



Case report

A porcelain gallbladder and a rapid tumor dissemination

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HIGHLIGHTS

- We report a patient with advanced gallbladder porcelain.
- Porcelain gallbladder is a very rare entity found in <1% of cholecystectomies.
- It consists of calcification of the gallbladder wall.
- The rapid progression of cancer in a porcelain gallbladder is more unusual.

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ABSTRACT

Introduction: Porcelain gallbladder is a very rare entity that consists of a calcification of the gallbladder wall, and is associated with carcinoma in 12.5–62% of patients, although recent studies suggest weaker association. **Case report:** We describe an 80-year-old woman who presented with colicky abdominal pain in the right upper quadrant, radiating to the back and associated with vomiting. Physical examination revealed jaundice, murphy's sign was negative.

Hepatic-biliary tract ultrasound revealed porcelain gallbladder, she was referred to the surgical team for a scheduled cholecystectomy. A month later, she presented diffuse abdominal pain. Imaging studies showed a disseminated process affecting liver's segments, capsule, and hilum; and lungs. An aggressive surgical treatment was dismissed, and was referred to the oncology department.

Discussion: There is controversy in the harboring risk of malignancy of the porcelain gallbladder. While it seems that the current data points towards a lower risk of degeneration, it is also demonstrated that patients with gallbladder wall calcifications are indeed statistically at risk of gallbladder cancer. Laparoscopic cholecystectomy has become a safe and efficient approach recommended for patients with gallbladder symptoms directly related or unrelated to gallbladder wall calcifications. In this case, a pathological gallbladder, very quickly evolved into an inoperable tumor with a poor prognosis.

Conclusion: This report heightens that with US evidence of porcelain gallbladder, an urgent CT scan should be carried out to assess an underlying malignancy, and a simple cholecystectomy should be done urgently rather than on a routine elective list to prevent possible malignant change if possible.

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1. Introduction

The porcelain gallbladder is a rare entity with an incidence ranging between 0.06 and 0.08% of cholecystectomies [1]. It consists of calcification of the gallbladder wall. The term originated from the description of a gallbladder wall extensively infiltrated with and replaced by calcium. This process results in a gallbladder with a fragile, brittle consistency and a blue discoloration.

Historically, since its first description in 1929 [2,3], it has been linked to a greater or lesser extent with the development of gallbladder cancer, although today there is much controversy between the relationship of porcelain gallbladder and gallbladder cancer [4].

We present a case of 80-years-old woman diagnosed with porcelain gallbladder.

2. Case report

An 80-year-old female patient with a history of: Hypertension in treatment, osteoarthritis and left crystalline eye extraction, came to the emergency department with a few hours history of abdominal colicky pain in the right upper quadrant, radiating to the

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back and associated with vomiting. A Diagnosis of porcelain gallbladder was made by ultrasound (Fig. 1).

Ultrasound: porcelain gallbladder, minimum intrahepatic bile duct dilatation. No ascites (Fig. 1).

Her symptoms resolved with conservative management, so she was discharged from the emergency department, with a surgical outpatient referral. It was decided to include her on the waiting list for a scheduled cholecystectomy. A month later, she came back to the Emergency Room with a sudden onset of jaundice and diffuse abdominal pain associated with fever and chills. On physical examination, she was jaundiced, her abdomen was soft with tenderness in the right upper quadrant although murphy's sign was negative and she had no signs of peritoneal irritation.

Laboratory tests showed: Total Bilirubin: 4.45 mg/dl; Direct bilirubin: 4.12 mg/dl; AST: 167 U/l; ALT: 181 U/l; γ -GT: 1109 U/l; Alkaline phosphatase: 442 U/l; LDH: 215 U/l; Hb: 8.8 g/dl; Hto: 27.3%; Leucocytes: $11 \cdot 240 / \mu\text{l}$; neutrophils: 77.3%.

Ultrasound: porcelain gallbladder, with bile ducts dilated, especially the left.

The patient was admitted under the Gastroenterology team. An ERCP showed hilar stenosis with a neoplastic character (Fig. 2). A sphincterotomy was performed and a right biliary plastic prosthesis was placed. ERCP brushing biopsies showed moderate cellular atypia.

A CT scan showed: porcelain gallbladder, with calcified walls of different thickness, up to 7.56 mm, associated with hypodense changes of an exophytic growth of a vesicular mass (Fig. 3A), affecting the adjacent liver segments IVb, V, VII, VIII (Fig. 3B) extending to the liver capsule and gastrohepatic ligament, adenopathic process in hepatic hilum, minimum amount of free intra-abdominal fluid, mild dilatation of biliary ducts with plastic stent in situ. Multiple nodules in the middle and lower lungs fields, 10 mm in diameter, well defined. Sub-diaphragmatic lymphadenopathy and a small pleural effusion.

Tumor markers showed CA 19-9: >50000 U/ml, CEA: 724.50 ng/ml. These markers are not useful for the diagnosis, because they are not specific for gallbladder cancer. They often are elevated only when the cancer is in an advanced stage.

A Cholangio-MRI was carried out, confirming multiple liver lesions suggesting metastatic dissemination. All these tests along with the rapid tumor dissemination, made us consider different therapeutic options, dismissing finally aggressive surgical treatment. The disease was inoperable and the patient was referred to the oncology department, who decided to perform ultrasound-

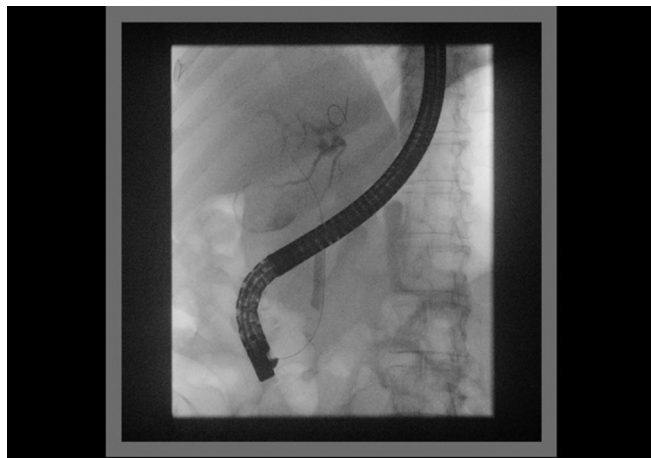


Fig. 2. ERCP showed hilar stenosis with a neoplastic character.

guided liver lesions' biopsy. The multi-disciplinary team conclusion was that this was an invasive gallbladder adenocarcinoma, rather than Bile duct tumor, which are classified as intrahepatic bile duct cancer (cholangiocarcinoma) and extrahepatic bile duct cancer. Because of the advanced age, rapid progression, pathologic features, advanced stage, along with the poor prognosis, the team decided to give palliative treatment, and the patient died in two months of follow up.

3. Discussion

Porcelain gallbladder is rare and has been found in <1% of cholecystectomy specimens. It is more common in women than men, and is most commonly diagnosed in people between 50 and 70 years of age [3]. Although porcelain gallbladder has been regarded as a precancerous lesion, the relationship between gallbladder cancer and the porcelain gallbladder remains unclear.

It is a common perception that gallbladder wall calcifications are associated with a very high risk of harboring gallbladder carcinoma; typically quoted between 13 and 61%. Schnellendorfer confirmed the suspicion that the actual risk is significantly less [5].

However, it still represents a risk factor of developing GBCA. Is the result of a chronic inflammatory process. Microscopically, it may be difficult to pathologically differentiate a primary calcified carcinoma of the gallbladder from a carcinoma arising in a porcelain gallbladder because there are no pathological findings or descriptions for the calcifications observed in the previously reported cases of carcinoma arising in the porcelain gallbladder. Calcification patterns with a broad continuous form along the fibrous stroma may be useful for the differential diagnosis.

Up to 50% of patients with gallbladder carcinoma (GBCA) do not have the diagnosis identified on initial imaging and erroneously proceed to simple cholecystectomy as the first surgical procedure. Further images are necessary in cases of porcelain gallbladder because a more careful consideration of the imaging findings should allow a higher number of patients to be identified preoperatively, and should subsequently translate to more patients being referred for appropriate preoperative workup and definitive oncological management [6].

The gallbladder wall thickening is, however, rarely considered, given the frequency with which the inflammatory conditions of acute and chronic cholecystitis are encountered daily by general surgeons and hepatobiliary surgeons alike [6]. The diagnosis can be challenging because of this common differential and also because the findings on US are generally nonspecific [6].

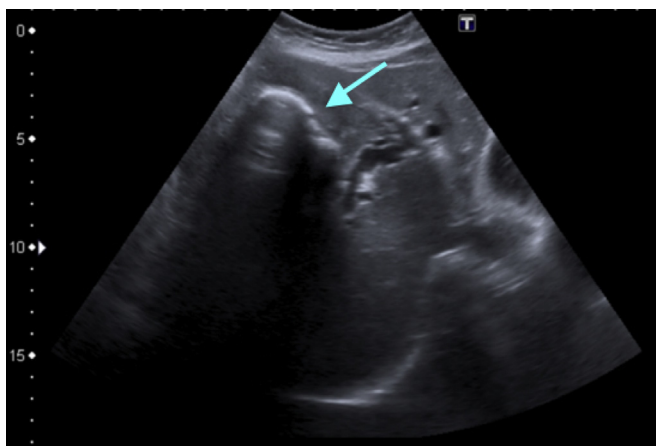


Fig. 1. Ultrasound: porcelain gallbladder with a posterior acoustic shadow. Minimum intrahepatic bile duct dilatation. No ascites.

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