Contents lists available at ScienceDirect

Annals of Medicine and Surgery

journal homepage: www.elsevier.com/locate/amsu

Case Report Cholecystopathia chronica calcarea (Porcelain gall bladder): A case report from Nepal

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ARTICLE INFO

Keywords: Porcelain gallbladder Calcified gall bladder Laparoscopic cholecystectomy Chronic cholecystitis Case report

ABSTRACT

Introduction and importance: Porcelain gall bladder is an uncommon end-stage modification of chronic cholecystitis, with an incidence ranging from 0.06 to 0.8% along with a plausibility of malignant transformation. *Case presentation:* We present a 55-year-old female presenting with complaints of epigastric and right hypochondriac region pain who underwent prophylactic laparoscopic cholecystectomy after making a provisional diagnosis of calcified gall bladder on a computed tomography workup. On histopathological examination, she was later diagnosed with a porcelain gallbladder devoid of features suggestive of malignant transformation. *Clinical discussion:* Porcelain gallbladder is a cholecystopathological condition in which the gallbladder wall gets calcified, either completely or partially. Though the exact pathomechanism of gallbladder calcification is unknown, it is believed to be due to chronic inflammation. Recent studies have shown that gallbladder calcification is associated with a lower risk of the development of gallbladder cancer. Imaging studies, followed by postoperative histopathological examinations, are used to diagnose the porcelain gallbladder. Though the management of asymptomatic patients is debatable, prophylactic cholecystectomy is the preferred treatment for symptomatic porcelain gallbladder patients.

Conclusion: Individual porcelain GB patients should be addressed based on the presenting condition, whether surgically or via clinical monitoring and follow-up, taking into consideration the advantages and limitations of both treatment modalities.

1. Introduction

Cholecystopathia chronica calcarea, commonly known as "porcelain gall bladder", is a rare end-stage modification of chronic cholecystitis characterized by bluish-hue discoloration and brittle gall bladder wall consistency [1–4]. It has been documented that females outnumber males (5:1), ranging in age from the 3rd decade to the 7th decade [5], with an incidence ranging from 0.06 to 0.8% and a risk of malignancy varying from 5% to 22% [2,6]. As it is a potentially pre-malignant condition with a plausibility of malignant transformation, cholecystectomy remains the favored surgical treatment of choice [2,7,8].

We herein report a case of a laparoscopically managed porcelain gall

bladder without malignant transformation, diagnosed via histopathological examination.

2. Method

This case is reported in accordance with SCARE 2020 guidelines [9].

3. Case Presentation

We present a 55-year-old female, known case of type 2 diabetes mellitus (under medication for 12 years), who presented with a onemonth history of abdominal pain. Pain was localized mostly to the

https://doi.org/10.1016/j.amsu.2022.104947

Received 8 September 2022; Received in revised form 2 November 2022; Accepted 13 November 2022 Available online 17 November 2022

Available online 17 November 2022







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epigastrium and right hypochondriac region and was insidious at onset, on and off, non-radiating, relieved on analgesics, and unrelated to the meal. She denied the history of fever, jaundice, significant weight loss, vomiting, pruritus, altered bowel habits, and a relevant family history of malignancy. She added that she had 2-3 episodes of a similar attack, which used to subside with analgesics taken from a nearby health center. She doesn't drink or smoke, and she has no serious allergies or psychosocial history.

On examination, her vital parameters were within normal limits. Systemic examinations were normal. Abdominal examination showed a soft, symmetric abdomen with no local rise in temperature and mild tenderness in the right hypochondriac region. Murphy's sign was negative. Organomegaly and renal angle tenderness were not evident. Routine baseline laboratory investigation and serology, as well as serum amylase and lipase, were within normal limits. A plain chest radiograph was unremarkable. Ultrasonography of the abdomen and pelvis reveals a contracted GB with an iso-echoic lesion with peripheral calcifications in the gall bladder (GB) fundus area and a similar iso-echoic lesion in the GB neck region. Further, she was also evaluated with plain computed tomography (Fig. 1) of the abdomen, which revealed diffuse wall calcifications involving the gall bladder wall in the fundic region with no focal lesion seen, consistent with a porcelain gall bladder. Evaluation of tumor markers revealed values within the normal limit.

After thorough discussion with the patient, a laparoscopic cholecystectomy was planned. The surgery was carried out by a qualified general and gastrointestinal surgeon in a tertiary care center. Following surgery, gross examination (Fig. 2) revealed a gall bladder measuring 5.5×4 cm with a wall thickness of 0.4 cm. The outside surface appears greyish to bluish-white. Mucosal loss was visible in the lumen. Gallstones were not present. Microscopic examination of gallbladder sections (Fig. 3) revealed extensive fibrosis and hyalinization of the wall, as well as areas of calcification and cholesterol cleft with loss of mucosal lining epithelium. Mild chronic inflammatory infiltrates were present, primarily composed of lymphocytes and plasma cells, with no evidence of cellular or nuclear atypia.

The postoperative period was uneventful, and follow-up of the patient after 7 days and 21 days remains unremarkable.

4. Clinical Discussion

A porcelain gallbladder is an unusual cholecystopathological condition in which the gallbladder wall gets calcified either completely or partially. During this process, it can become brittle and bluish, resulting



Fig. 1. CT abdomen showing a calcified rim of the gall bladder wall (or-ange arrow)



Fig. 2. Gross specimen (cut section) of the gall bladder.

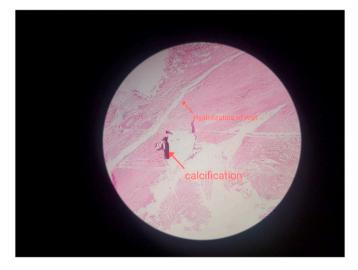


Fig. 3. Histopathological picture showing calcification of the gall bladder along with hyalinization of the gallbladder wall.

in a "porcelain" like appearance [1,2,7,8,10].

The incidence of porcelain gallbladders (in routine cholecystectomy specimens) is less than 1%, and risk factors include female gender, old age, and gallstones. In about 90% of cases of porcelain gallbladder, gallstones are present; therefore, the risk factors for the formation of gallstones and the prolonged presence of gallstones carry a significant risk of gallbladder calcification. The prevalence is highest in the sixth decade of life, with a 5:1 female to male ratio [1]. Our patient is a female patient in her mid-fifties.

The exact pathomechanism of gallbladder calcification is not clearly elucidated. However, it is believed to be due to chronic inflammation that causes bleeding, scarring, and hyalinization of the gallbladder wall. Among the potential etiologies, which include ischemia, inflammation, and dysfunction in calcium homeostasis (such as metabolism and dystrophic calcification), cystic duct obstruction with bile stagnation and persistent gallbladder wall irritation secondary to gallstones is thought to be the primary etiopathology, resulting in mucosal calcium carbonate precipitation [1,11].

Its presentation ranges from selective mucosal calcification to complete calcification of the entire gallbladder wall [1,11]. Gallbladder calcification is associated with the development of gallbladder cancer [1,11–13]. But throughout the assessment, our patient reports no indications of a malignant transformation.

Previously, the incidence of gallbladder cancer in patients with gallbladder wall calcification was reported to be anywhere from 12% to 61% [10], but recent reports have shown the incidence of gallbladder cancer in patients with porcelain gallbladder to be around 6% [10–13]. The incidence of gallbladder cancer is higher in cases of incomplete calcification than in those of extensive calcification [10,11]. Most porcelain gallbladder cases are asymptomatic, but some patients present with pain in the right upper quadrant as in our case and other signs of chronic cholecystitis, and it is frequently detected incidentally on imaging studies [2,4,6,8].

Imaging studies such as plain abdominal X-rays, ultrasound, abdominal computed tomography (CT) scans, and diffusion-weighted magnetic resonance imaging are used to diagnose porcelain gallbladder [1,6]. An ultrasound scan was used to evaluate our patient. After an ultrasound scan revealed peripheral calcifications in the gallbladder fundus and neck region, plain computed tomography was performed, which revealed diffuse calcification of the gallbladder wall and confirmed the diagnosis of porcelain gallbladder by histopathological examination.

Prophylactic cholecystectomy is the preferred treatment for a porcelain gallbladder in patients who are young and fit, symptomatic, or have a selective mucosal type of calcification. The role of preventative cholecystectomy in circumstances other than these is, however, debatable [1,11–13]. A single-incision laparoscopic cholecystectomy has been demonstrated in some publications [14] to be an effective treatment option; however, other studies recommend an open surgical technique due to the gallbladder's technical hurdles due to the stony but brittle nature of the gallbladder during surgery [4]. Because she was presented with intermittent symptoms and upon her own request, our patient underwent a laparoscopic cholecystectomy.

5. Conclusion

Porcelain GB is a rare end-stage chronic cholecystitis variant. Despite studies finding a lower but plausible risk of malignant transformation, a diagnostic workup is to be undertaken and management (either conservative or surgical intervention) relient on the clinical spectrum of the patient at presentation.

Provenance and peer review

Not commissioned, externally peer-reviewed.

Ethical approval

This case report did not require ethical approval from ethics committee.

Sources of funding

All authors declare that this research did not receive any specific grant or funding from funding agencies in the public, commercial, or not-for-profit sectors.

Author contribution

Authors and their contribution are depicted as follows:

1. Samikshya Karki, Sujan Bohara, Gaurab Mainali: Study conception, drafting, revision of the manuscript, and manuscript submission.

2. Pawan Singh Bhat, Binit Upadhaya Regmi, Saugat Khatri: Editing the draft and revision of the content of the manuscript.

3. **Sushil Bahadur Rawal, Srijan Malla:** Complete supervision, critical appraisal of the manuscript, and performed the surgery.

4. All authors have approved the final article for submission.

Conflicts of interest

All authors declare that they have no conflict of interest.

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2. Unique identifying number or registration ID: N/A.

3. Hyperlink to your specific registration (must be publicly accessible and will be checked): N/A.

Guarantor

Dr. Sujan Bohara.

Consent

Written informed 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 this journal on request.

Acknowledgement

The patient depicted in this article is acknowledged by the authors.

Appendix A. Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.amsu.2022.104947.

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