Multiple intraductal papillary neoplasms of the bile ducts: a case report

Muhammad Raheel Anjum, MBBS, MRCP (UK), FCPS (Pak),[1] Bobby V. M. Dasari, MS, MSc, FRCS,[2] Shyam Menon, MSc, MD, FRCP[1]

Intraductal papillary neoplasms of the bile duct (IPN-B) are rare.[1] These tumors can obstruct bile flow either by luminal occlusion or by secretion of excessive mucin (mucinous subtype). No standard guidelines on management or surveillance of IPN-B exist. Surgical resection remains the definitive treatment of IPN-B.[2] The radiologic appearance of IPN-B, mimicking cholangiocarcinoma, can pose a significant diagnostic dilemma. Direct visualization with cholangioscopy may clarify the diagnosis and facilitate targeted biopsy.

A 75-year-old man presented with abdominal pain and progressive cholestatic jaundice. He had a history of laparoscopic cholecystectomy for adenocarcinoma of the gallbladder (pT1, pN0, R0) in 2010. He underwent MRCP (Fig. 1), which demonstrated dilated intrahepatic ducts in both right and left lobes of the liver with multiple intraluminal filling defects, believed to be potentially arising from the duct walls. Differential diagnosis included metastasis from previous gallbladder cancer, primary sclerosing cholangitis, Caroli disease, and hepatolithiasis. The extrahepatic ducts were normal.

Because the diagnosis was unclear and the patient needed biliary decompression, he underwent ERCP and Spyglass (Boston Scientific, Marlborough, Mass) cholangioscopy (Video 1), which demonstrated fleshy intraluminal polypoidal mass lesions (Fig. 2) in the right anterior and right posterior intrahepatic ducts and at the origin of the left main intrahepatic duct (Fig. 3). These lesions were discrete and had papillary projections with evidence of mucin in the duct. Biopsy results confirmed IPN-B with high-grade dysplasia.

Liver transplantation was considered inappropriate because of his age and comorbidity (significant ischemic heart disease). Intraoperative cholangioscopy demonstrated another lesion in a duct in segment IV-b of the liver. The patient underwent an extended right hepatectomy with an access loop (Fig. 4) to facilitate access to the left intrahepatic ductal system for future surveillance. Postoperative histology results also confirmed IPN-B with high-grade dysplasia.

The optimal management and surveillance of IPN-B is unclear. Limited liver resection followed by surveillance of the remnant liver with imaging and cholangioscopy seems a reasonable strategy. Therapeutic options in the future may involve cholangioscopy-assisted local resection/ablation (Video 1, available online at www.VideoGIE.org).

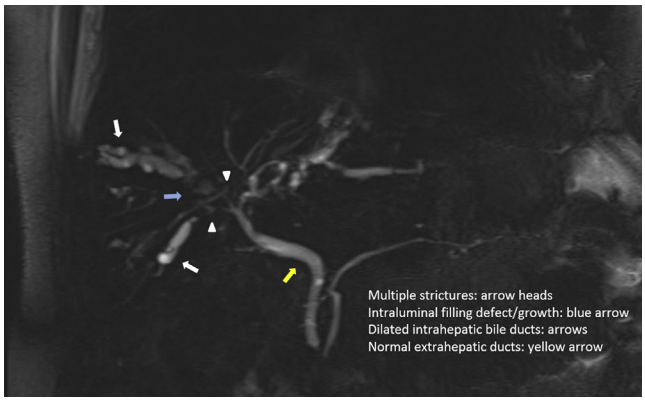

Figure 1. MRCP image demonstrating multiple strictures *(arrowheads)*, intraluminal filling defect/growth *(blue arrow)*, dilated intrahepatic bile ducts *(arrows)*, and normal extrahepatic ducts *(yellow arrow)*.

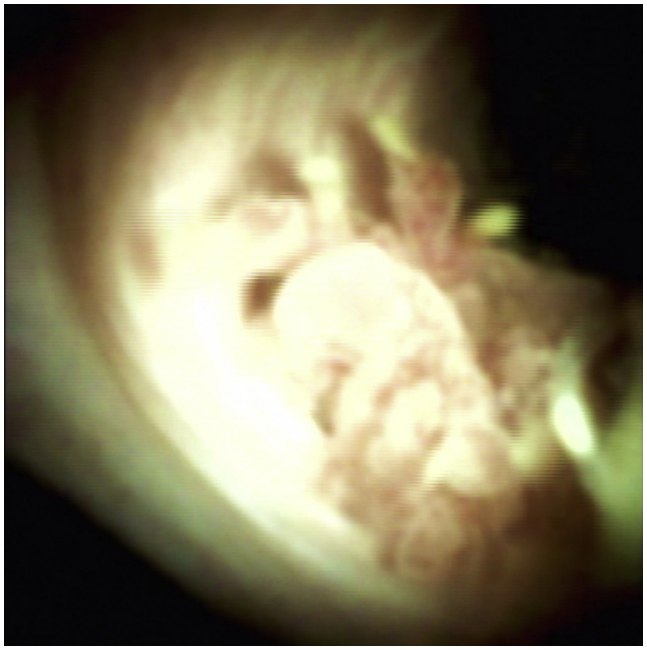

Figure 2. Cholangioscopic view of intraductal papillary neoplasm of the bile duct lesion.

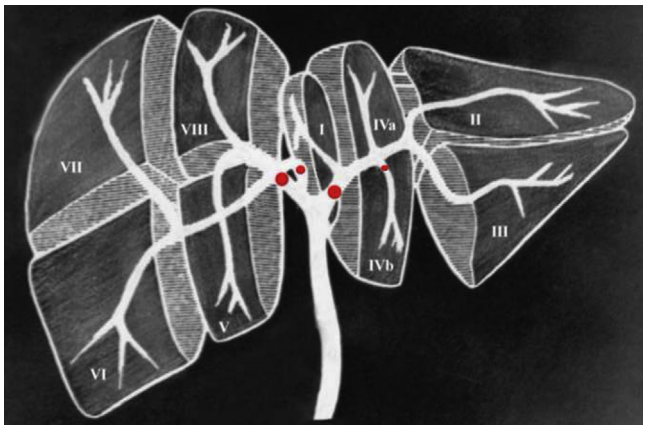

Figure 3. Diagrammatic representation of positions of intraductal papillary neoplasms of the bile duct (IPN-B) in relation to biliary tree.

DISCLOSURE

All authors disclosed no financial relationships.

Abbreviation: IPN-B, intraductal papillary neoplasms of the bile duct.

REFERENCES

1. Lim JH, Yoon KH, Kim SH, et al. Intraductal papillary mucinous tumor of the bile ducts. Radiographics 2004;24:53-66.
2. Ohtsuka M, Shimizu H, Kato A, et al. Intraductal papillary neoplasms of the bile duct. Int J Hepatol 2014;2014:459091.

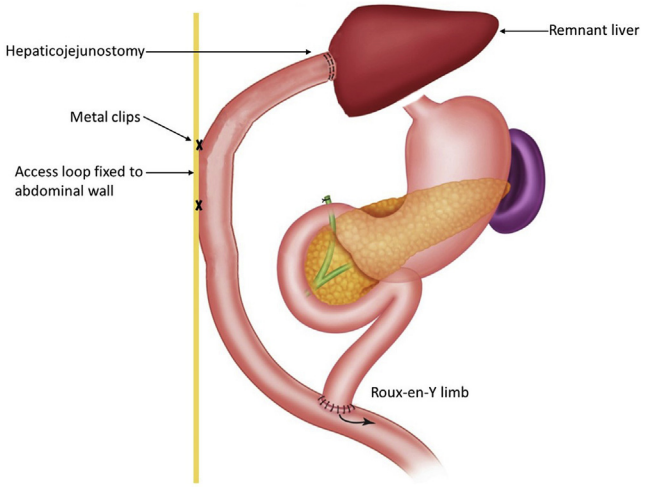

Figure 4. Diagrammatic representation of Roux-en-Y hepaticojejunostomy with access loop fixed to abdominal wall.

Department of Gastroenterology, The Royal Wolverhampton NHS Trust, Wolverhampton, United Kingdom (1), University Hospitals Birmingham NHS Foundation Trust, Birmingham, United Kingdom (2).

https://doi.org/10.1016/j.vgie.2020.08.008