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CASE REPORT

Bile Duct Hamartoma Mimicking Liver Metastasis in Suspected Porcelain Gallbladder: a Case Report

Gautham Krishnamurthy[1] · Harjeet Singh[1] · Sravya Deepika Ganti[1] · Ganga Ram Verma[1]

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Introduction

Bile duct hamartoma, also called as von Mayenburg complexes, are rare hepatic lesions arising due to a disturbance in the ductal plate remodeling during embryogenesis. [1] The incidence is estimated to be 0.69 to 5.6% in autopsy series. [2, 3] Solitary multicystic bile duct hamartoma are extremely rare and till 2014 only ten cases have been reported in the English literature. [4] We report a case of incidental solitary multicystic biliary hamartoma in a patient undergoing laparoscopic cholecystectomy for suspected porcelain gallbladder.

Case Report

A 45-year-old female presented with bilateral flank pain and mild right hypochondrial pain for 4 months. She was detected to have bilateral renal calculi and porcelain gallbladder on sonography and non-contrast computed tomography (Fig. 1a). Clinical examination was unremarkable. On reevaluation with ultrasonography, the presence of porcelain gallbladder was confirmed along with tiny renal calculi. Hemogram, liver function tests, and coagulation profile were normal. Patient was planned for day care laparoscopic cholecystectomy. Intraoperatively, a 3 × 2 cm non-contiguous nodular lesion was noted in segment 4b adjacent to the gallbladder (Fig. 1b). The rest of the liver was normal. There was no evidence of ascites or peritoneal nodules. In view of the association of carcinoma gallbladder in the porcelain gallbladder, the possibility of metastasis was considered. However, the frozen section of the lesion revealed multiple cystic spaces lined by cuboidal lining epithelium. Dystrophic calcification was noted in its fibrotic wall. Overall features were suggestive of bile duct hamartoma. Cholecystectomy was completed laparoscopically. There was a large stone occupying the entire gallbladder (Fig. 1c). Cut section of the specimen did not reveal nodularity or mucosal thickening. Therefore, no frozen section of the gallbladder specimen was done. Final histopathological examination of the liver lesion confirmed the diagnosis of bile duct hamartoma chronic cholecystitis of gallbladder specimen (Fig. 2). Patient is doing well after 1 year of surgery.

Discussion

Bile duct hamartoma is usually detected incidentally during imaging for other indications, autopsy, or surgery. [2] They may be detected as an isolated lesion or in association with Caroli's disease, congenital hepatic fibrosis [1], and polycystic disease of the kidneys [3] but have never been reported in association with chronic cholelithiasis. They are characterized by small multiple nodules measuring up to 1.5 cm in size. They appear as well-circumscribed whitish nodules or subcapsular plaques involving both lobes of the liver, hence can mimic hepatic metastasis preoperatively during diagnostic laparoscopy or during surgical exploration. [1, 2, 5] Solitary multicystic bile duct hamartoma is extremely rare and known to occur adjacent to the fissure of the falciform ligament and protrude from the liver [6] as in index case.

✉ Ganga Ram Verma
grvermapgi@gmail.com

Gautham Krishnamurthy
k.gautham@gmail.com

Harjeet Singh
harjeetsingh1982@gmail.com

Sravya Deepika Ganti
deepika972@gmail.com

[1] Department of General Surgery, Post Graduate Institute of Medical Education and Research, Sector 12, Chandigarh 160012, India

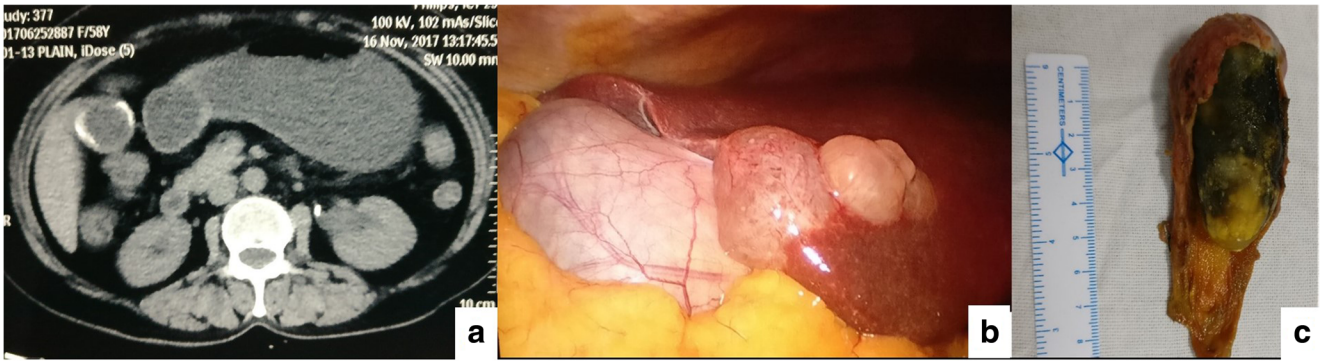

Fig. 1 **a** Plain computed tomography showing calcification appearing to involve the gallbladder wall. No SOL are noted in the adjacent liver. **b** Intraoperative photograph showing non-contiguous multinodular lesion in segment 5 liver. Rest of the liver surface was normal. **c** Cut section of cholecystectomy specimen showing a large gallstone occupying the entire gallbladder lumen. Gallbladder wall and mucosa were normal

When in multiple, they are uniform in size and distribution, unlike liver metastases which are of varying sizes and irregular distribution. [6] The presence of clusters of dilated bile ducts embedded in fibrous stroma confirms the diagnosis [7]. Excision of the lesion is curative.

On contrast-enhanced-computed tomography, they appear as multiple small hypodense nodules distributed throughout the liver. On magnetic resonance imaging, they appear as hypointense lesions on T1-weighted images and hyperintense lesions on T2-weighted images with no communication with the biliary tree. [8] Important differential diagnosis includes liver metastasis especially in patients with extrahepatic malignant tumors, simple hepatic cyst, peribiliary cyst, Caroli disease, and rarely primary hepatocellular carcinoma with cystic changes [9]. While simple hepatic cysts are usually larger in size (> 1 cm), peribiliary cysts are exclusively seen around the hepatic hilum and larger portal tracts [8, 10]. Presence of biliary communication in Caroli disease differentiates it from bile duct hamartoma. [8] Rim enhancement on gadolinium-enhanced MR images mimics metastasis in the presence of a known primary. [11] In the presence of extrahepatic primary, final exclusion of metastatic lesions relies on liver biopsy or follow-up imaging studies. [9] Since the disease may not be detected on preoperative imaging owing to their small size [5], there are reports of the abandoning the planned procedure on suspicion of metastasis during surgery [1, 5].

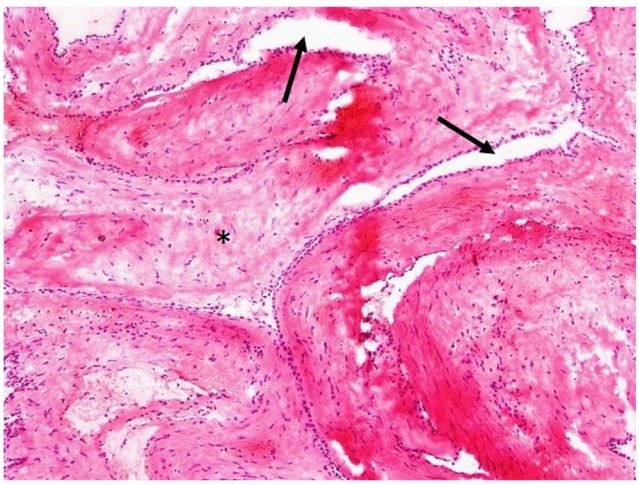

Fig. 2 Microscopic image with H&E stain showing multiple cystic spaces lined by cuboidal lining epithelium (black arrow) with fibrocolllagenous myxoid stroma (*) suggestive of bile duct hamartoma (hematoxylin and eosin, × 40)

Eight cases of biopsy-proven biliary hamartoma in surgically treated malignant cases have been reported in English literature. Summary of these cases is depicted in Table 1. While three of them had a preoperative diagnosis of bile duct hamartoma, five were an intraoperative surprise. It should be noted that the frozen section was performed in the former three cases despite classical preoperative imaging. Though diffuse involvement was the predominant form, solitary lesions were present in two cases. In contrary to solid nodular lesions found in other cases, our patient had solitary cystic lesion making it atypical.

Presence of suspicious lesion, solitary or multiple, during surgery in association with potentially malignant disease of the gallbladder, mandates the careful approach towards laparoscopic cholecystectomy in an endemic region. [19] Intraoperative frozen section is invaluable in differentiating it from metastasis and planning appropriate surgery.

Conclusion

Incidental solitary biliary hamartoma in an extremely rare entity. Preoperative diagnosis is difficult. Incidental detection during surgery raises the suspicion of liver metastasis especially in association with porcelain gallbladder, a premalignant disease. Frozen section clinches the diagnosis. Excision is curative.

Table 1 Summary of cases of surgically treated malignant patients with biopsy proven concomitant bile duct hamartoma published in the English literature

Author, year	Age/sex	Suspected/proven primary	First detection of BDH	Nature of histological confirmation	Nature of lesion*			
					Character	Size	Distribution	Number
Eguchi 2004 [12]	56/male	Hilar cholangiocarcinoma	Intraoperative surprise	Frozen section	Gray-white nodules	3–5 mm	Segments 4 and 6	Multiple
Fritz 2006 [13]	66/female	Esophageal squamous cell carcinoma (biopsy proven)	Intraoperative surprise	Frozen section	White nodular lesion	2–3 mm	Both lobes	Multiple
Nagano 2006 [14]	72/male	Distal cholangiocarcinoma (biopsy proven)	Preoperative imaging (classical)	Frozen section	Gray-white nodules, subcapsular	0.5 cm	Both lobes	Multiple
Mimatsu 2008 [15]	60/male	Esophageal carcinoma (biopsy proven)	Intraoperative surprise	Frozen section	Gray-white nodule	1 cm	Segment 3	Solitary
Lorenzon 2011 [16]	68/male	Synchronous and right colon (biopsy proven)	Intraoperative surprise	Frozen section	Gray-white nodular lesions	Less than 5 mm	Both lobes	Multiple
Bieze 2013 [17]	44/female	Liver SOL (FNH on preoperative imaging)	Intraoperative surprise	Frozen section	Small white lesions	NA	Both lobes	Multiple
Watanabe 2014 [18]	30/female	Carcinoma stomach (biopsy proven)	Preoperative imaging (classical)	Excision and frozen section	White nodule	15 × 10 mm	Segment 4	Solitary
Elsoueidi 2016 [2]	63/male	Pancreas (biopsy proven)	Preoperative imaging (classical)	Frozen section	Gray-white nodules, subcapsular	Largest 2.5 × 4.7 cm. Rest less than 1 cm	Both lobes	Multiple
Index case	45/female	Gallbladder carcinoma in view of suspected porcelain gallbladder	Intraoperative surprise	Excision and frozen section	Multicystic lesion	3 × 2 cm	Segment 4	Solitary

BDH, bile duct hamartoma; *SOL*, space occupying lesion; *FNH*, focal nodular hyperplasia; *NA*, detail not available

*Intraoperative characters. In case size is not mentioned in intraoperative findings, size was derived from preoperative imaging

Compliance with Ethical Standards

Conflict of Interest The authors declare that they have no conflicts of interest.

Informed Consent Informed consent was obtained from the patient regarding the usage of her information for the purpose of publication.

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