Mucinous cystic neoplasm (MCN) of the liver is a biliary counterpart of pancreatic MCN, usually occurring in the liver, but occasionally extending to the extrahepatic bile duct [1,2]. We present a case of MCN with extrahepatic bile duct extension diagnosed by peroral video cholangioscopy (POVCS).

A 49-year-old woman was admitted with a week of vomiting without abdominal pain, and jaundice. Abdominal ultrasound (Fig. 1), contrast-enhanced computed tomography (Fig. 2), and magnetic resonance cholangiopancreatography (Fig. 3) revealed a multilocular perihilar cystic lesion with dilation of $B_2$, $B_3$, and $B_4$. Endoscopic retrograde cholangiography showed an elliptical defect from the left hepatic duct to the perihilar bile duct (Video 4). Intraductal ultrasonography demonstrated multilocular, cyst-in-cyst structures in the extrahepatic bile duct (Fig. 5). Subsequent POVCS (CHF-B260; Olympus, Tokyo, Japan) revealed a mass filling the bile duct covered with smooth and translucent bile duct mucosa, suggesting a subepithelial growth without bile duct communication (Fig. 6; Video 1). A directly visualized biopsy showed no evidence of malignancy. We diagnosed intraductal MCN of the liver based on the findings of the imaging modalities. The patient underwent left lobectomy and caudate lobectomy. A cholangiogram of the resected specimen showed no biliary communication. Histologically, the dilated ducts were filled with a polypoid cystic mass consisting of lining columnar epithelium and ovarian-like stroma, features consistent with MCN with low grade dysplasia. The tumor was located beneath the lining epithelium of the left hepatic duct and there was no communication between the ductal lumen and cystic spaces. There was no evidence of recurrence at 3-year follow-up. This is the first report of biliary intraductal MCN, suggesting an origin from Glisson’s capsule of the large hepatic bile duct. A recent report was the first to describe extrahepatic bile duct intraductal MCN using POVCS [3]. A cystic mass with smooth surface and compression due to distension without bile duct communication may be the characteristic findings of biliary intraductal MCN with POVCS.

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Competing interests: None

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Fig. 4 Endoscopic retrograde cholangiography showing an elliptical defect from the left hepatic duct to the perihilar bile duct.

Fig. 5 Intraductal ultrasonography showing multilocular, cyst-in-cyst structures in the extrahepatic bile duct.

Fig. 6 Endoscopic image showing the smooth and hyperpermeable bile duct mucosa, suggesting intraductal growth.

Video 1

Peroral video cholangioscopy (CHF-B260, Olympus, Tokyo, Japan) performed using a mother–baby system under fluoroscopic guidance.