

## Hepatic Mucinous Cystic Neoplasm with Fistulation to The Duodenum: A Rare Case of Recurrent Cholangitis

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### Abstract

Non-invasive mucinous cystic neoplasms (MCN) are rare biliary cystic tumours occurring predominantly in females. We report the case of a 56-year-old female presenting with abdominal pain, a mass and subsequently jaundice in the background of previous biliary surgery. Cross sectional imaging was suggestive of a MCN with possible fistulation to the duodenum. She underwent an extended right hepatectomy with extrahepatic bile duct resection and a left and caudate hepaticojejunostomy. Histology confirmed a non-invasive MCN. The patient made an uneventful recovery and remains well more than a year after surgery.

**Keywords:** Hepatic MCN; Recurrent Cholangitis; Extended Right Hepatectomy

## Introduction

Hepatic mucinous cystic neoplasms (MCN) originate primarily from intrahepatic bile ducts and account for 5% of liver cysts [1,2]. They include non-invasive MCNs or biliary cystadenomas (BCA) and MCNs with invasive carcinoma, or biliary cystadenocarcinomas (BCAC) [1]. Non-invasive MCNs occur predominantly in middle aged females[3] and have a 20% risk of malignant transformation [4]. They are often asymptomatic but may present with abdominal pain and distention due to extrinsic compression in the gastro-duodenal region [5] and less commonly with obstructive jaundice, cholangitis, haemorrhage, and cyst rupture [6] Confirmation is by pathology characterised by the presence of ovarian-type stroma that expresses oestrogen and progesterone receptors lined by biliary type mucus secreting cuboidal or columnar epithelium [2]. The risk of malignant transformation and the propensity for recurrence warrants complete surgical excision [7] . We report the case of a middle-aged female with a non-invasive MCN with possible fistulation to duodenum.

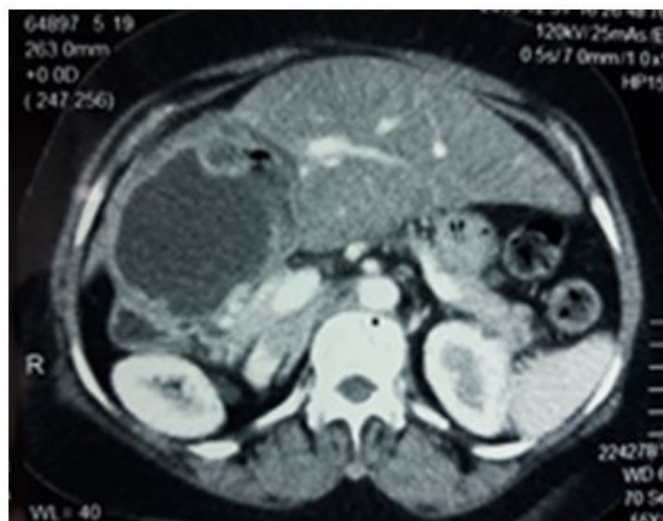
## Case presentation

A 56-year-old female was referred with upper abdominal pain, early satiety, and a right upper quadrant mass. She had undergone surgery twice, 18 years prior to presentation, via right paramedian incisions; a cholecystectomy followed by another biliary procedure, for calculi and cholangitis, according to available records. She had been relatively asymptomatic during the intervening years until the current presentation.

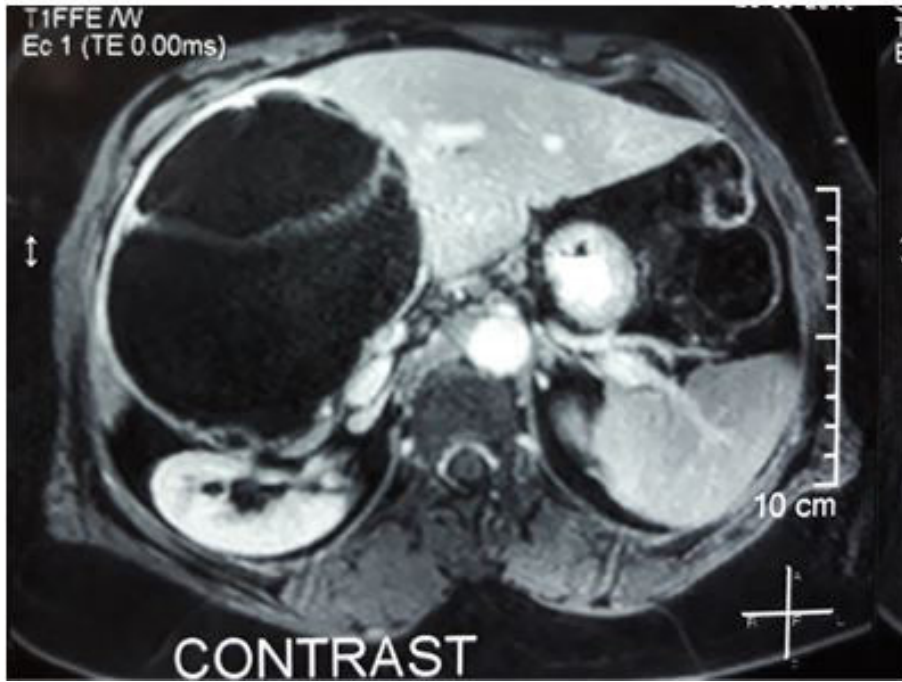
Her liver biochemistry was normal. Abdominal CT and MRI / MRCP showed an encapsulated lesion (13 x 12 x 13 cm) in the right hepatic lobe (Figure 1) suggestive of a non-invasive

MCN (Figure 2). US guided cyst aspiration performed as a temporising measure for symptomatic relief produced inspissated bile, while simultaneous contrast injection failed to demonstrate communication with the bile ducts. She preferred to defer surgery and returned home but presented a few months later with non-bilious vomiting, significant weight loss and jaundice. A repeated CT abdomen showed the cyst, now reduced in size (8.1 x 6.7 x 9.5 cm) with enhancing, thickened, irregular walls and internal septae in segments IV, V and VI compressing the proximal duodenum. There was mild intrahepatic duct dilatation with aerobilia, which in the absence of sepsis raised the possibility of a cysto-duodenal fistula.

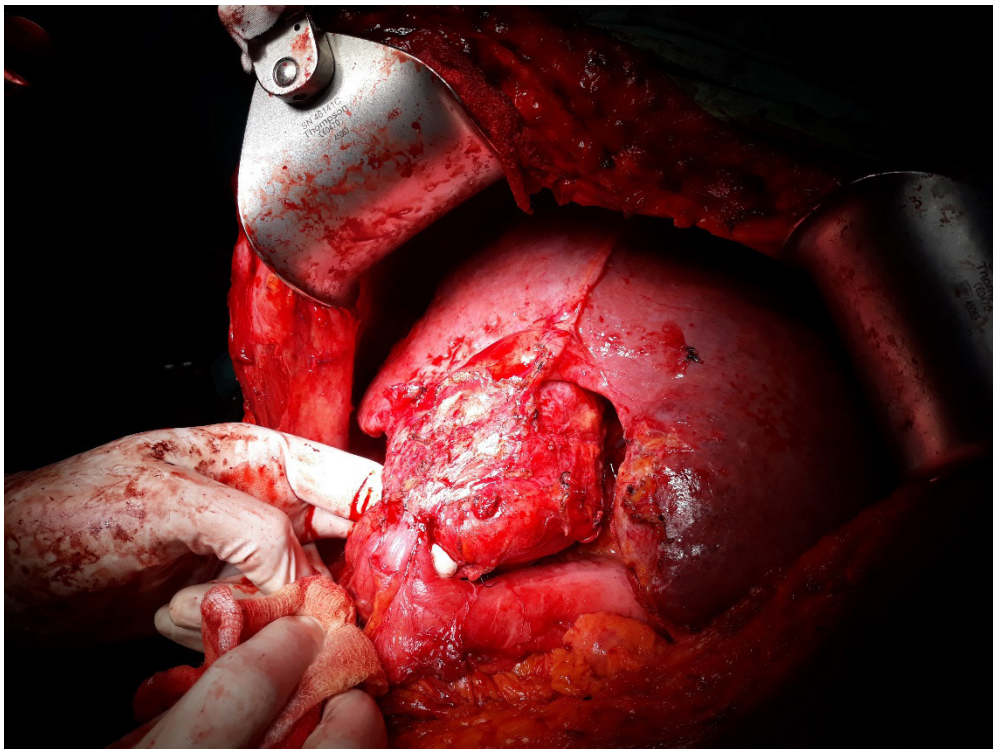
Surgery was performed via an inverted L incision. Dense adhesions to the visceral surface of the liver and hilum were divided. The right lobe was atrophic with an exophytic cystic lesion arising from segments IVB and V adherent to porta hepatis and the junction of the 1st and 2nd parts of the duodenum (Figure 3) and compensatory hypertrophy of the left lateral segment. An extended right hepatectomy and extrahepatic bile duct resection was performed. A cuff of the sero-muscular layer of the duodenum was excised with the cyst and oversewn, though no fistula was demonstrated. Reconstruction was with a Roux-en-Y left hepaticojejunostomy and an additional anastomosis to incorporate a dilated caudate lobe duct (Figure 4). Histology confirmed a uniloculated 65 x 55 x 45 mm benign MCN (Figure 5 A, B) demonstrating ovarian like stroma with progesterone receptor positivity on immunohistochemistry (Figure 5-C, D). There was no evidence of malignancy. The postoperative period was complicated by a right subdiaphragmatic collection on day 7 which resolved with ultrasound guided percutaneous drainage and antibiotics. She was discharged on the 18th postoperative day and remains well more than a year after surgery.



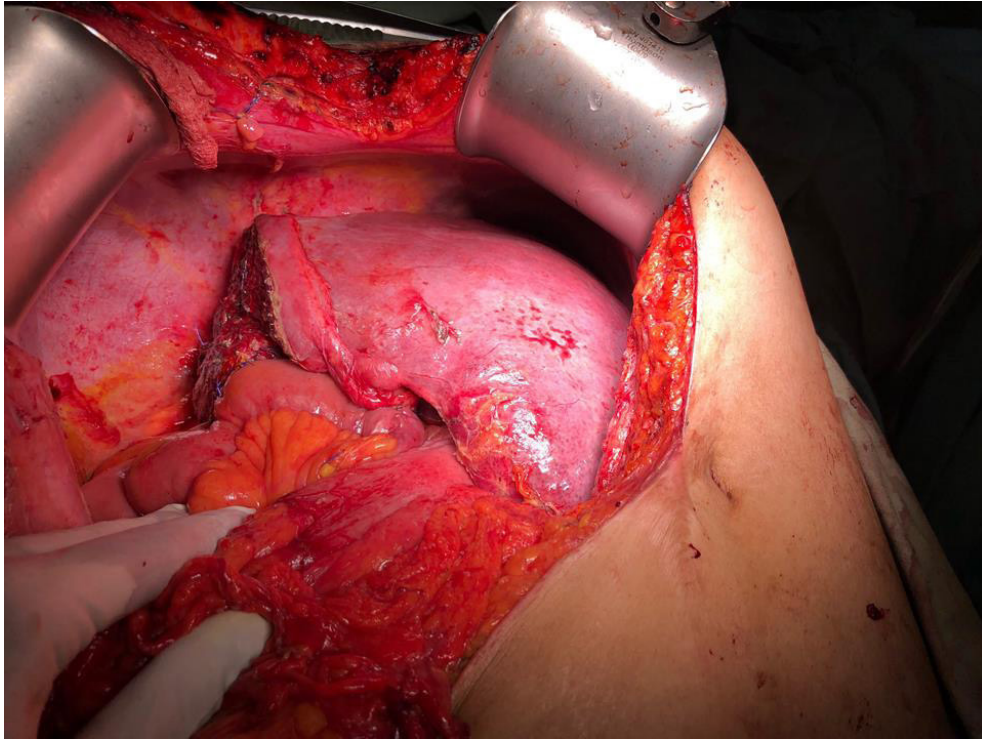
**Figure 1:** Contrast enhanced CECT with hypodense encapsulated non enhancing lesion in segment IV,V/VI



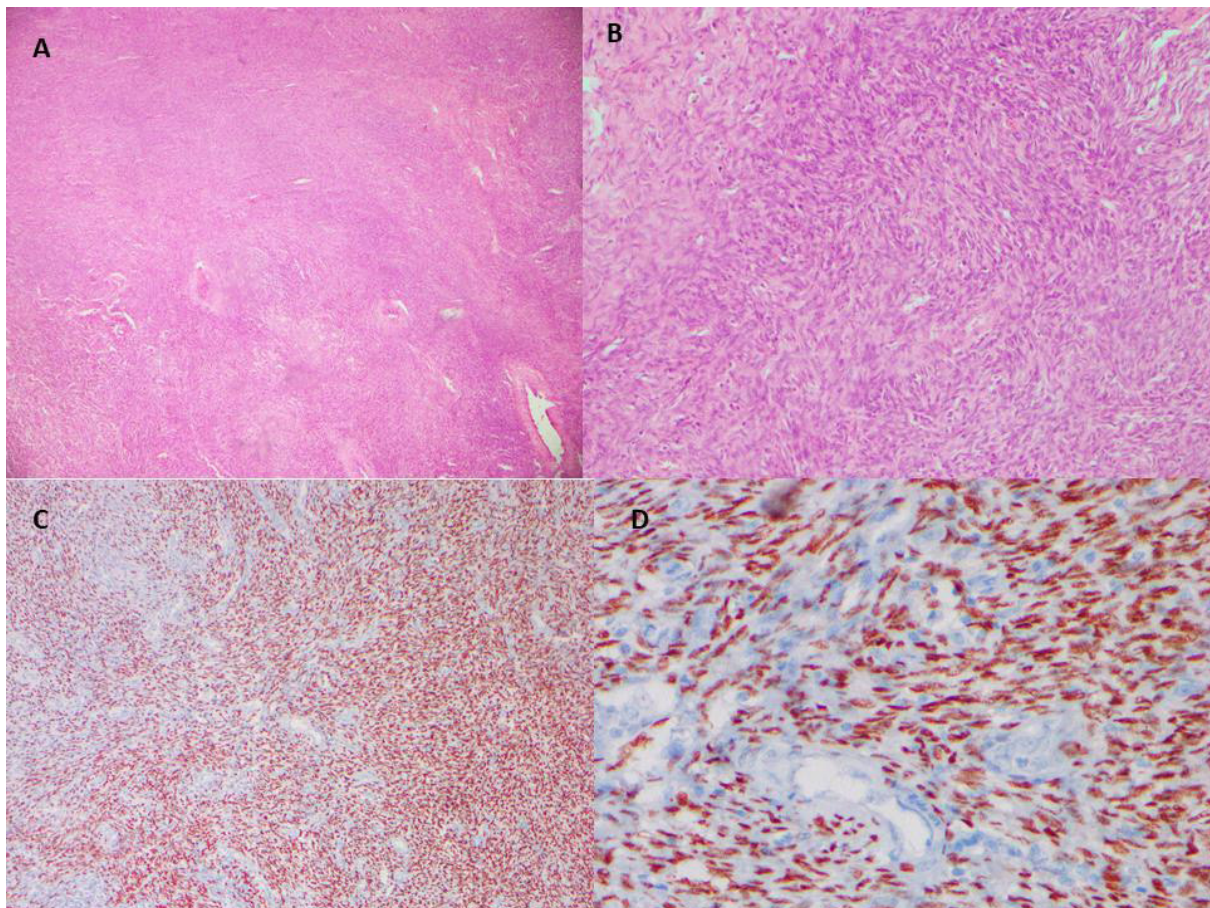
**Figure 2:** MRI liver showing cystic lesion in segment V,VI



**Figure 3:** Cystic lesion arising from the right lobe segments IVB/V adherent to porta hepatis infiltrating into D1/D2



**Figure 4:** Extended right hepatectomy with bile duct excision and a left hepaticojejunostomy and caudate hepaticojejunostomy



**Figure 5:** Histopathology images. A, B- H and E slides showing ovarian stroma of biliary MCN (A-Low power, B high power). C, D- Immunohistochemistry showing progesterone receptor positivity

## Discussion

Non-invasive hepatic MCNs, are rare benign neoplasms of the biliary tract that mostly originate from the right lobe [8] The presence of mesenchymal stroma suggests its origin from ectopic embryonal tissue forming the gallbladder [9,5]

Though mostly asymptomatic, epigastric pain or a mass are presentations with vomiting in the presence of pressure effects [5] Obstructive jaundice and cholangitis are less common [6] and intracystic haemorrhage, cyst rupture and malignant transformation are complications [9] The jaundice seen later in this patient may have been due to raised intracystic pressure secondary to infection or haemorrhage causing common bile duct compression or alternatively the presence of mucobilia [10]. Although our patient's initial presentation was abdominal pain subsequently she developed jaundice possibly due to compression which was evident by dense adherence of cyst to bile duct and invasion of bile duct .

A high index of suspicion for MCNs is imperative when investigating complex cysts of the liver, mainly due to their propensity to undergo malignant change. Differential diagnoses include simple cysts, hydatid cysts, liver abscesses, polycystic disease, Caroli's disease, embryonal sarcomata, and biliary intra-ductal papillary mucinous neoplasms (IPMN). Most have a normal liver biochemistry and about 20% present with derangement such as hyperbilirubinaemia [6]. Differentiation of MCNs from other complex cystic hepatic lesions is by ultrasonography and cross sectional imaging including CT and MRI [11]. MCNs appear as anechoic lesions with irregular thickened walls and internal septae characterised by septal thickening, papillary infolding, and mural nodules on ultrasonography. On CT, isodense lesions with enhancing nodularity and bile duct dilatation are predictive of MCN[12] while on MRI, multi-locular, irregular thick walls with low intensity T1 signals and high intensity T2 signals are seen[13]. Although imaging may help differentiate MCNs from other cystic lesions of the liver, malignant transformation is primarily a histopathological diagnosis[7]. MCNs are usually multilocular with nodular internal septae surrounded by a dense fibrostroma, though rarely they may be unilocular as in this case[14]. Imaging studies [12,13] usually reveal complex cystic mass with internal septa, mural or septal nodules and papillary projections, which may enhance with contrast administration.

The MCNs exhibit a mass effect on imaging because of higher viscosity of fluid compared with simple hepatic cysts containing serous fluid [13].

Our patient's histopathology showed biliary cyst epithelium with ovarian stroma and progesterone receptor positivity on immunohistochemistry. Microscopically, the cysts contain three layers including a biliary type epithelium, a moderate to dense cellular ovarian stroma, and dense collagenous connective tissue[6]. Stromal cells are immunoreactive to vimentin, muscle-specific actin, alpha- smooth muscle actin, desmin, oestrogen, and progesterone receptors[15].

Surgical resection is the treatment of choice for hepatic MCNs in view of their malignant potential[7]. Peripherally situated MCNs confined to one lobe are amenable to resection whereas central lesions involving bilio-vascular structures may warrant enucleation[7]. Recurrence rates after complete resection are between 5-10% [16]. Unlike other primary hepatic tumours, systemic therapies have not been found to be particularly effective in the treatment of primary invasive MCNs. Given that chemotherapy has been utilized as a primary, adjuvant or salvage therapy have been reported in the literature, no definitive recommendations in this area can be made [17]. The dense adhesion of the cyst wall to the bile duct necessitated extrahepatic bile duct resection in this patient.

## Conclusion

Mucinous cystic neoplasms (MCN) are rare benign lesions with the potential for malignant change. Diagnosis is aided by multimodal imaging and surgical excision is the treatment of choice.

**Authors Contribution-** RJ and DS prepared the manuscript and figures. DS, VD, SS were involved in patient management. MVCDS involved with histopathological assessment of the surgical specimen and final pathology report. SS supervised the manuscript preparation. All authors approved the submitted manuscript.

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**Ethics approval** - Not applicable.

**Patient consent for publication**- Informed written consent was obtained from the patient for publication of this case report and accompanying images. A copy of the consent form is available for review by the Editor of this journal.

### **Availability of data and materials**

Data sharing is not applicable to this article as no datasets were generated or analyzed in this study.

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