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Case report

# A porcelain gallbladder and a rapid tumor dissemination

Juan-Ramón Gómez-López [a], Beatriz De Andrés-Asenjo [a], Christian Ortega-Loubon [b, *]

[a] Department of General Surgery, University Clinic Hospital of Valladolid, Spain
[b] Department of Cardiac Surgery, University Clinic Hospital of Valladolid, Spain

## 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.


## 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, osteoarthrosis 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

* Corresponding author. Department of General Surgery, University Clinic Hospital of Valladolid, Avenue Ramón y Cajal 3, 47003 Valladolid, Spain. Tel.: +34 983420000.
*E-mail address:* Christlord26@gmail.com (C. Ortega-Loubon).

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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; <gamma>GT: 1109 U/l; Alkaline phosphatase: 442 U/l; LDH: 215 U/l; Hb: 8.8 g/dl; Hto: 27.3%; Leucocites: 11·240/<mu>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 intraabdominal 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-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.

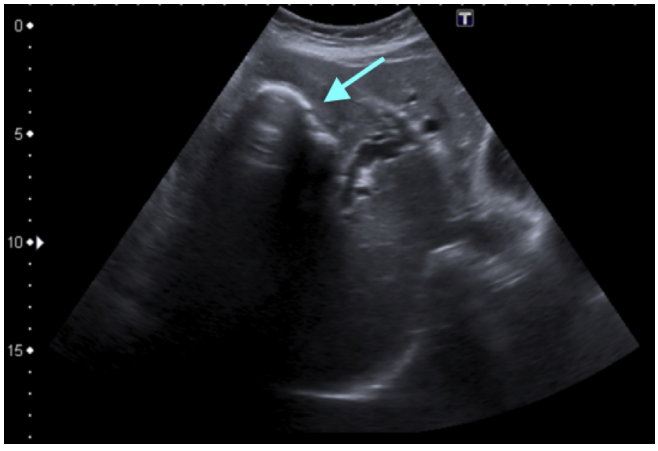


**Fig. 1.** Ultrasound: porcelain gallbladder with a posterior acoustic shadow. Minimum intrahepatic bile duct dilatation. No ascites.

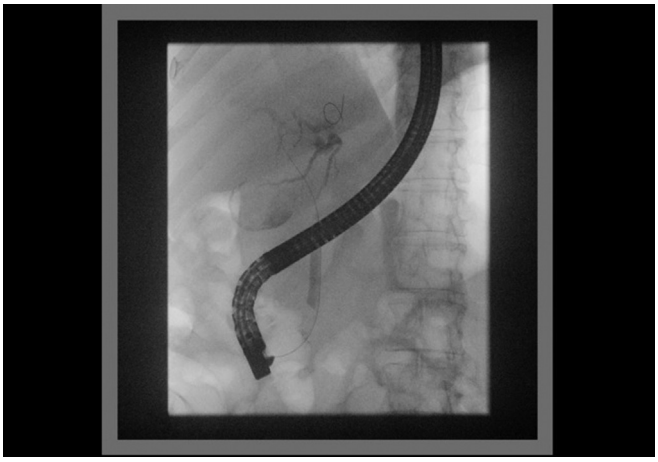

**Fig. 2.** ERCP showed hilar stenosis with a neoplastic character.

## 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%. Schnelldorfer 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].

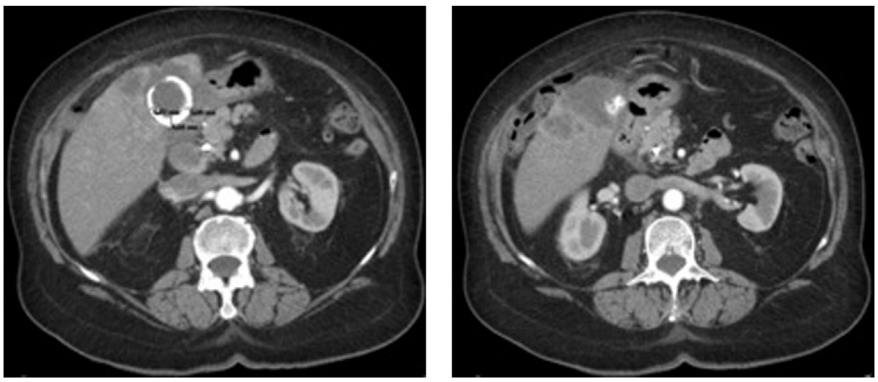


**Fig. 3.** (A) CT showed exophytic growth of a vesicular mass and a calcified wall thickening. (B) Affected liver segments (IV b, V, VII, VIII) extended to the liver capsule, hepatic hilum, and gastrohepatic ligament.

Additional imaging is usually necessary to delineate the nature of gallbladder wall thickening, but unfortunately this is undertaken all too infrequently. Multidetector CT is ideally suited to the careful assessment of the nature of gallbladder wall thickening, and will also frequently document evidence of distant nodal or metastatic spread.

There is emerging evidence that CT findings related to the nature of gallbladder wall thickening can differentiate benign from malignant causes [7] and can also assign T stage with some degree of accuracy preoperatively.

Kim et al. describe 5 distinct patterns of wall enhancement on CT that correlate well with the differentials of GBCA, adenomyomatosis, and acute and chronic cholecystitis (Table 1). And Liang et al. describe some features that might be suggestive of GBCA (Table 2).

The presence of enlarged nodes before any surgery, on the other hand, is more ominous for malignancy, and involvement of nodal groups beyond the hepatoduodenal ligament is viewed as a contraindication to surgery in some patients, as there have traditionally been very few longterm survivors among patients with N2 nodal disease, and this N staging classifies patients as stage IVB disease in the current 7th edition of the AJCC Cancer Staging Manual (http://www.cancer.gov/cancertopics/pdq/treatment/gallbladder/HealthProfessional/page3) [10].

In those few cases where the diagnosis of GBCA is established preoperatively, an accurate assessment of T, N, and M stage is critical to plan treatment and surgery when appropriate.

The presence of symptoms typical for gallbladder cancer and the presence of a gallbladder mass are the only statistical significant predictors for the presence of malignancy. These predictors, however, have only limited clinic value, since the presence of these predictors are stigmatic for advanced gallbladder cancer at which point the chance for cure is frequently limited. Our case was a stage III B with invasion of adjacent liver parenchyma and the presence of right adenopatics hepatic hilum and subdiaphragmatic processes; pulmonary nodules also were suggestive of neoplastic dissemination. For all this, along with rapid disease progression and the age of the patient, we dismissed radical surgery.

**Table 1**
Computed tomography features of gallbladder wall thickening with likely differential diagnosis [7,8].

| Type | Description | Most common diagnosis |
|---|---|---|
| 1 | Heterogeneously enhancing thick one layer | Gallbladder cancer |
| 2 | Strongly enhancing thick inner layer (≥2.6 mm)<br>Weakly enhancing/nonenhancing thin outer layer (≤3.4 mm) | Gallbladder cancer |
| 3 | Borderline pattern | Adenomyomatosis |
| 4 | Weakly enhancing thin inner layer<br>Nonenhancing thin outer layer | Chronic cholecystitis |
| 5 | Weakly enhancing thin inner layer<br>Nonenhancing thick outer layer | Acute cholecystitis |

**Table 2**
Computed tomography features suggesting gallbladder cancer in a series of 26 patients with cholecystitis complicating GBCA matched with patients with simple cholecystitis alone.[9].

| Features suggesting gallbladder cancer |
|---|
| Higher frequency of nodal involvement (65% vs 16.7%) |
| More extensive wall thickness (8.9 mm vs 5.9 mm) |
| Focal irregularity in wall thickness |
| Less distension of gallbladder (volume 71 mL vs 95 mL) |

## 4. Conclusion

This report heightens that with US evidence of porcelain gallbladder, an urgent CT scan should be carried out to assess an underlying malignancy. Further, a simple cholecystectomy should be done urgently rather than on a routine elective list to prevent possible malignant change if there are no co-morbidities.

## Conflict of interest

None.

## Funding

None.

## Ethical approval

Written consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in Chief of Annals of Medicine and Surgery.

## Author contributions

All authors contributed equally in this case report.

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