SPMC JHCS

CASE REPORT

Hepatic cystadenocarcinoma presenting as an ovarian new growth in a 65-year-old female: case report

Stephen Matthew B Santos,[1] Dulce De Castro,[1] Joffrey S Betanio,[1] Walter Batucan[1]

[1]Department of General Surgery, Southern Philippines Medical Center, JP Laurel Ave, Davao City, Philippines

Correspondence
Stephen Matthew B Santos, stephensantos4@gmail.com

Received
17 January 2020

Accepted
22 June 2021

Published online
30 June 2021

Cite as
Santos SMB, De Castro D, Betanio JS, Batucan W. Hepatic cystadenocarcinoma presenting as an ovarian new growth in a 65-year-old female: case report. SPMC J Health Care Serv. 2021;7(1):9. http://n2t.net/ark:/76951/jhcs7f6kp3

ABSTRACT

Biliary cystic tumors (BCT), which include the subgroups biliary cystadenoma (BCA) and biliary cystadenocarcinoma (BCAC), affect 5 to 10% of the global population. BCTs are solitary, multiloculated cysts that are usually intrahepatic in location. BCACs are rare tumors that arise from the malignant transformation of BCA. The presentation of BCT often mimics simple hepatic cysts and other hepatic cystic lesions making diagnosis difficult. With the recent advances in medical imaging, BCTs have been diagnosed more often. Patients with BCT are often asymptomatic. When symptoms are present, however, patients usually manifest with abdominal pain and distention. Given the high risk of recurrence, complete surgical resection by formal hepatic resection or enucleation is the best treatment of choice for patients. We present a case of a 65-year-old female who came in due to a four-year history of an enlarging abdomen. She was initially treated, preoperatively, as a case of ovarian new growth but was later managed as hepatic cystadenocarcinoma.

Keywords. Biliary cyst, pelvic mass, complete surgical resection, neoplasm

INTRODUCTION

Differentiating between a gynecologic from a non-gynecologic abdominopelvic mass, both of which appear solid on ultrasonography, can prove to be difficult, especially when the mass appears considerably large in size. Intra-abdominal masses with malignant clinical features can come from the gastrointestinal tract, hepatobiliary tract, or urinary tract, or they can be gynecologic in origin.[1] Hepatobiliary tumors can grow large and may mimic an abdominopelvic mass such as an ovarian new growth.

Biliary cystic tumors (BCT), which affect 5 to 10% of the global population, were once thought to be relatively uncommon. With advances in cross-sectional abdominal imaging, BCTs have recently been diagnosed more frequently.[2 3] However, its presentation is not straightforward—oftentimes mimicking simple hepatic cysts and other hepatic cystic lesions (e.g. hydatid cysts and metastatic tumors that undergo cystic degeneration), thus making diagnosis more difficult.[4]

The clinical evaluation of patients with BCT is often nonspecific. Many patients are often asymptomatic, and they are often diagnosed with this disease due to an incidental finding on abdominal imaging. Most symptomatic patients, however, present with abdominal pain and distention, bloating, and nausea and vomiting. Rarely, patients may also present with jaundice and signs of cholangitis, especially when significant biliary obstruction is present.[5 6]

The diagnosis of BCT is usually based on information from a combination of multiple sources, including ultrasonography, computed tomography (CT), magnetic resonance imaging (MRI), and histopathologic examination. Although imaging findings may be suggestive of the diagnosis, they are often overlapping with other cystic lesions of the liver often leading to a diagnostic dilemma and inadequate management.[3 7]

Complete surgical resection by formal hepatic resection or enucleation is the management of choice for BCTs, given their high risk of recurrence.[8 9] Since surgery alone may provide excellent long-term outcomes

IN ESSENCE

Biliary cystadenocarcinomas are rare cystic hepatic neoplasms that are a malignant transformation of biliary cystadenoma.

In this case report, we describe the case of a 65-year-old female with an abdominopelvic mass initially managed as ovarian new growth by the OB-Gyne Department, but was subsequently referred to the Hepatobiliary Service of the Surgery Department when an intraoperative finding of a hepatobiliary mass was found.

We performed an exploratory laparotomy, intraoperative ultrasound, excision of hepatic mass segment IVB, and we also did a transcystic intraoperative cholangiogram to check if the biliary tract was left intact after removal of the mass.

for biliary cystic adenocarcinoma (BCAC), there is limited role for adjuvant chemo- and radiotherapy in the management of this disease.[3]

We present the case of a 65-year-old female who came in due to a four-year history of gradual abdominal enlargement. The patient was initially managed as a case of ovarian new growth, but an intraoperative finding of a globular hepatic mass led to the diagnosis of hepatic cystadenocarcinoma.

CLINICAL FEATURES

A 65-year-old female was referred to our service by the the Obstetrics and Gynecology Department (OB-Gyne) during the intraoperative phase of the patient's surgery. She initially consulted at the Gynecology Outpatient Department (OPD) of our institution, complaining of gradual abdominal enlargement. The patient noted that her abdomen was gradually enlarging over the past four years. This was associated with intermittent, vague abdominal pain. She did not manifest any other symptoms. A year prior to admission, the patient consulted a private physician. An abdominal ultrasonography done on the patient revealed a right ovarian mass. The patient decided not to act on the findings until three months prior to admission, when she opted to seek consultation in our institution. A transvaginal ultrasound done revealed a huge abdominopelvic mass of probable ovarian origin, and its laterality undetermined. The mass exhibited a malignant sonomorphology. The endometrium was thickened and the ovaries were not seen. The patient was then scheduled for elective surgery and a consent for exploratory laparotomy, which included total abdominal hysterectomy with bilateral salpingo-oophorectomy was obtained.

The patient had been hypertensive for three years and had been taking Losartan 50 mg tablet and Amlodipine 10 mg tablet daily. She didn't smoke nor drink alcoholic beverages, and she has no family history of cancer. She denied other accompanying symptoms such as weight loss, profuse vaginal bleeding, or changes in bowel movement. Her obstetrical history was Gravida 5 Para 5 (5005) and all five children were delivered via spontaneous vaginal delivery with no fetomaternal complications. She had no prior history of abnormal uterine bleeding, abortion, or oral contraceptive pill use. Her menarche age was at 13, coitarche age at 25, and menopause age at 52. She had one sexual partner and has no history of sexually transmitted infection.

On physical examination, the patient had pale palpebral conjunctivae and a globular abdomen with a palpable, movable non-tender mass measuring approximately 30x40 cm. There were no ictericia, lymphadenopathies or edema. The rest of the physical examination findings were unremarkable.

DIAGNOSTIC APPROACHES

The patient's complete blood count results showed anemia of 102 g/L. Serum electrolytes, liver function tests, serum bilirubin levels, hepatitis profile and bleeding parameters were all normal. The transvaginal ultrasound done a few months prior to admission showed a huge heterogeneous multilocular solid mass which measures 28.01 x 30.28 x 12.89 cm. The mass exhibits more than four papillary structures, the largest of which measures 3.72 x 4.68 cm, with minimal vascularity.

THERAPEUTIC APPROACHES

The OB-Gyne Department admitted the patient under their service, continued her antihypertensive medications, and obtained her cardiopulmonary clearance. On the third hospital day, they performed an exploratory laparotomy which revealed a huge abdominopelvic mass that seemed to be connected to the liver and gallbladder. There was no ascites or hemoperitoneum. The uterine corpus, ovaries, and fallopian were grossly normal. The OB-Gyne Department immediately transferred the patient to the Hepatobiliary Service of the Surgery Department. Upon inspection, we noted that the mass had a smooth surface with an approximate size of 34x30x20 cm. The hepatic mass was attached superiorly to the liver edge at segment IVB, which displaced the gallbladder laterally to the right, the stomach to the left, and the bowel inferiorly. The mass is moveable at the base where the porta hepatis is located. (Figure 1) We performed an intraoperative ultrasound using a T-shaped probe, which revealed a well-demarcated, multiloculated mass. (Figure 2) Using electrocautery, we dissected the mass from the hepatic surface and removed the gallbladder together with the mass, which left a large defect on the surface of segments V/IVb. (Figure 2) There was no bile leak or active bleeding during the

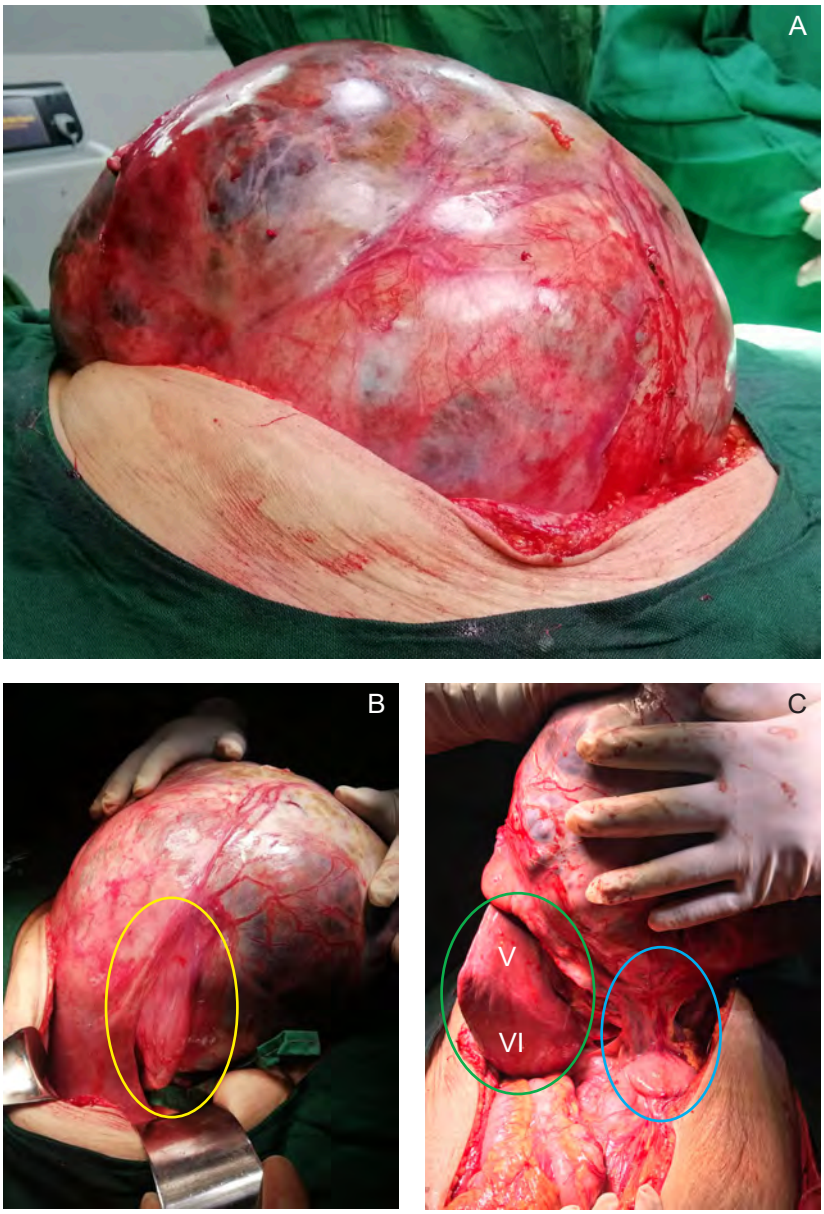

Figure 1 Intraoperative findings of the hepatic mass (A) and its adjacent structures. Lateral view with liver and gallbladder (B: yellow circle). Inferior attachment of the mass showing segments V and VI of the right lobe of the liver (C: green circle) and the porta hepatis (C: blue circle).

excision. We also performed a transcystic intraoperative cholangiogram, which revealed an opacification on both right and left intrahepatic ducts, an absence of filling defects in the common bile duct, and an egress of the dye into the laterally displaced duodenum. These findings indicate an intact biliary tract. There were also no bowel masses and mesenteric lymphadenopathies noted.

Grossly, the excised specimens are composed of the polypoid hepatic mass and the gallbladder. The hepatic mass consisted of gray, irregular cystic tissue with smooth external surface, which measured 32x30x19 cm and weighed 9kg. (Figure 3) Cut sections of the mass showed a multiloculated cyst filled with thick brown gelatinous material. Cut section of the gallbladder showed a dark-green velvety mucosa and an average wall thickness of 0.2 cm. There were no masses or stones noted.

Histopathological examination of the mass showed diffuse infiltrative sheets of atypical cells with peripherally located pleomorphic nuclei. These atypical cells, floating in lakes of mucin, exhibited glandular formation with a papillary growth pattern. There was no lymphovascular formation noted. (Figure 4) Sections of the gallbladder showed numerous mononuclear infiltrates with presence of Rokitansky-Aschoff sinuses. All these findings were consistent with mucinous cystadenocarcinoma and chronic cholecystitis.

OUTCOMES

The patient's postoperative course was uneventful and she was discharged four days after surgery. She followed-up at the OPD one week after discharge and had a regular weekly follow-up for one month. Laboratory tests showed normal serum bilirubin and alkaline phosphatase levels. She had hypoalbuminemia [26.54 (35-50 g/L)] and moderately elevated liver enzymes [SGPT 116.5 (14-54 U/L); SGOT 94.14 (15-41 U/L], which may be attributed to pathophysiological changes following liver resection. She

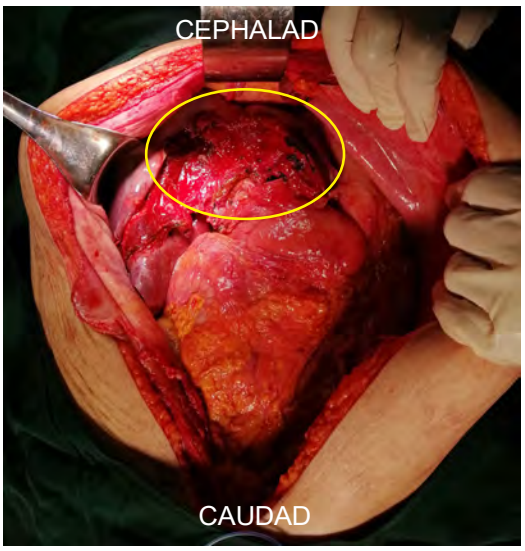

Figure 2 Defect (yellow circle) formed after excision of the hepatic mass and cholecystectomy.

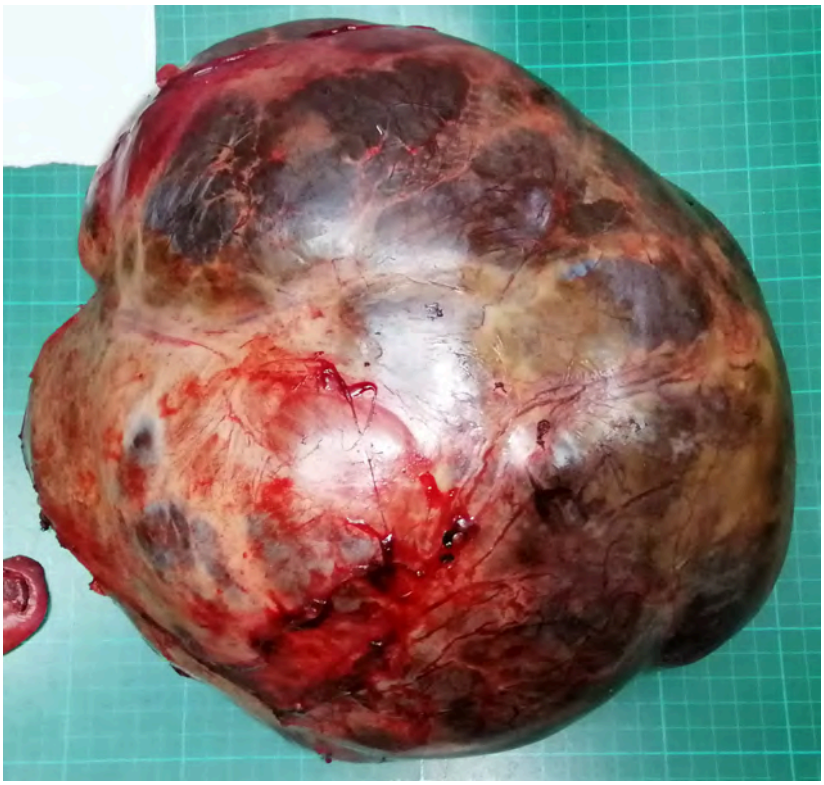

Figure 3 The excised polypoid hepatic mass showing an irregular cystic tissue with smooth external surface. The hepatic mass measures 32x30x19 cm and weighs 9kg.

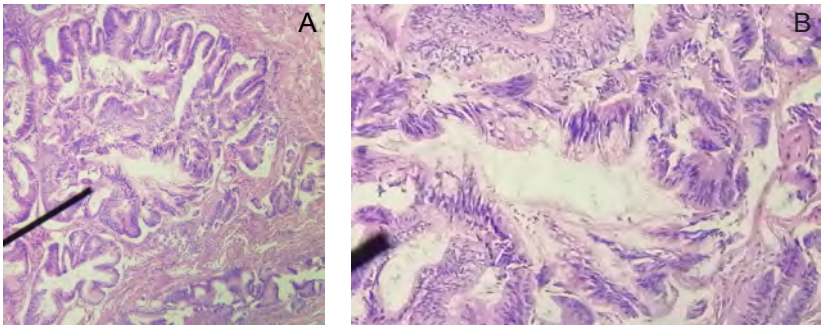

Figure 4 Histopathology of the hepatic mass at low power field (A:hematoxylin-eosin stain, x10) and high power field (B:hematoxylin-eosin stain, x40) showing diffuse infiltrative sheets of atypical cells with peripherally located pleomorphic nuclei surrounded by mucin-filled areas.

remained under observation and monitoring and was advised follow-up after 3 months for repeat abdominal ultrasound and liver function tests. The patient, however, was lost to follow-up starting a month after discharge.

DISCUSSION

The presence of an intraabdominal mass in a patient often poses a significant diagnostic dilemma for a surgeon. The mass can originate from many different causes, ranging from benign to malignant, and from the most common to the rarest. The pitfalls in diagnostic imaging of abdominal masses usually present a challenge to the surgeon in terms of prognosis and treatment outcomes of the disease. Our patient had an initial diagnosis of an ovarian new growth based on ultrasound but was later on managed differently after an intraoperative finding of a globular hepatic mass was found.

Mucinous cystic neoplasm is a subset within the hepatic cyst differential and it includes the subgroups called hepatic/biliary cystadenoma (BCA) and hepatic/biliary cystadenocarcinoma (BCAC).[2] BCAC are rare tumors that arise from the malignant transformation of BCA or from sequestered ectopic tissues of the foregut into the liver10 While BCA is more common in females (90%), BCAC is more evenly distributed between both sexes.3 BCA typically presents at ages 40 to 50 years old,[6 11] while BCAC occurs a decade later.[12]

BCACs clinically manifest from no symptoms to nonspecific symptoms such as vague abdominal pain (74%), jaundice (20%) and abdominal mass (14%).8 The size of the tumor may range from 1.5 to 35 cm.[13] Extrahepatic biliary cystic neoplasms commonly present with obstructive jaundice (85%) and the most common site of occurrence was the common bile duct (32%).[14] Our patient presented with nonspecific symptoms of vague abdominal pain and abdominal distention making it difficult to diagnose BCT with only these taken into consideration.

Although most patients present with normal laboratory values, approximately 20% of patients have elevated liver function tests.[15] A critical step in the management of BCTs is differentiating them from other complicated cystic lesions of the liver such as hydatid cyst, hemorrhagic cyst, liver abscess, atypical simple cyst, posttraumatic cyst, polycystic disease, and intraductal papillary mucinous neoplasm, which may only require a conservative management.[3 16]

Presently, imaging findings have nonspecific and overlapping characteristics that makes BCAC difficult to diagnose preoperatively.[2] Radiologically, both BCA and BCAC appear similar on ultrasound and CT—both demonstrating multilocular, septated lesions.[4 17] However, BCAC is more likely to demonstrate mural nodules, papillary projections, calcifications, and/or irregular cyst wall thickness on CT.[18] Our

patient's ultrasound findings showed a huge multilocular, solid, heterogeneous mass with papillary structures. These findings, however, are not pathognomonic for BCAC. MRI is another useful tool in evaluating these tumors. Fine-needle aspiration and core biopsy were sometimes undertaken, which demonstrate bile-tinged mucin and malignant glandular cells,[19] but have been widely associated with pleural and peritoneal dissemination.[3 17] Hence, our patient did not undergo these diagnostic procedures due to their associated risks.

On gross pathology, BCTs generally appear as solitary, multiloculated cysts that have dense fibrosis and calcifications.[20] BCA is characterized by the presence of a single layer of cuboidal to columnar while BCAC, on the other hand, exhibit a cytologically malignant multilayered epithelium. It can be distinguished from BCAs by the loss of polarity, mild nuclear pleomorphism, prominent nucleoli, and frequent mitotic figures.[21] Two types of BCAC exist, one with mesenchymal "ovarian-like" stroma that appears exclusively in females, and the other type which lacks the mesenchymal stroma which appears in both males and females. BCAC without the mesenchymal stroma follows a more aggressive course and has a worse prognosis than BCAC with mesenchymal stroma.[20 21]

Current data on the surgical management of patients with biliary cystic neoplasms are minimal and are limited to case reports and therefore inadequate to guide clinical practice.[22] Surgical options include complete liver resection with negative margins or careful enucleation depending on the anatomic position of the cyst and surgeon experience.[3 17 23] Since the mass of our patient was extrahepatic and did not involve major vessels and lobes, a complete excision of the hepatic mass was performed. Complete excision of BCAC had a 65% to 100% 5-year survival rate with only 13% recurrence rate.[12 23]

There is limited role of chemo- and radiotherapy for BCAC due to insufficient data coming from very few published case reports.[12 24] Currently, surgery alone for biliary cystadenocarcinoma showed excellent outcomes and prognosis even without adjuvant chemo- and radiotherapy.[3]

In summary, we were presented with a female patient having a four-year history of gradual abdominal enlargement. She was initially managed as a case of ovarian new growth, underwent exploratory laparotomy in order to remove the mass, but was managed differently when a globular hepatic mass was found during the intraoperative period. The diagnosis of BCTs—especially extrahepatic biliary neoplasms—require a high index of suspicion, since these tumors are usually misdiagnosed preoperatively with other liver lesions and thus managed inadequately.

Contributors
SMBS, DDC, JB, and WB contributed to the diagnostic and therapeutic care of the patient in this report. All of them acquired relevant patient data, and searched for and reviewed relevant medical literature used in this report. All wrote the original draft, performed the subsequent revisions, approved the final version, and agreed to be accountable for all aspects of this report.

Patient consent
Obtained

Reporting guideline used
CARE Checklist
(http://www.care-statement.org/downloads/CAREchecklist-English.pdf)

Article source
Submitted

Peer review
External

Competing interests
None declared

REFERENCES

1. Alessandrino F, Dellafiore C, Eshja E, Alfano F, Ricci G, Cassani C, et al. Differential Diagnosis for Female Pelvic Masses. IntechOpen [Internet]. 2013 Feb 20 [cited 2021 Jun 28]. Available from: https://www.intechopen.com/books/medical-imaging-in-clinical-practice/differential-diagnosis-for-female-pelvic-masses.

2. Del Poggio P, Jamoletti C, Forloni B, De Benedictis R, Mattiello M, Corti D, Pezzica E. Malignant transformation of biliary cystadenoma: a difficult diagnosis. Dig Liver Dis. 2000 Nov;32(8):733-6.

3. Soares KC, Arnaoutakis DJ, Kamel I, Anders R, Adams RB, Bauer TW, Pawlik TM. Cystic neoplasms of the liver: biliary cystadenoma and cystadenocarcinoma. J Am Coll Surg. 2014 Jan;218(1):119-28.

4. Alobaidi M. Biliary (Hepatic) Cystadenoma/Cystadenocarcinoma Imaging and Diagnosis. 2020 Sep 22 [cited 2021 Jun 29]. In: Medscape [Internet]. New York: Medscape. c1994-2021. Available from: https://emedicine.medscape.com/article/364156-overview.

5. Jawad N, Woolf AK, Chin-Aleong JA, Greaves R, Kocher HM. Biliary Cystadenoma Causing Obstructive Jaundice: Case Report and Literature Review. Case Rep Gastroenterol. 2009 Sep 12;3(3):269-274.

6. Soochan D, Keough V, Wanless I, Molinari M. Intra and extra-hepatic cystadenoma of the biliary duct. Review of literature and radiological and pathological characteristics of a very rare case. BMJ Case Rep. 2012 Apr 4;2012:bcr0120125497.

7. Metussin A, Telisinghe P, Kok K, Chong V. Extrahepatic biliary cystadenoma: a rare cause of biliary obstruction. Oman Med J. 2015 Jan;30(1):66-8.

8. Pitchaimuthu M, Aidoo-Micah G, Coldham C, Sutcliffe R, Roberts JK, Muiesan P, Isaac J, Mirza D, Marudanayagam R. Outcome following Resection of Biliary Cystadenoma: A Single Centre Experience and Literature Review. Int J Hepatol. 2015;2015:382315.

9. Arnaoutakis DJ, Kim Y, Pulitano C, Zaydfudim V, Squires MH, Kooby D, Groeschl R, Alexandrescu S, Bauer TW, Bloomston M, Soares K, Marques H, Gamblin TC, Popescu I, Adams R, Nagorney D, Barroso E, Maithel SK, Crawford M, Sandroussi C, Marsh W, Pawlik TM. Management of biliary cystic tumors: a multi-institutional analysis of a rare liver tumor. Ann Surg. 2015 Feb;261(2):361-7.

10. Lantinga MA, Gevers TJ, Drenth JP. Evaluation of hepatic cystic lesions. World J Gastroenterol. 2013 Jun 21;19(23):3543-54.

11. Morris M, Anderson C, Drake L, Redfield S, Subramony C, Vanderlan W. Giant biliary cystadenoma. J Surg Case Rep. 2012 Sep 1;2012(9):15.

12. Läuffer JM, Baer HU, Maurer CA, Stoupis C, Zimmerman A, Büchler MW. Biliary cystadenocarcinoma of the liver: the need for complete resection. Eur J Cancer. 1998 Nov;34(12):1845-51.

13. Ahanatha Pillai S, Velayutham V, Perumal S, Ulagendra Perumal S, Lakshmanan A, Ramaswami S, Ramasamy R, Sathyanesan J, Palaniappan R, Rajagopal S. Biliary cystadenomas: a case for complete resection. HPB Surg. 2012;2012:501705.

14. Davies W, Chow M, Nagorney D. Extrahepatic biliary cystadenomas and cystadenocarcinoma. Report of seven cases and review of the literature. Ann Surg. 1995 Nov;222(5):619-25.

15. Nakagawa M, Matsuda M, Masaji H, Goro W. Successful preoperative diagnosis of biliary cystadenoma with mesenchymal stroma and its characteristic imaging features: report of two cases. Turk J Gastroenterol. 2011 Dec;22(6):631-5.

16. Simo KA, Mckillop IH, Ahrens WA, Martinie JB, Iannitti DA, Sindram D. Invasive biliary mucinous cystic neoplasm: a review. HPB (Oxford). 2012 Nov;14(11):725-40.

17. Sang X, Sun Y, Mao Y, Yang Z, Lu X, Yang H, Xu H, Zhong S, Huang J. Hepatobiliary cystadenomas and cystadenocarcinomas: a report of 33 cases. Liver Int. 2011 Oct;31(9):1337-44.

18. Wilkinson N. Hepatobiliary cystadenocarcinoma. Case Reports Hepatol. 2012;2012:298957.

19. Logroño R, Rampy BA, Adegboyega PA. Fine needle aspiration cytology of hepatobiliary cystadenoma with mesenchymal stroma. Cancer. 2002 Feb 25;96(1):37-42.

20. Devaney K, Goodman ZD, Ishak KG. Hepatobiliary cystadenoma and cystadenocarcinoma. A light microscopic and immunohistochemical study of 70 patients. Am J Surg Pathol. 1994 Nov;18(11):1078-91.

21. Banerjee A, Shah SR, Singh A, Joshi A, Desai D. Rare biliary cystic tumors: a case series of biliary cystadenomas and cystadenocarcinoma. Ann Hepatol. 2016 May-Jun;15(3):448-52.

22. Arnaoutakis DJ, Kim Y, Pulitano C, Zaydfudim V, Squires MH, Kooby D, et al. Management of biliary cystic tumors: a multi-institutional analysis of a rare liver tumor. Ann Surg. 2015 Feb;261(2):361-7.

23. Vogt DP, Henderson JM, Chmielewski E. Cystadenoma and cystadenocarcinoma of the liver: a single center experience. J Am Coll Surg. 2005 May;200(5):727-33.

24. Mezale D, Strumfa I, Vanags A, Bahs G, Strumfs B, Silovs A, et al. Mucinous Cystic Neoplasms of the Liver and Extrahepatic Biliary Tract. IntechOpen [Internet]. 2018 Aug 29 [cited 2021 Jun 29]. Available from: https://www.intechopen.com/books/topics-in-the-surgery-of-the-biliary-tree/mucinous-cystic-neoplasms-of-the-liver-and-extrahepatic-biliary-tract.