Rare Presentation of Malignant Porcelain Gallbladder with Intrahepatic Ductal Calcification and Surgical Obstructive Jaundice

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Introduction

Porcelain gallbladder is a pathological entity characterized by the presence of a brittle calcified gallbladder wall. The prevalence of porcelain gallbladder in cholecystectomy specimens range from 0.06 to 0.8%, with a female predominance [1]. Calcified gallbladder is associated with an increased risk of gallbladder cancer (5–10%). The incidence, however, depends on the pattern of calcification—selective mucosal calcification poses a significantly higher risk of cancer compared to diffuse intramural calcification [2].

Case Summary

A 50 years old lady was admitted with complaints of progressively increasing jaundice, intermittent low grade fever and pruritus for the past 2 months. There was history of progressive weight loss and loss of appetite. No history of abdominal pain, hematemesis, melena or vomiting was present. Apart from icterus, physical examination was grossly normal. Blood investigations revealed total bilirubin 22.0 mg/dl, alkaline phosphatase 183 IU/L, and SGOT 313 IU/L. Computed tomographic scans (CT scan) revealed densely calcified and thickened gallbladder wall (Fig. 1a) with dense intraductal calcification, mainly involving the confluence and central intrahepatic bile ducts (Fig. 1b). There was significant bilateral intrahepatic biliary radical

dilatation and multiple hypodense lesions in bilateral lobes of liver suggestive of metastasis from gall bladder primary. Ultrasonography guided fine needle aspiration cytology from gall bladder mass was positive for adenocarcinoma (Fig. 2). Endoscopic retrograde cholangiopancreatography (ERCP) revealed patent confluence with long subhilar benign stricture extending to supraduodenal common bile duct (CBD) with cholangiogram revealing no evidence of malignancy or intraductal mass. Two stents each of 15 cm × 7 French were placed across the common bile duct stricture to achieve internal biliary drainage. Following ERCP procedure the patient's jaundice was relieved and bilirubin levels normalized. The patient was advised supportive care in view of her advanced disease.

Discussion

Calcification of the gallbladder wall (porcelain gallbladder) is a relatively rare manifestation of chronic cholecystitis. The incidence of such calcification is low, ranging between 0.06%–0.8% of cholecystectomy specimens [1]. It is associated with cholelithiasis in more than 95% of cases reported [1]. Calcification is more common in women (female: male ratio of 5:1) and female predominance is similar to the incidence of cholelithiasis; with a mean age of 54 years (range 38–70 years) [3].

The pathogenesis of gallbladder calcification remains controversial. It is postulated that it can be as a result of obstruction of the cystic duct leading to mucosal precipitation of calcium carbonate salts, which in turn causes bile to stagnate within the gallbladder [1], or this calcification is a dystrophic process resulting from chronic low-grade infection and/or compromised circulation due to impacted cystic duct stones, resulting in hemorrhage, scarring, and

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Fig. 1 **a** CECT revealing dense transmural biconvex curvilinear calcification of gallbladder. **b** CECT showing dense central intrahepatic ductal calcification with peripheral ductal dilatation but no calcification

hyalinization of the wall which in turn provides a matrix for deposition of lime salts [3]. A third pathogenetic mechanism could be chronic irritation of the wall by a stone or other foreign body [4].

Histologically, two types of calcification may be seen. One consists of a continuous broad band within the muscularis, seen radiographically as large plaque-like areas. In the second pattern, numerous calcified microliths are scattered diffusely throughout the mucosa and submucosa, localized within glandular spaces and in the Rokitansky-Aschoff sinuses. This may also be associated with

calcification in the muscularis, producing a radiographic pattern of both granular and plaque-like calcifications [5].

The risk of malignancy within a porcelain gallbladder as previously reported in the literature was extremely high (10–50%) [6, 7]. However modern series have shown a much lower incidence (5–10%) [2, 8]. Two types of calcification patterns were noted—complete intramural calcification and selective mucosal calcification. The incidence of cancer in selective mucosal pattern of calcification is approximately 7% with an odds ratio of 13.89 ($P=.01$). However, there is no consistent association between diffuse intramural calcification and cancer [2].

We report a rare case of porcelain gallbladder that presented as obstructive jaundice. Radiological imaging revealed dense intramural calcification of the gallbladder (Fig. 1a) with cholelithiasis and bilateral intrahepatic biliary radicle dilatation with patent confluence and long benign subhilar stricture extending to supraduodenal CBD. There was significant intraductal calcification involving only the confluence and bilateral central intrahepatic ducts (Fig. 1b). No extra-hepatic or peripheral intrahepatic ductal calcification was seen. Central bile duct calcification could be due to long standing ductal obstruction or due to same dystrophic calcification process, which led to pathogenesis of porcelain gallbladder. Such calcification associated with a porcelain gall bladder is extremely rare and not reported in literature.

Fig. 2 Microphotograph of fine needle aspirate from Porcelain Gallbladder mass showing papillary clusters of neoplastic cells with high nucleocytoplasmic ratio, vesicular nuclei and moderate amount of cytoplasm (H&E $\times 200 \times$ digital magnification)

Conclusion

Porcelain gallbladder is an uncommon entity, even in North Indian Gangetic plains, which has one of the highest incidence

of carcinoma gallbladder in the world [9], and its presentation with obstructive jaundice and associated intra-hepatic ductal calcification has not been reported in literature.

References

1. Polk HG Jr (1966) Cancer and the calcified gallbladder. *Gastroenterology* 50:582–585
2. Stephen AE, Berger DL (2001) Cancer in the porcelain gallbladder: a relationship revisited. *Surgery* 129:699–703
3. Cornell CM, Clarke R (1959) Vicarious calcification involving the gallbladder. *Ann Surg* 149:267–272
4. Ashur H, Siegal B, Oland Y, Adam YG (1978) Calcified gallbladder (porcelain gallbladder). *Arch Surg* 113:594–596
5. Ochsner SF, Carrera GM (1963) Calcification of the gallbladder (“porcelain gallbladder”). *Am J Roentgenol Radium Ther Nucl Med* 89:847–853
6. Berk RN, Armbuster TG, Salzstein SL (1973) Carcinoma in the porcelain gallbladder. *Radiology* 106:29–31
7. Shimizu M, Miura J, Tanaka T et al (1989) Porcelain gallbladder relation between its type by ultrasound and incidence of cancer. *J Clin Gastroenterol* 11:471–476
8. Kwon AH et al (2004) Laparoscopic cholecystectomy in patients with porcelain gallbladder based on preoperative ultrasound findings. *Hepato-gastroenterology* 51:950–953
9. Misra S, Chaturvedi A, Misra NC, Sharma ID (2003) Carcinoma of the gallbladder. *Lancet Oncol* 4:167–176