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Case Report

Biliary cystadenoma: Case report with MRI findings and surgical confirmation ☆

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ABSTRACT

Biliary cystadenoma is a very uncommon benign cystic neoplasm involving the liver and the biliary tract. Most common presentations include right upper quadrant pain, nausea, vomiting, obstructive jaundice, and enlarging liver size. It can mimic many more commonly occurring diseases such as hepatic cyst, hepatic abscess, hydatid disease of the liver, and hepatic tuberculosis. Hence it becomes very challenging for physicians to correctly diagnose it due to its rarity and similarity with other conditions. Furthermore, very few pieces of literature guide physicians in correctly identifying the disease. Based on his physical examination and detailed investigation, we present a case report of a 72-year-old female diag-

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nosed with biliary cystadenoma. We hope that this case report will significantly add to the existing literature on this subject

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Introduction

Biliary cystadenoma (BCA), a rare (<5%) benign cystic tumor of the liver, originates from either an aberrant bile duct or directly from primitive hepatobiliary stem cells with a lining of the mucin-secreting columnar or cuboidal epithelium [1–7]. It constitutes around 85% intrahepatic, most commonly in the right lobe in the liver (55%), followed by the left lobe (29%) and rarely bilobed (16%) and extrahepatic being sparse in 15% of cases [1,2]. In 1887, Hueter reported the first case of BCA, then in 1892, the first reported case of BCA resection was done by Keen, with less than 200 cases till 2012 [6,7]. More than 85% of the cases occur in middle-aged (40-50 years) women, with an

incidence rate of 4:1 for males suggesting there might be a hormonal influence [1,3,5,7]. The large multiloculated histologic pattern is further divided into 2 types according to the presence or absence of mesenchymal (ovarian-like) stroma [4,6]. The clinical presentation is primarily asymptomatic. Those who are symptomatic present with mainly abdominal pain and distension (55%-90%) with derangement of liver function in around 20% of the cases [3,7]. It can result in complications such as obstructive jaundice, hemorrhage, cyst rupture, or malignant transformation [2–4,6,7]. Ultrasonography, CT, and MRI remain the primary diagnostic modalities, and tissue diagnosis is required to differentiate between benign and malignant neoplasm and other cystic lesions found in the biliary tract and hepatic region [1,3–5,7]. There is a need to prevent recur-

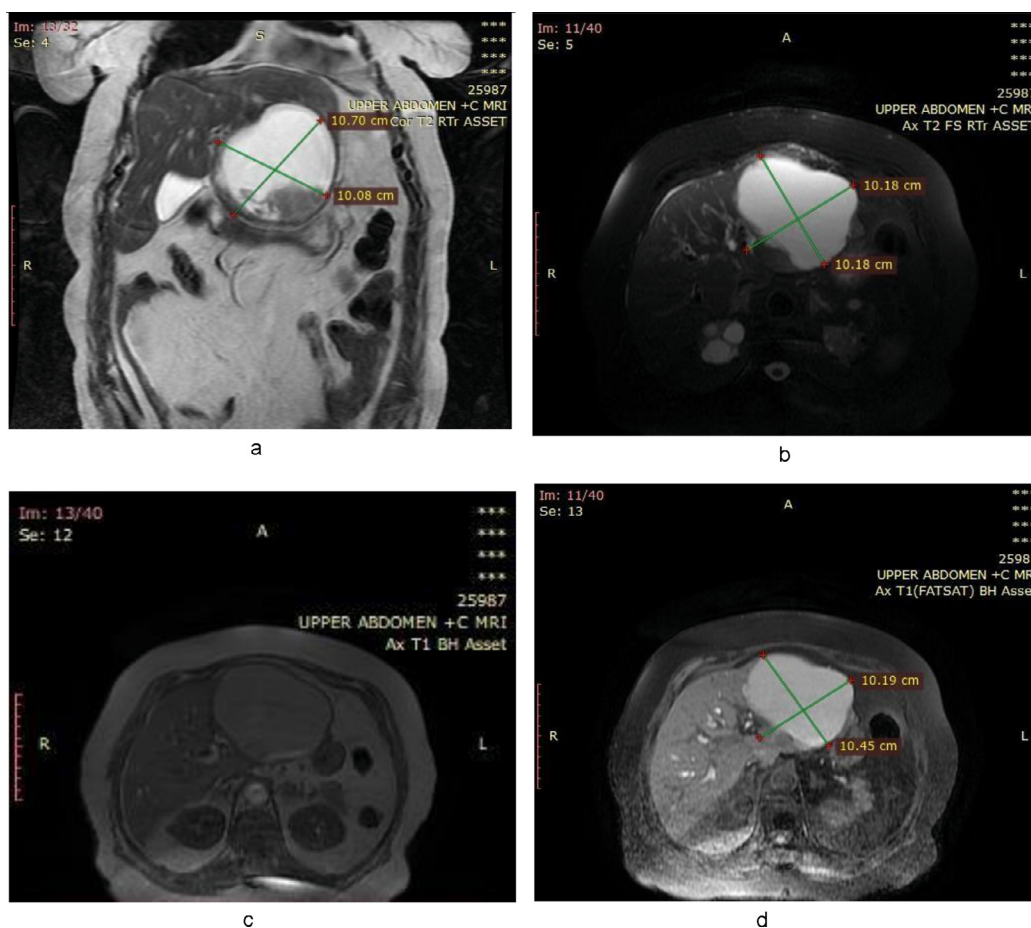


Fig. 1 – (a) coronal sequence precontrast an exophytic large cystic lesion, showing hyperintense T2 measuring 110 × 100 mm in size. The is debridement in its inferior aspect. (b) sequence axial T2 Fat set an exophytic large cystic lesion, showing hyperintense T2 measuring 110 × 100 mm in size. The is debridement in its inferior aspect. (c) sequence T1 fat set precontrast and exophytic large cystic lesion, showing hypointense T1 measuring 110 × 100 mm in size. The is debridement in its inferior aspect. (d) sequence axial Fat set T1 precontrast and exophytic large cystic lesion, showing hyperintense T2 measuring 110 × 100 mm in size. The is debridement in its inferior aspect.

rence and malignant transformation, seen in approximately 30% of the cases which are misdiagnosed or incompletely resected; hence the definite treatment is complete surgical resection [3–5,7]. The postoperative follow-up is imperative to see postoperative surgical complications or risk of recurrence [1–7]. Our case of a 72-year-old female gives insight into the radiological findings and management plan of BCA.

Case report

We report the case of a 72-year-old patient who was admitted by an internist-gastroenterologist due to pain in the upper left hypochondrium associated with abdominal distension, nausea, and occasional vomiting. She also complained of back pain.

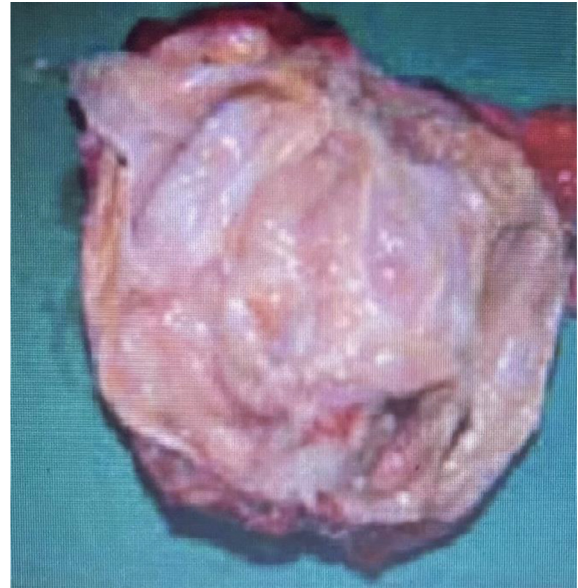
She has a history of hypertension and chronic obstructive pulmonary disease, which are managed with therapy by a cardiologist and pulmonologist.

On physical examination, there was nothing significant. Her gastroenterologist ordered an abdominal ultrasound, which revealed cysts with exophytic septa in the left lobe of the liver and renal cysts. An MRI of the upper abdomen was recommended to visualize the head of cystic changes.

On MRI with contrast and postcontrast study, the findings were:

- There was an exophytic large cystic lesion involving a significant part of the left liver lobe, showing hyperintense T2 and intermediate T1 signal intensity and measuring 110 × 100 mm in size. It showed peripheral contrast enhancement, and there was debridement in its inferior aspect.
- The mass showed fat suppression in the T1 out phase and peripheral nodular contrast enhancement in the post-contrast study.
- Multiple renal cortical cysts were also noted, the largest in the upper aspect of the right kidney, measuring 18 mm in size. A minimal increase in thickness of the left adrenal gland was noted.
- Dilatation of the abdominal aorta was evident.

After receiving the result, a consultation was arranged with the surgeon. Open surgery was recommended, and extrusion of the cystic lesion was planned, involving both the liver's left lobe and the cystic mass and parenchyma in the vicinity.



After surgery, Fig. 3 the mass showed a trabecular, multilocular cystic lesion with thickened wall and no papillary excrescences, polypoid areas, or significant solid components.

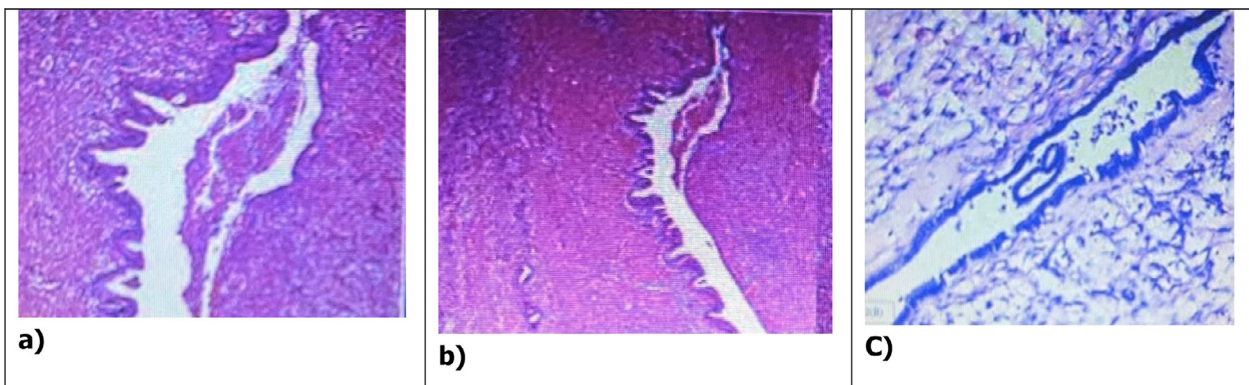
The resected mass showed a trabecular, multilocular cystic lesion with a thickened wall and no papillary excrescences, polypoid areas, or significant solid components.

Fig. 4 Histopathology showed Biliary Cystadenocarcinoma – increased nuclear pleomorphism and chromatin irregularity with increased epithelial cell stratification and tubule-papillary growth compared to biliary cystadenoma.

Figure HP showing Biliary cystadenocarcinoma – Stain: Hematoxylin and eosin (H& E), magnification a. x100, b. x 40 dhe c x 100 showing: Increased nuclear pleomorphism and chromatin irregularity with increased epithelial cell stratification and tubulopapillary growth ascompared with the biliary cystadenoma).

Discussion

We present a 72-year-old woman with biliary cystadenoma, a family of very rare cyst neoplasm of the biliary ductal system



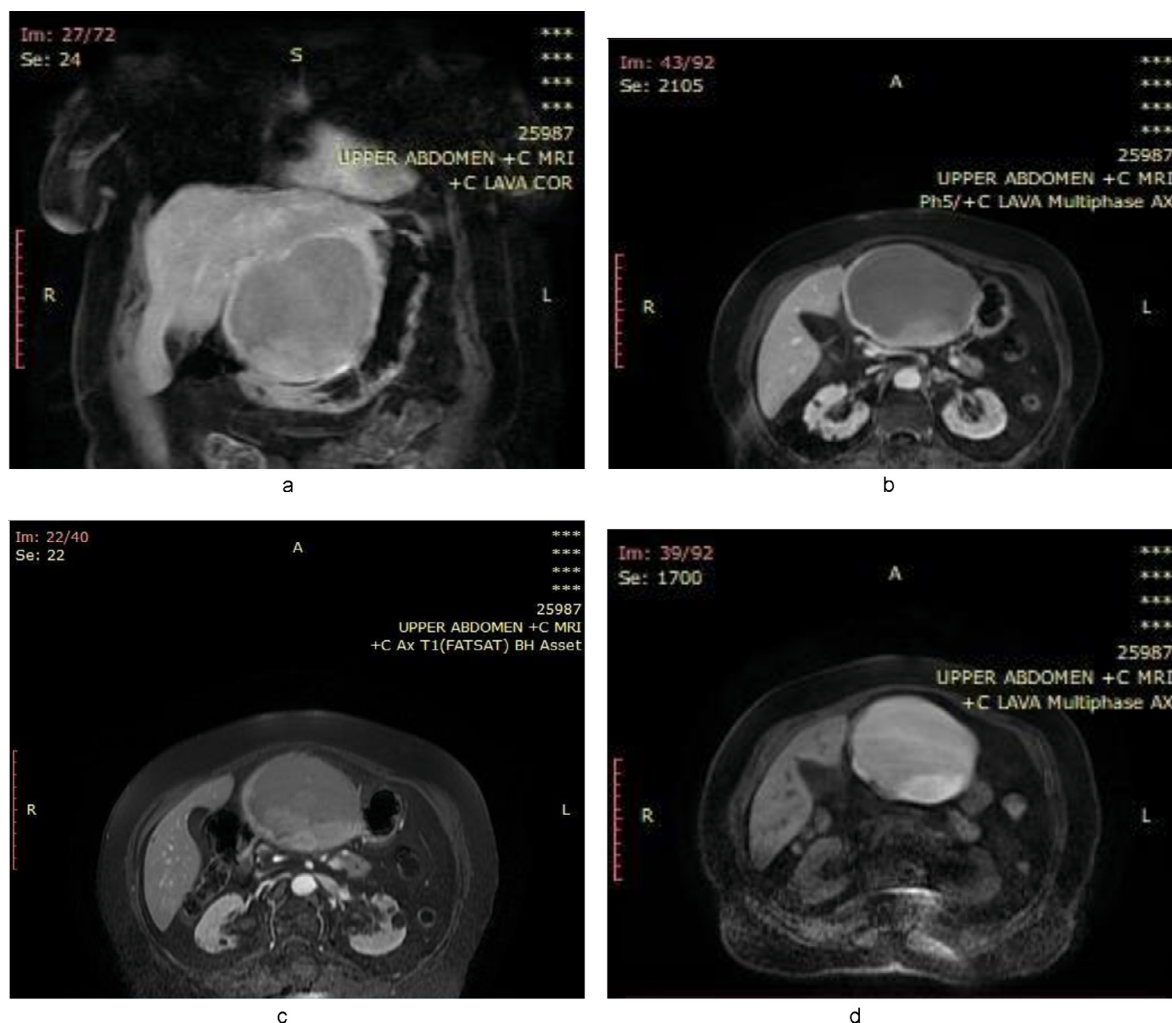


Fig 2 – (a) shows coronal postcontrast lava showing: an exophytic large cystic lesion measuring 110 x 100 mm in size; the mass shows fat suppression in the T1 out phase and peripheral nodular contrast enhancement in the postcontrast study. The debris in its inferior aspect. (b) sequence axial T1 multiphase Ph 5 postcontrast showing an exophytic large cystic lesion measuring 110 x 100 mm in size. It shows peripheral contrast enhancement. The debris in its inferior aspect. The mass shows fat suppression in the T1 out phase and peripheral nodular contrast enhancement in the postcontrast study. (c) sequence T1 postcontrast showing an exophytic large cystic lesion measuring 110 x 100 mm in size. It shows peripheral contrast enhancement. The is debris in its inferior aspect. (d) sequence axial multiphase postcontrast showing an exophytic large cystic lesion measuring 110 x 100 mm in size. It shows peripheral contrast enhancement. The is debris in its inferior aspect. The mass shows fat suppression in the T1 out phase and peripheral nodular contrast enhancement in postcontrast study.

[8]. Biliary cystadenomas are benign and belong to the 5% or less of the hepatic cyst neoplasms [1,3,5–7,9]. In most cases, they are asymptomatic and the diagnosis is made incidentally by imaging or as a trans-surgical finding [8,9]. In our case, the patient had unspecific abdominal symptoms such as pain in the upper left hypochondrium, abdominal distension, nausea, and vomiting. Clinical manifestations include extrinsic compression of adjacent structures (stomach, duodenum, and biliary tree) [8,9]. The most frequent symptoms are (60%) abdominal discomfort, swelling, abdominal pain, palpable abdominal mass in the right upper quadrant, dyspepsia, nausea, vomiting, and weight loss. The compression of the biliary tree may lead to jaundice, biliary colic, and fever.

Furthermore, when there is compression of the inferior vena cava or hepatic vein, patients can present abdominal swelling or ascites [8,9]. Clinical signs were unspecific at the physical examination. Some literature reviews affirm that, in most patients, laboratory studies are normal; however, 20% can have high liver functions test [7], and high levels of CA 19.9 in blood tests are consistent with this neoplasm [5]; in our case laboratory test were not valuable for the diagnosis.

The diagnosis of biliary cystadenoma can be incidental. Abdominal ultrasound and computed tomography are the imaging studies most frequently used, with a sensitivity of 90% and 87%, respectively. We must consider the two studies complementary to the accuracy of diagnosis [6,8,9]. Tomography

is less valuable to identify septations. However, it is superior in detecting lesion size and extension [8]. Magnetic resonance imaging has been proven beneficial to differentiate biliary cystadenoma from other cystic liver lesions and when we need to rule out biliary tree lesions with a magnetic resonance cholangiopancreatography [9]. Magnetic resonance can characterize the cyst fluid content by the variation of the signal intensity and the protein content (T1) and identification of high-intensity cyst septation (T2) [7–9]. In our patient, MRI has shown T1 and T2 components and provided exact cyst localization, improving the posterior surgical planning Figure 1 and 2.

Regardless of whether the biliary cystadenoma is a benign lesion, the management must be surgical. The complete resection with a wide margin of normal liver is the treatment of choice [3,6–9]. Although biliary cystadenoma has a high potential for malignant transformation, the prognosis is excellent when appropriate surgical management is performed [9]. Also, cystadenoma and cystadenocarcinoma cannot be differentiated in the pre-surgical period with radiological criteria [9]. Other conservative therapies are cyst aspiration, laparoscopic fenestration. However, there is a high risk of recurrence between 80% and 90%, even 100% [8,9].

The histopathologic report shows a solitary, multilocular cystic lesion with fluid content [7,9]. Microscopically it is composed of three layers: a biliary epithelium, cellular stroma, and collagenous connective tissue [9]. The distinction between cystadenoma and cystadenocarcinoma is the presence of increasing malignant epithelium, such as multilayered epithelium, frequent mitotic figures, loss of polarity, and nuclear pleomorphism [7,8]. In our case, a complete surgical resection was performed, and the histopathological diagnosis was a cystadenoma (Figures 1, 2 and Fig. 3).

In large case series, the survival rate is greater than 90% with 18 years of follow-up [7]. Abdominal ultrasound or computed tomography is recommended to monitor the prognosis in these patients [3].

Conclusion

A high index of suspicion is required to diagnose the rare disease of biliary cystadenoma. Although benign, it can lead to dangerous complications such as malignant transformation, obstructive jaundice, massive enlargement, rupture, secondary bacterial infection, bleeding, and involvement of other vital structures nearby, such as inferior vena cava. Hence it is imperative for physicians to correctly and quickly diagnose this condition. Prompt management and successful removal of this tumor are essential to prevent adverse events and im-

prove patients' quality of life. This case report provides a comprehensive presentation of the disease, guiding physicians in adequately diagnosing and treating this relatively rare phenomenon known as biliary cystadenoma.

Patient consent

I, give my consent for the publication of identifiable details, which can include photograph(s) and/or videos and/or case history and/or details within the text (“Material”) to be published in the Radiology Case Reports and Article “Biliary cystadenoma: case report with MRI findings and surgical confirmation”.

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