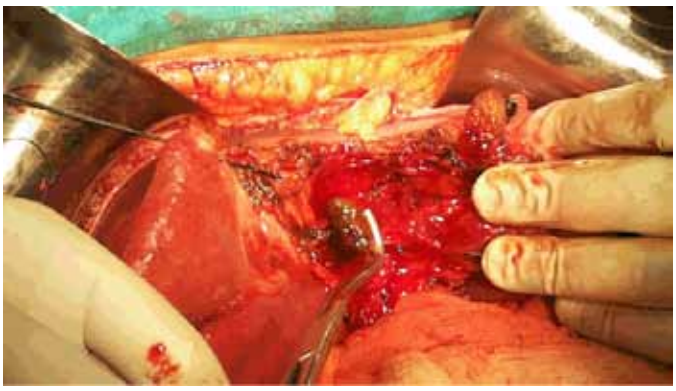

Figure 2: Intraluminal cystic lesion from tumor extricated from bile duct.

the cyst, that is, blood versus mucin. Despite all the aforementioned radiological features, the preoperative radiological diagnostic accuracy may be as low as 30%. A preoperative percutaneous biopsy has no additional value, as it rarely produces a definitive diagnosis. Pre- and intraoperative diagnosis of biliary cystadenomas and cystadenocarcinomas can be very difficult and it can be safely done only after histopathologic examination. In addition, it is essential to differentiate cystadenoma from liver hydatid cyst especially in endemic areas like us since both imaging features are similar, as in our patient. Elevated CEA and CA 19-9 in the serum or the cystic fluid aids in diagnosis, and follow-up of the patient. However, there are no definitive tumor markers found with high diagnostic accuracy. Complete resection is advised as there is risk of malignant transformation. Thomas et al. [10] reported that tumor recurrence occurs in two-thirds of the patients, who have only local or pericystic excision. In contrast, only 10% of the patients, who have hepatic lobectomy, hemihepatectomy or radical excision of cystadenoma with a rim of normal tissue with a diameter of 2 cm, require further surgery for recurrence. In our case wide local excision of the cyst in the segment 4b done along with cholecystectomy with removal of the protruding mass into the bile duct. The rent in the bile duct sutured with 3-0 vicryl along the T-tube drainage of common bile duct. The T-tube was removed four weeks later, after confirming absence of a bile leak. Patient is free of biliary sequelae or other complications in the follow-up period.

CONCLUSION

The diagnosis of biliary cystadenoma requires a high degree of suspicion. Biliary cystadenoma should be suspected when radiologic imaging studies suggest a multilocular cystic hepatic lesion, especially in middle aged women. Histopathological examination establishes definitive diagnosis. Intrahepatic BCA can present with episodic surgical jaundice. They are best treated radically as they have a definite potential to transform into malignant tumors in nearly 10%.

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Author Contributions

Saravanan Janakiraman – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published
Jeswanth Sathyanesan – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Ravichandran Palaniyappan – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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