ORIGINAL ARTICLE

# Peribiliary cysts masquerading as choledocholithiasis

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## CASE PRESENTATION

A 70-year-old man presented to his primary care physician with progressive shortness of breath. A subsequent workup revealed a normal chest radiograph, but the CT scan of the chest revealed heterogeneous hepatic cysts. Cross-sectional abdominal imaging revealed "beaded" bilobar intrahepatic biliary ductal dilation and narrowing of the distal common bile duct concerning for obstruction (Fig. 1). Laboratory analysis showed normal liver function tests. He reported 15 years of heavy alcohol use. He denied other risk factors for chronic liver disease, fevers, chills, or night sweats. His physical examination was unremarkable. He was referred to gastroenterology department and underwent an EUS showing multiple anechoic structures in both liver lobes, extrahepatic tree, and the pancreatic head suspicious for a type IV choledochal cyst in the liver and side-branch intraductal papillary neoplasms in the pancreatic head. He was referred to surgery and was offered a Whipple pancreaticoduodenectomy to remove all cystic portions of his extrahepatic biliary tree and pancreatic head with lifelong surveillance for the intrahepatic ducts. Seeking a second opinion, the patient was referred to our institution where an ERCP was performed.

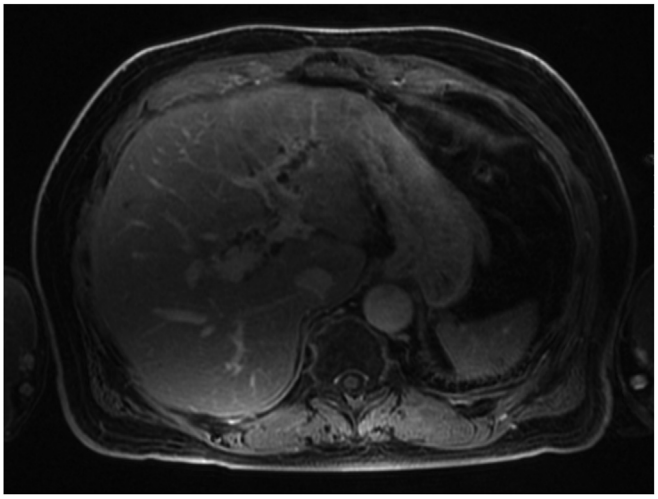

**Figure 1.** Liver acquisition with volume acceleration T1 phase magnetic resonance cholangiogram revealing bilobar intrahepatic polypoid structures.

## PROCEDURE

The ERCP demonstrated filling defects in the mid to proximal bile duct with a chain-like appearance in the left main bile duct on cholangiogram suggestive of stones (Fig. 2). Balloon sweeps did not retrieve any stones or debris. Cholangioscopy showed filling defects representing polypoid structures suggestive of peribiliary cysts (Fig. 3; Video 1, available online at www.videogie.org).

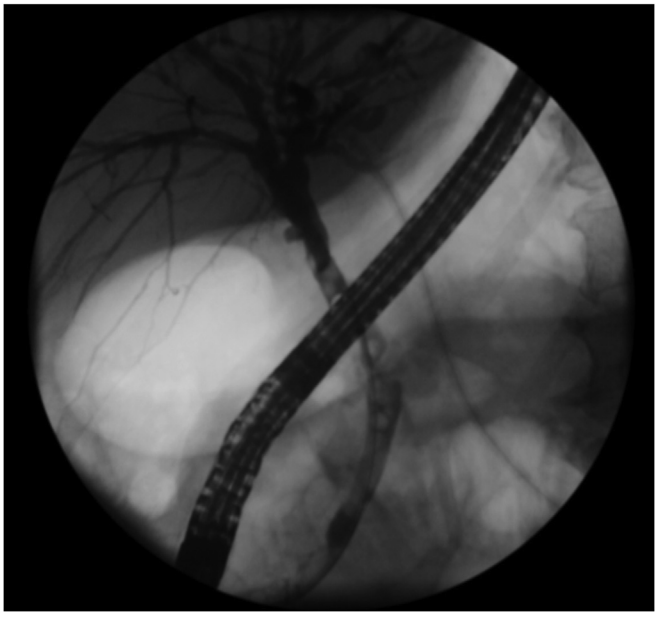

**Figure 2.** Filling defects noted along the mid to proximal bile duct.


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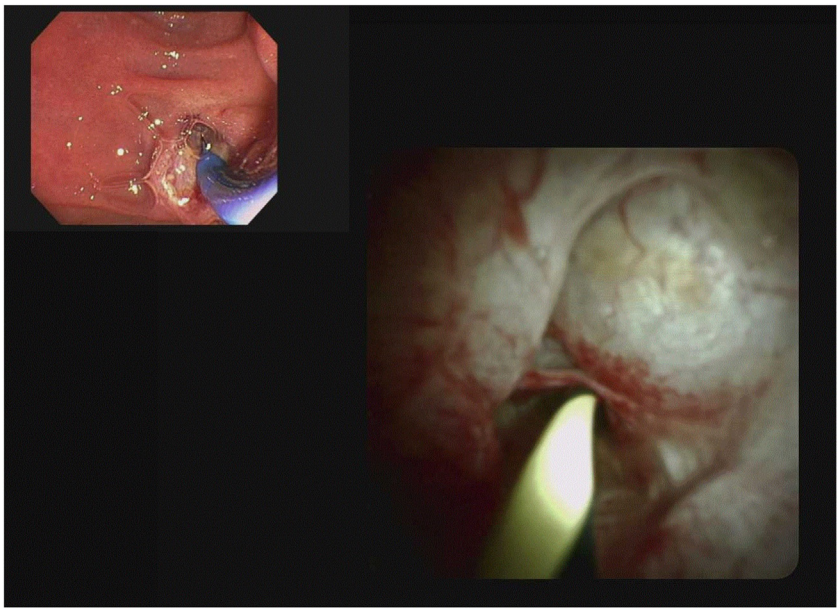

**Figure 3.** Cholangioscopy revealing dilated polypoid structures in the biliary tree with normal overlying mucosa.

## OUTCOME

His clinical history and endoscopic findings were reviewed with a multidisciplinary team. Surveillance was recommended with MRI in 6 months, which showed no change on interval follow-up (Fig. 4). The patient remains asymptomatic and undergoes yearly surveillance with MRI.

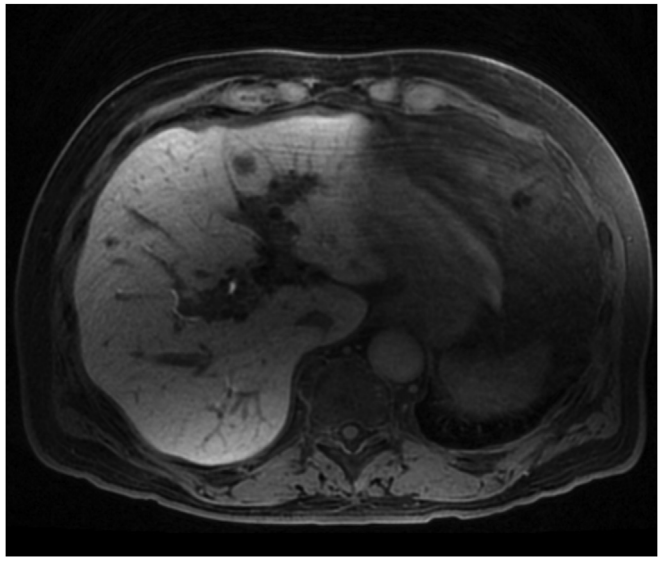

**Figure 4.** Liver acquisition with volume acceleration T1 phase MRI of the abdomen with 14-minute delay from gadoxetate disodium injection performed at 6-month follow-up revealing unchanged bilobar intrahepatic polypoid structures.

## DISCUSSION

This unusual case presents the workup and diagnosis of peribiliary cysts, which may be mistaken for a malignancy leading to unnecessary interventions.[1] A recent systematic review of the few reported cases described 3 types of peribiliary cysts: type 1 (87%), isolated intrahepatic; type 2 (7%), isolated extrahepatic; and type 3 (6%), both intrahepatic and extrahepatic (Fig. 5).[2] This patient had type 3 distribution, which is often grouped with type 1 for presenting most commonly in men with cirrhosis or portal hypertension. Although two-thirds of cases have been reported in Japan, peribiliary cysts can occur from underlying hepatic disease where alcohol abuse (like with this patient) is the most likely predisposing factor. Asymptomatic presentation, as in this case, occurs in 30%.[2] For those presenting with symptoms, these often are related to cirrhosis (52%) or biliary obstruction (19%).[2] Hepatobiliary malignancies have been noted in 17% of patients (most commonly, hepatocellular carcinoma), but these were exclusively in circumstances of cirrhosis or multiple intrahepatic cysts.[2] Autopsy reports note that 10% of cases will have epithelial changes in peribiliary glands; however, it is unclear whether these findings are associated with malignant degeneration.[3]

Although CT and MRI may be diagnostic in 48% and 64%, respectively, cholangioscopy may be necessary to characterize and access foci for biopsy to clench the diagnosis.[2] As opposed to choledochal cysts, peribiliary cysts

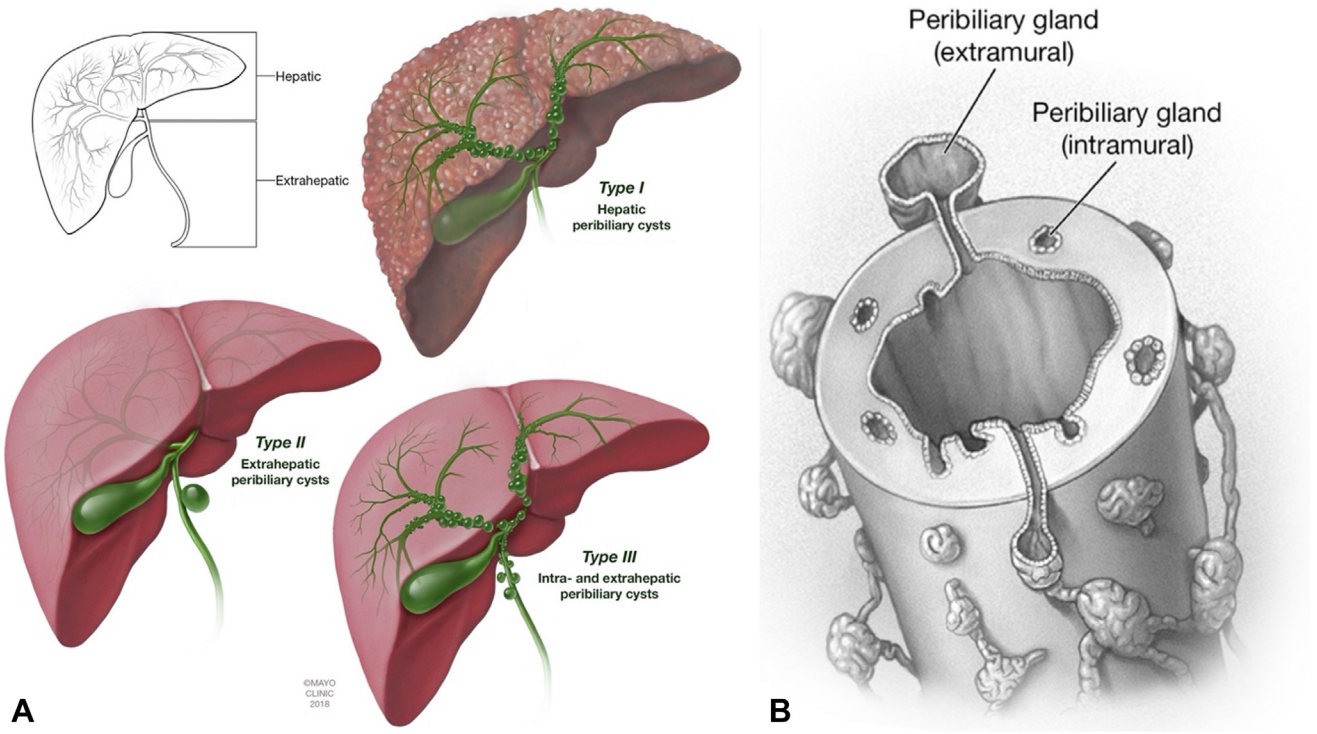


**Figure 5. A,** Graphical representation of the 3 types of peribiliary cysts. **B,** Histopathologic representation of peribiliary cysts. (Adapted from Bazerbachi et al.[2])

have intermittent saccular outpouchings along the biliary tree. Once malignancy can be excluded, the best treatment for type 1 and 3 peribiliary cysts is to address the underlying condition that resulted in cyst formation. In the case of this patient with both intra- and extrahepatic biliary cysts, that included alcohol cessation. For patients with symptomatic type 2 (extrahepatic) peribiliary cysts, surgical resection can be considered. In the absence of malignant features, surveillance should be considered given the unclear association between cyst formation and malignancy. Often, malignancy arises from the underlying liver disease rather than biliary degeneration; thus, surveillance for hepatocellular carcinoma may be necessary.

## DISCLOSURE

*The authors did not disclose any financial relationships.*

### REFERENCES

1. Lim J, Nissen NN, McPhaul C, Annamalai A, et al. Peribiliary hepatic cysts presenting as hilar cholangiocarcinoma in a patient with end-stage liver disease. J Surg Case Rep 2016;2016:rjw130.
2. Bazerbachi F, Haffar S, Sugihara T, et al. Peribiliary cysts: a systematic review and proposal of a classification framework. BMJ Open Gastroenterol 2018;5:e000204.
3. Sato Y, Harada K, Sasaki M, et al. Cystic and micropapillary epithelial changes of peribiliary glands might represent a precursor lesion of biliary epithelial neoplasms. Virchows Arch 2014;464:157-63.