Received: 18 August 2021 | Revised: 3 February 2022 | Accepted: 16 February 2022

DOI: 10.1002/ccr3.5531

CASE REPORT

Clinical Case Reports Open Access WILEY

Extrahepatic mucinous biliary cystadenoma: A rare cause of gastric outlet obstruction

Arkadeep Dhali[1] | **Sukanta Ray**[1] | **Gopal Krishna Dhali**[2] | **Sujan Khamrui**[1] | **Ranajoy Ghosh**[3]

[1]Department of GI Surgery, IPGME&R, School of Digestive & Liver Diseases, Kolkata, India

[2]Department of Gastroenterology, IPGME&R, School of Digestive & Liver Diseases, Kolkata, India

[3]Department of GI Pathology, IPGME&R, School of Digestive & Liver Diseases, Kolkata, India

Correspondence
Arkadeep Dhali and Sukanta Ray, Department of GI Surgery, IPGME&R, School of Digestive & Liver Diseases, Kolkata, India.
Emails: arkadipdhali@gmail.com (AD); drsukantaray@yahoo.co.in (SR)

Funding information
Nil.

Abstract

Extrahepatic mucinous biliary cystadenoma is an extremely rare clinical entity that can present with varieties of vague clinical signs and symptoms. Gastric outlet obstruction caused due to this has never been reported before. We highlighted the diagnostic dilemma we faced when radiological investigations could not suggest the accurate organ of origin.

KEYWORDS

biliary cystadenoma, extrahepatic, gastric outlet obstruction, mucinous neoplasm

1 | INTRODUCTION

Biliary cystadenoma is an extremely rare clinical entity, even more so when located in the extrahepatic biliary tree in comparison to their intrahepatic counterparts.[1] It accounts for less than 5% of all cystic lesions of the liver.[2] Less than 50 cases are reported in the existing medical literature. It is typically seen in middle-aged females and is thought to have hormonal factors such as estrogen and progesterone responsible for its pathogenesis.[3] Many a time, it is hard to diagnose due to its vague clinical signs and symptoms, prolonged clinical course, and difficult radiological interpretations. Although there are no cases of extrahepatic biliary cystadenomas presenting with gastric outlet obstruction and ours is the first of its kind. The case report was realized according to international Surgical CAse REport (SCARE) guidelines.[4] The aim of reporting the case report was to highlight the diagnostic dilemma we faced when radiological investigations could not suggest the accurate organ of origin.

2 | CASE REPORT

A 26-year-old female presented to the outpatient department with history of dull-aching, epigastric pain, radiating to the back for the last 7-months. This was accompanied by early satiety and abdominal fullness and progressed to non-bilious vomiting after taking food since the last 7-days. There was no history of fever,

weight loss, jaundice, or altered bowel habits. There was no relevant past or family history. Physical examination was unremarkable. Laboratory investigations (complete hemogram, serum electrolytes, transaminases, and serum bilirubin) were within normal limits. Contrast-enhanced computed tomography of the abdomen revealed a fairly enlarged pancreas. A 13.5 × 10.5 × 9.3 cm complex cystic space-occupying lesion (SOL) showing septation was seen (Figure 1A,B). The main pancreatic duct was not dilated. Overall, it was suggestive of a pancreatic cyst. To rule out any intraluminal cause of gastric outlet obstruction, an upper gastrointestinal endoscopy was performed which was unremarkable. Magnetic resonance imaging showed a well-defined cystic (T2 hyperintensity) lesion arising from left lobe of liver (Figure 1C).

An endoscopic ultrasound (EUS) was warranted to better characterize the pancreatic SOL. It showed large exophytic multiloculated cystic SOL with mural nodule arising from the neck of pancreas (Figure 2). EUS-guided aspiration of cyst fluid was done which showed cyst fluid amylase-23 U/L, CEA- 34.8 ng/ml. Cytology revealed few benign epithelial cells. Considering the provisional diagnosis as cystic neoplasm arising from head of the pancreas, Whipple's pancreaticoduodenectomy was planned. Intraoperatively, we found a distended gallbladder with non-dilated common bile duct along with a 18 × 10 × 10 cm septated cystic mass arising from the inferior surface of segments IVB and V of the liver (Figure 3). It was having minimal peri-cystic adhesions and was free from the hepatoduodenal ligament.

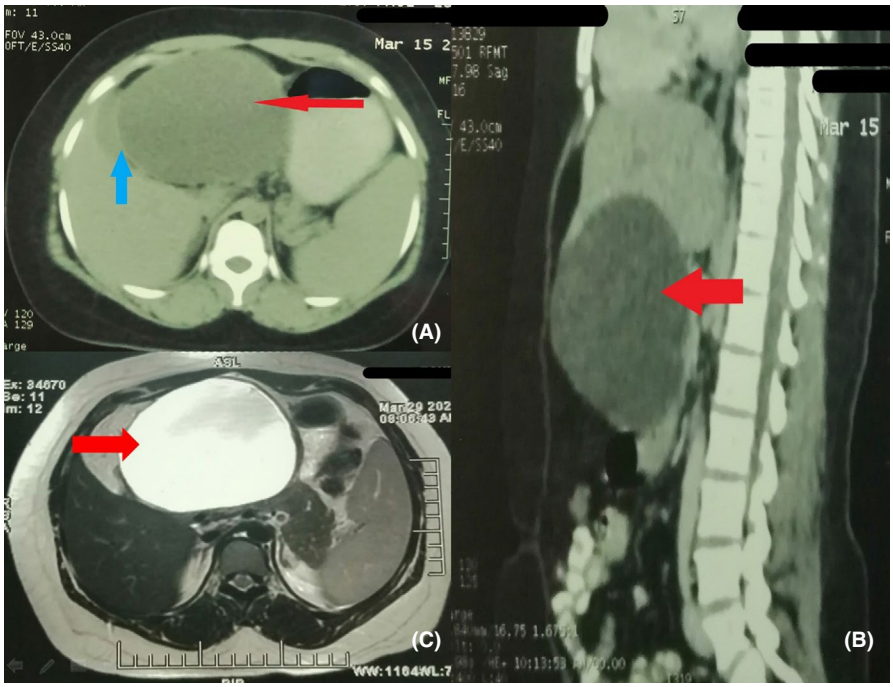

FIGURE 1 Contrast-enhanced computed tomography (A) transverse section, (B) saggital section) showing 13.5 × 10.5 × 9.3 cm complex cystic space-occupying lesion (red arrow) with septation (blue arrow). Magnetic resonance imaging (C) showing a well-defined cystic (T2 hyperintensity) lesion arising from left lobe of liver (red arrow)

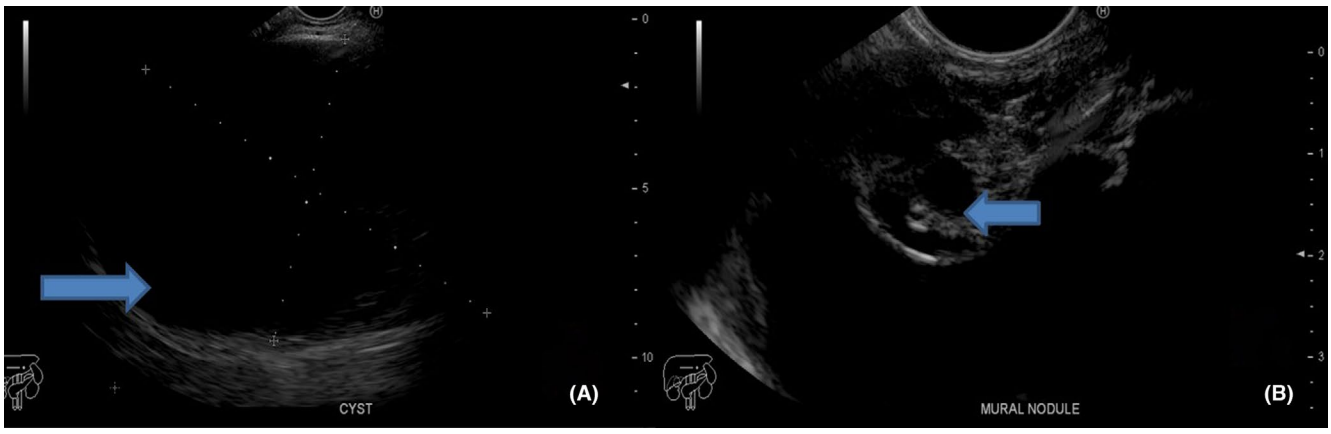

FIGURE 2 Endoscopic ultrasound showing, (A) large exophytic multiloculated cystic space-occupying lesion, (B) mural nodule arising from neck of pancreas

After careful adhesiolysis, and dividing the feeding vessels, enucleation of the mass was done from the liver bed along with cholecystectomy. Resected specimen (Figure 4) was sent for histopathological examination. It showed a cyst wall lined by cuboidal to columnar epithelium with apical mucin. Dense ovarian type of stroma was seen without any cellular and architectural atypia. Overall, it was suggestive of mucinous biliary cystadenoma (Figure 5). She had an uneventful recovery and was discharged on the 7th postoperative day. She was found to be well at 18-month follow-up.

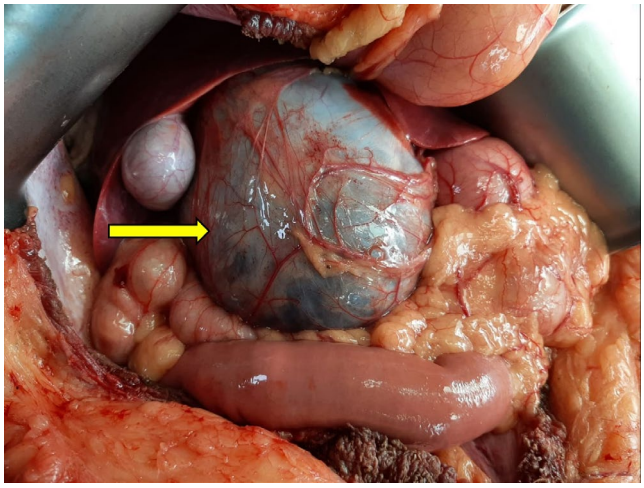

FIGURE 3 Intraoperative image showing distended gallbladder with non-dilated common bile duct along with a 18 × 10 × 10 cm septated cystic mass (yellow arrow) arising from inferior surface of segments IVB and V of liver

3 | DISCUSSION

The incidence of biliary cystadenoma ranges between 1:20,000 and 1:100,000, although the malignant counterpart is even rarer with an estimated incidence of 1:10 million.[5] These are generally sporadic in nature.[6] The exact origin of the neoplasm is unknown. Two schools of thought say that either they are derived from ectopic ovarian tissue or from ectopic embryonic gallbladder rests.[7] Estrogen-receptor positivity in the dense ovarian stroma points toward the hormonal dependence in the tumorigenesis.[8] This could very well explain its exclusive occurrence in females. Clinically, it can present with non-specific symptoms such as epigastric pain, vague abdominal discomfort obstructive jaundice, or even ascending cholangitis.[9] Although this was not the case for us. Here, the tumor was big enough to compress the stomach to cause clinical features of gastric outlet obstruction. Moreover, as it was extramural and partial obstruction, electrolytes were within normal limits.

Noninvasive imaging modalities such as computed tomography (multiloculated lesion, internal septation, papillary projections, intramural nodule, and septal enhancement), magnetic resonance imaging (characteristic of the fluid), endoscopic or transabdominal ultrasound helps in characterizing the lesion, but a definitive diagnosis can only be attained after histopathological evaluation. In our case, these investigations failed to point out the origin of the tumor. Intraoperatively, the origin of the tumor was made out and the surgeons took a call to modify the treatment plan as stated above. Preoperative needle aspiration or needle biopsy is not routinely performed as

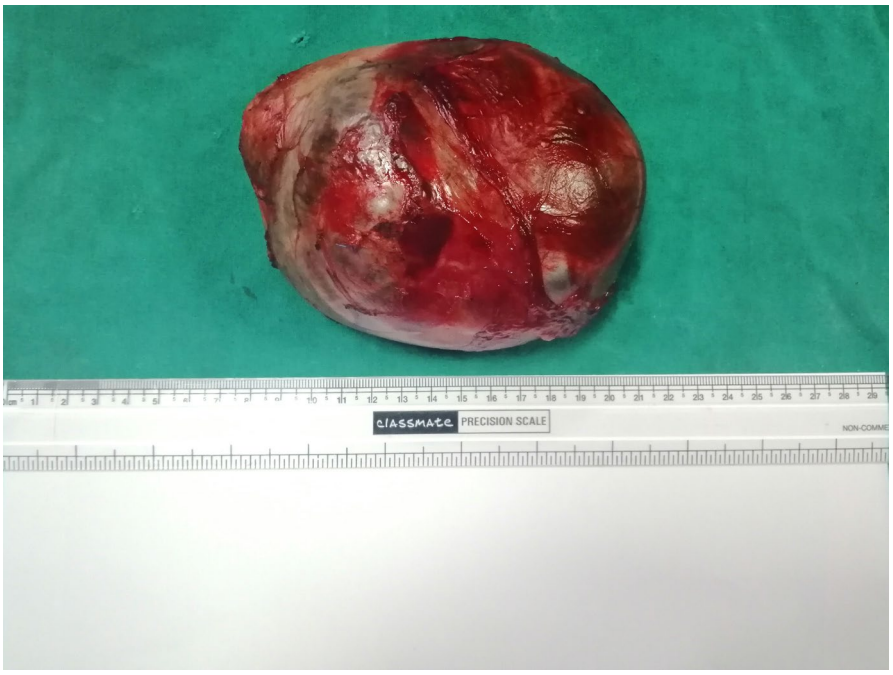

FIGURE 4 Enucleated specimen

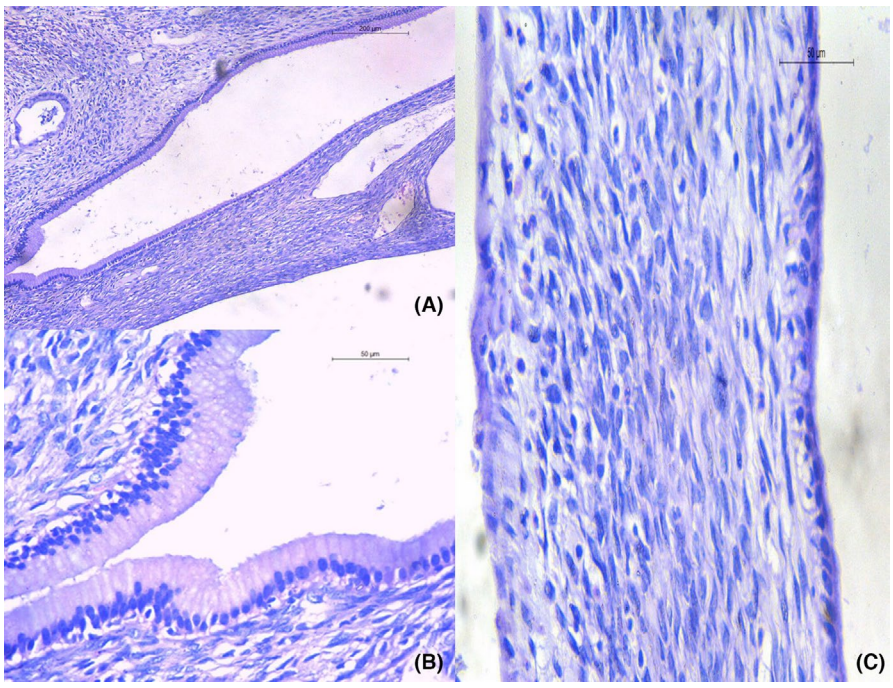

FIGURE 5 Hematoxylin and eosin image, A (10X) and B (40X): cyst wall lined by cuboidal to columnar epithelium with apical mucin, C (40X): dense ovarian type of stroma seen without any cellular and architectural atypia. Overall, it is suggestive of mucinous biliary cystadenoma

it increases the risk of secondary infection, intra-cystic bleeding, rupture of cyst, needle seeding of tumor cells, and dissemination, which may complicate the diagnosis and further management. Although in our case, the pre-operative diagnosis was thought to be pancreatic cystic lesion and differentials considered for that were cystic neoplasms and pseudocyst. Hence, cystic fluid aspiration was done to distinguish them.

Surgical excision (complete) is the treatment of choice and other treatment modalities such as partial resection, marsupialization, fenestration, and sclerosis are shown to be associated with recurrence with a rate that can be as high as 10%–90%.[10]

The resected specimen on histological examination gives the definitive diagnosis. The cysts are lined by gastric or biliary columnar epithelial cells with apical mucin. Dense ovarian stroma is also another hallmark finding which suggests the benign nature of the disease with a good prognosis where as its absence suggests poor prognosis.[11] Immunohistochemistry is also beneficial to better characterize the tumor and is done when the histopathology is inconclusive.

4 | CONCLUSION

Extrahepatic biliary cystadenoma is an extremely rare slow-growing tumor that arises predominantly in female and has malignant potential. It presents with vague clinical symptoms and clinicians should be aware of it as a differential of gastric outlet obstruction. Moreover, they might face a diagnostic dilemma when radiological investigations cannot suggest the accurate organ of origin, which can lead to confusion regarding the treatment to be followed. Hence, careful intraoperative evaluation is required before commencing with the procedure. Complete surgical resection remains the treatment of choice to prevent both recurrence and malignant transformation.

ACKNOWLEDGEMENT
None.

CONFLICT OF INTEREST
None declared.

AUTHOR CONTRIBUTIONS
Arkadeep Dhali, Sukanta Ray, and Gopal Krishna Dhali involved in conception, design of the study, acquisition of the data, drafting the manuscript, and final approval of the version to be submitted. Sujan Khamrui and Ranajoy Ghosh involved in acquisition of the data and final approval of the version to be submitted.

ETHICAL APPROVAL
Not required in our institution to publish anonymous case reports.

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.

DATA AVAILABILITY STATEMENT

The data that support the findings of this study are available from the corresponding author upon reasonable request.

ORCID

Arkadeep Dhali https://orcid.org/0000-0002-1794-2569
Gopal Krishna Dhali https://orcid.org/0000-0002-6759-5067

REFERENCES

1. Davies W, Chow M, Nagorney D. Extrahepatic biliary cystadenomas and cystadenocarcinoma. Report of seven cases and review of the literature. *Ann Surg*. 1995;222(5):619-625. doi:10.1097/00000658-199511000-00003
2. Wang YJ, Lee SD, Lai KH, Wang SS, Lo KJ. Primary biliary cystic tumors of the liver. *Am J Gastroenterol*. 1993;88:599-603.
3. Pedram-Canihac M, Le Bail B, Rivel J, Blanc J, Saric J, Bioulac-Sage P. Hepatobiliary cystadenoma with mesenchymal stroma: a hormone dependent tumor. Report of five cases with immunohistochemical study of hormone receptors [Article in French]. *Ann Pathol*. 2000;1:14-18.
4. Agha RA, Franchi T, Sohrabi C, Mathew G. The SCARE. guideline: updating consensus Surgical CAse REport (SCARE) guidelines. *Int J Surg*. 2020;84(2020):226-230.
5. Manouras A, Markogiannakis H, Lagoudianakis E, Katergiannakis V. Biliary cystadenoma with mesenchymal stroma: report of a case and review of the literature. *World J Gastroenterol*. 2006;12(37):6062-6069. doi:10.3748/wjg.v12.i37.6062
6. Dixon E, Sutherland FR, Mitchell P, McKinnon G, Nayak V. Cystadenomas of the liver: a spectrum of disease. *Can J Surg*. 2001;44(5):371-376.
7. Lee CW, Tsai HI, Lin YS, et al. Intrahepatic biliary mucinous cystic neoplasms: clinicoradiological characteristics and surgical results. *BMC Gastroenterol*. 2015;15:67. doi:10.1186/s12876-015-0293-3
8. Soares KC, Arnaoutakis DJ, Kamel I, et al. Cystic neoplasms of the liver: biliary cystadenoma and cystadenocarcinoma. *J Am Coll Surg*. 2014;218(1):119-128. doi:10.1016/j.jamcollsurg.2013.08.014
9. Srinivas T, Bhat SP, Sunder G, Sivarajan R. Mucinous cystic neoplasm of the liver and extrahepatic biliary tract with ascending cholangitis: a case report and review of the literature. *Indian J Surg Oncol*. 2020;11(Suppl 2):204-207. doi:10.1007/s13193-020-01121-7
10. Simo KA, Mckillop IH, Ahrens WA, Martinie JB, Ianniti DA, Sindram D. Invasive biliary mucinous cystic neoplasm: a review. *HPB*. 2012;14(11):725-740.
11. Soochan D, Keough V, Wanless I, Molinari M. Intra and extrahepatic cystadenoma of the biliary duct: review of the literature and radiologic and pathological characteristics of a very rare case. *BMJ Case Rep*. 2012;2012:bcr0120125497.

How to cite this article: Dhali A, Ray S, Dhali GK, Khamrui S, Ghosh R. Extrahepatic mucinous biliary cystadenoma: A rare cause of gastric outlet obstruction. *Clin Case Rep*. 2022;10:e05531. doi:10.1002/ccr3.5531