# Cystic intraductal papillary mucinous neoplasm of the bile duct in the left lobe of the liver with associated liver atrophy: A case report

Sushma Jaishi[1], Kunal Bikram Deo[1], Barurendra Yogi[1], Yamuna Agrawal[1], Samiksha Lamichhane[1], and Shailesh Adhikary[1]

[1]BP Koirala Institute of Health Sciences

March 04, 2025

**Key Clinical Message**

Cystic IPMN-B is a rare liver disease with a high risk of malignant transformation (64%-89%) and poses diagnostic challenges. We describe a case of a 74-year-old woman with a large bilobar mucinous cyst in the left liver lobe associated with its atrophy managed with left hepatectomy. The cyst was lined with columnar cells with apical mucin and had radiological communication with the bile duct.

**Key words:** cystic IPMN-B; Mucinous Liver Cyst; Hepatectomy

## 1 Introduction

Primary cystic hepatobiliary neoplasm is a rare entity that can be benign and malignant.[1] The overall risk of malignant transformation or even harboring malignancy warrants careful evaluation and timely management of these cysts. Biliary cystadenoma/ mucinous cystic neoplasm and intraductal papillary mucinous neoplasm of bile duct (IPMN-B) are two broad categories of benign/premalignant cysts.[1] Cystic IPMN-B is a recently described distinct entity occurring exclusively in intrahepatic bile ducts. The risk of malignant transformation is 64% to 89% compared to 2% to 30 % in biliary cystadenoma.[1] However, making the clinical diagnosis of these cysts is a diagnostic challenge. We present a unique case of cystic IPMN-B of the left lobe of the liver with associated liver atrophy in an elderly lady.

## 2 Case Report

### 2.1 Case History, Examination and Diagnosis

A 74-year-old lady presented with right upper abdominal pain and lump associated with early satiety for three months. There was no history of fever or jaundice. She was a smoker for 50 pack years and had hypertension and chronic obstructive airway disease (COPD). A general physical examination revealed stable vitals parameters with body mass index of 20.7 $kg/m^2$. Abdominal examination revealed non-tender, smooth 5cm x 5cm smooth lump over the epigastrium. Blood investigation revealed a normal hemogram and liver function test with an alkaline phosphatase of 61 U/ml. Serum Cancer antigen (CA) 19-9 and carcinoembryonic antigen (CEA) were 7.8 U/ml and 0.5 ng/ml, respectively. Cyst fluid CEA was 0.5 ng/ml and was negative for malignant cells. Although the string test was suggestive, objective evidence of mucin could not be evaluated at our hospital. Transabdominal ultrasonography showed a large, well-defined cystic lesion with internal septation and no internal vascularity in the left lobe of the liver. Subsequently contrast enhanced computed tomography (CECT) of the abdomen (Fig 1a &1b) showed a hepatic cyst in segment IV and Spigelian caudate lobe with few enhancing septation with adjacent transient hepatic attenuation difference with compression and atrophy of the left lobe of the liver. The atrophic left lobe had a dilated intrahepatic biliary duct, and there was communication of a few bile ducts with the cyst. (Figure 1a & 1b)

### 2.2 Treatment

With the diagnosis of cystic IPMN-B, left hepatectomy with en-block lobectomy of Spiegel caudate lobe along with the entire cyst was performed. (Figure 1c &1d) Intraoperatively the right side of the cyst wall was partially adherent to the middle hepatic vein. The left hepatic vein had a common trunk with the middle hepatic vein and was ligated subsequently after the identification. The cyst was bilobar of size 5cm x 5cm and 10cm x 10cm respectively with atrophy of the left hepatic lobe. The left hepatic duct was compressed and was seen ending blindly into the cyst. The cyst fluid was clear and mucinous.

### 2.3 Outcome and Follow-up

The patient had good postoperative recovery after postoperative intensive care management for COPD exacerbation. The histopathological evaluation of the resected specimen revealed variable-sized cysts lined with columnar cells having basal nucleus and apical mucin. The cyst contained mucinous material. The surrounding liver tissue showed fibrosis encircling hepatocytes forming parenchymal nodules but lacked ovarian-like stroma. (Figure 2) The patient is doing well in the follow-up of 1 year with no recurrence.

## 3 Discussion

The cyst leading to atrophy of the left lobe of the liver, radiological communication of dilated intrahepatic bile ducts within, and absence of ovarian-like stroma makes our case unique. World Health Organisation (WHO) in 2010 proposed uniform terminology of mucinous cystic neoplasm in the presence of ovarian-like stroma (OS) or intraductal papillary mucinous neoplasm of bile duct (IPMN-B) in the presence of biliary tree communication.[2] However, the presence of OS and demonstration of bile duct communication is not present in all cases.[3] The literature does not clarify whether cystadenoma without OS is a discrete entity or IPMN-B where biliary communication could not be established. The prevalence of these cysts compared to those with OS is lower[4] but few studies have shown equal prevalence as well.[5] Most IPMN-B cases have been described as tubular or fusiform dilatation of bile ducts similar to IPMN of the pancreas.[6] The rate of malignancy in IPMN-B (60% - 70%) is higher than that of the pancreatic counterpart of IPMN (23%-30%).[7,8] The presence of mural nodules suggesting malignancy is commonly present except in benign cases.[7] The stage of malignancy in IPMN-B is however less advanced.[1]

Cystic IPMN-B is an uncommon variant as in the index case resembling branch duct IPMN of the pancreas.[6] These cysts have been thought to arise from prebiliary glands located within the wall or scattered in the surrounding connective tissue of the larger bile duct.[9] It has equal sex predilection and peaks in the sixth decade.[1,10] The cystic IPMN-B is frequently associated with mural nodules and bile duct dilatation distal to the cyst.[7] This is attributed to mucus from cysts in the bile duct leading to obstruction and dilatation, which in the long run could lead to liver atrophy as in the index case. Mucobilia may result in transient biliary obstruction or recurrent cholangitis.[1] The risk factors are shown to be hepatolithiasis, Clonorchis infections, etc.[11]Although biliary communication is a key feature, CECT or magnetic resonance imaging cannot always demonstrate luminal communication, probably due to narrow communications.[12] However, the biliary communication in the background of the atrophic liver was well demonstrated by a CT scan in the index case. Cyst fluid CEA in the index case was normal. The exact correlation of CEA to cystic IPMN-B is scarcely described in literature, but higher fluid CEA has been associated with cystadenocarcinoma.[13]

All Cystic IPMN-B requires resections, preferably with intraoperative cholangiogram to demonstrate biliary communication and to rule out mucus plugs in the bile duct. Preoperative endoscopic retrograde cholangiography or intraoperative cholangiography (IOC) is more reliable as macroscopic examination is often accurate.[14] IOC was not used in the index case due to logistic issues. Although the index case had no malignant foci, most of the cases have malignant foci which warrant formal hepatectomy in all cases. Additionally, extrahepatic bile duct resection and lymph node sampling may also be required in case of malignant cysts. The prognosis after resection is good compared to intrahepatic or hilar cholangiocarcinoma with 5-year survival of 60% to 80%.[10] The columnar cells and MUC1 expression adversely impact survival.[1,10]

## 4 Conclusion

Hepatobiliary cysts require careful evaluation due to the risk of malignant transformation. Cystic IPMN-B is an uncommon variant that requires formal hepatectomy due to the high risk of malignancy, and it poses a preoperative diagnostic challenge. A timely and precise management results in a good prognosis for these cysts.

**Disclosures**

**List of abbreviations:**

IPMN-B: Intraductal papillary mucinous neoplasm of bile duct

CEA: carcinoembryonic antigen

CA 19-9: Cancer antigen 19-9

CECT: Contrast-enhanced computed tomography

COPD: chronic obstructive airway disease

OS: ovarian-like stroma

IOC: Intraoperative cholangiography

**Ethical approval and consent to publication**

This case study was ethically approved by the Institutional Review Committee, B P Koirala Institute of Health Sciences, Dharan, Nepal. Written informed consent from the patient and her guardian for the publication of the case study was taken.

**Conflict of interest:** nothing to disclose

**Funding** : none

**Data statement:** All relevant data about this patient is in the case report, and further data about the patient can be made available on request

**Authors contributions:**

*Sushma Jaishi:* Conceptualization, Data curation, Formal analysis, Methodology, Writing -original draft

*Kunal Bikram Deo* : Conceptualization, Data curation, Formal analysis, Investigation, Methodology, Project administration, Resources, Supervision, Writing - original draft, Writing - review & editing

*Barurendra Raj Yogi* : Conceptualization, Investigation, Methodology, Project administration, Writing - original draft

*Yamuna Agrawal: Investigation, Project administration, Resources, Writing - review & editing*

*Samiksha Lamichhane: Investigation, Project administration, Resources, Writing - review & editing*

*Shailesh Adhikary: Formal analysis, Supervision, Validation, Visualization, Writing – review, editing & approval*

**REFERENCES**

1. Farges O, Paradis V. Cystic hepatobiliary neoplasia. In:*BLUMGART'S Surgery of the Liver, Biliary Tract and Pancreas* . Vol 2. 7th ed. Elsevier; 2023:1203-1217.

2. Tsui W, Adsay N, Crawford J. Mucinous cystic neoplasms of the liver. In: *World Health Organization Classification of Tumours. Pathology and Genetics of Tumours of the Digestive System* . IARC Press; 2010:236-238.

3. Kishida N, Shinoda M, Masugi Y, et al. Cystic tumor of the liver without ovarian-like stroma or bile duct communication: Two case reports and a review of the literature. *World J Surg Oncol* . 2014;12:229. doi:10.1186/1477-7819-12-229

4. Devaney K, Goodman ZD, Ishak KG. Hepatobiliary cystadenoma and cystadenocarcinoma: A light microscopic and immunohistochemical study of 70 patients. *Am J Surg Pathol* . 1994;18:1078-91. doi:10.1097/00000478-199411000-00002

5. Vogt DP, Henderson JM, Chmielewski E. Cystadenoma and cystadenocarcinoma of the liver: A single center experience. *J Am Coll Surg* . 2005;200:727-33. doi:10.1016/j.jamcollsurg.2005.01.005

6. Sakamoto E. Clinicopathological studies of mucin-producing cholangiocarcinoma. *J Hepatobiliary Pancreat Surg* . 1997;4:157-62. doi:10.1007/BF02489781

7. Kwang YP, Jin SH, Seong HC, Dong WC. Intraductal papillary neoplasm of the bile ducts: The clinical features and surgical outcome of 25 cases. *J Surg Oncol* . 2008;97:508-12. doi:10.1002/jso.20994

8. Minagawa N, Sato N, Mori Y, Tamura T, Higure A, Yamaguchi K. A comparison between intraductal papillary neoplasms of the biliary tract (BT-IPMNs) and intraductal papillary mucinous neoplasms of the pancreas (P-IPMNs) reveals distinct clinical manifestations and outcomes.*Eur J of Surg Oncol* . 2013;39:554-8. doi:10.1016/j.ejso.2013.02.016

9. Nakanishi Y, Zen Y, Hirano S, et al. Intraductal oncocytic papillary neoplasm of the bile duct: The first case of peribiliary gland origin.*J Hepatobiliary Pancreat Surg* . 2009;16:869-73. doi:10.1007/s00534-009-0070-1

10. Shibahara H, Tamada S, Goto M, et al. Pathologic Features of Mucin-producing Bile Duct Tumors: Two Histopathologic Categories as Counterparts of Pancreatic Intraductal Papillary-mucinous Neoplasms.*Am J Surg Pathol* . 2004;28:327-38. doi:10.1097/00000478-200403000-00005

11. Chen TC, Nakanuma Y, Zen Y, et al. Intraductal papillary neoplasia of the liver associated with hepatolithiasis. *Hepatology* . 2001;34:651-8. doi:10.1053/jhep.2001.28199

12. Lim JH, Zen Y, Jang KT, Kim YK, Nakanuma Y. Cyst-forming intraductal papillary neoplasm of the bile ducts: Description of imaging and pathologic aspects. AJR *Am J Roentgenol* . 2011;197:1111-20. doi:10.2214/AJR.10.6363

13. Pinto M M, Kaye A D. Fine needle aspiration of cystic liver lesions. Cytologic examination and carcinoembryonic antigen assay of cyst contents. *Acta Cytol* . 1989;33:852-56.

14. Zen Y, Fujii T, Itatsu K, et al. Biliary cystic tumors with bile duct communication: A cystic variant of intraductal papillary neoplasm of the bile duct. *Mod Pathol* . 2006;19:1243-54. doi:10.1038/modpathol.3800643

**Legends**

**Figure 1: Computed tomography and intraoperative images**

**a and b** : Cross-sectional and coronal Computed tomography images showing hepatic cyst in the left lobe of the liver with enhancing sepatation giving a bilobed appearance with adjacent transient hepatic attenuation difference and left lobar atrophy. The cyst abuts the middle hepatic vein. Mild dilated intrahepatic bile duct ducts in the atrophic liver seem to communicate with the cyst (yellow arrow).

**c:** Intraoperative images showing bilobed cyst in the left lobe of the liver (white arrows).

**d:** Image after left lobe and Spigelian caudate resection. White arrow indicates inferior vena cava and middle hepatic vein.

**Figure 2: Histopathology images**

**a:** variable size of cyst

**b:** Larger view of cyst containing fluid

**c:** Cyst lined by single layer of columnar to cuboidal cells

**d:** Section from rest liver shows fibrosis forming a incomplete nodules

**e:** Masson's trichrome stain highlights the fibrosis

**f:** Reticulin stain highlights the fibrosis encircling hepatocytes forming a parenchymal nodule