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GI IMAGE

Extrahepatic Neuroendocrine Tumor Causing Biliary Obstruction

Jonathan P. Rehfuss[1] · Ryan M. Thomas[1,2]

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Case

A 57-year-old man with a past medical history of hyperlipidemia and gastroesophageal reflux disease presented for surgical consultation after a comprehensive work up for abdominal pain. He reported vague abdominal discomfort and a sensation of bloating but no other symptoms. He denied a history of jaundice, liver disease, or malignancy and had no prior abdominal operations. Physical examination revealed a well-nourished, healthy-appearing middle-aged man with no abdominal distention or tenderness, no palpable lymphadenopathy, and no jaundice. Hepatic function panel as well as the tumor markers CEA, CA19-9, and AFP was normal. Computed tomography revealed significant right intrahepatic ductal dilatation and a 1-cm hilar mass associated with the right hepatic duct (Fig. 1). An attempted endoscopic retrograde cholangiopancreatography to obtain a tissue biopsy had failed because of a technical inability to cannulate the ampulla, and endoscopic ultrasound failed to reveal a mass.

Based on imaging, the mass was suspected to be a Bismuth-Corlette class 3A cholangiocarcinoma without evidence of vascular invasion or systemic disease that would preclude resection, and the patient was brought to the operating room for potential resection. Intraoperative findings included a firm mass at the hepatic ductal confluence with right hepatic duct involvement necessitating a right hepatectomy with common bile duct resection and Roux-en-Y hepaticojejunostomy. Frozen sections of the common bile duct and distal left hepatic duct margins were negative for malignancy. Postoperatively, the patient recovered well and was discharged 7 days following surgery. Interestingly, the final pathology revealed a 1-cm well-differentiated neuroendocrine tumor with a very low proliferation index (Ki-67 < 1%) emanating from the right hepatic duct with luminal epithelial erosion and muscularis propria involvement (Figs. 2 and 3). All margins and lymph nodes were free of tumor.

Approximately 90% of biliary tract tumors are cholangiocarcinoma, a rare malignancy with an incidence of approximately one or two cases per 100,000 annually in the USA. Much rarer still are neuroendocrine tumors (NETs) of the extrahepatic biliary tract which comprise less than 1% of all gastrointestinal NETs. Only 20% of these tumors are well-differentiated. Unlike pancreatic NETs, they almost never display hormonal function. A review published in 2014 determined that the median age of presentation was 47 with a 1.6/1 female predominance and 90% had symptoms which were primarily related to mass effect by the tumor or metastases rather than hormonal secretion.

✉ Ryan M. Thomas
ryan.thomas@surgery.ufl.edu

[1] Department of Surgery, College of Medicine, University of Florida, PO Box 100109, Gainesville, FL 32610, USA

[2] North Florida/South Georgia Veterans Health System, Gainesville, USA

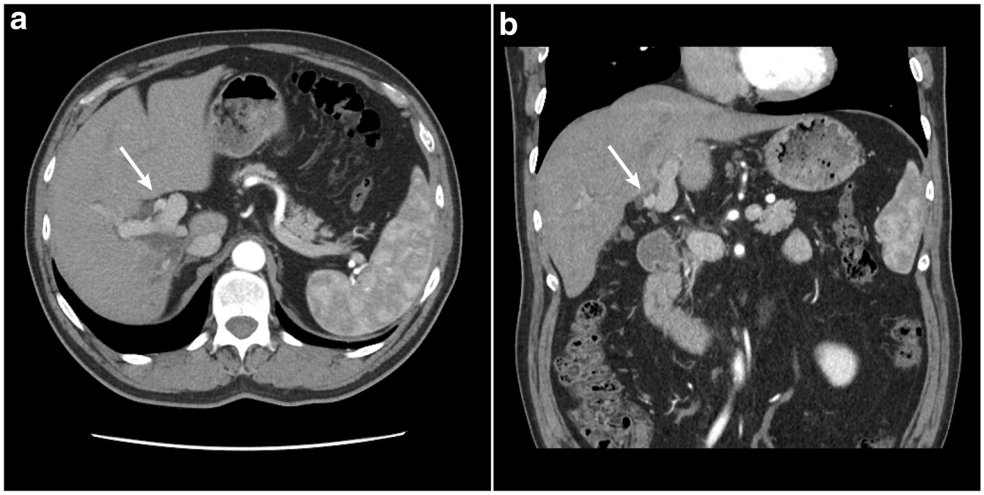

Fig. 1 Axial (**a**) and coronal (**b**) computed tomographic images demonstrate a well-circumscribed 1-cm mass at the hepatic ductal confluence denoted by the arrow

Approximately one third of patients with extrahepatic bile duct NETs have metastases to either local lymph nodes or the liver.[1]

The preoperative diagnosis of a biliary tract mass as an NET is difficult because these tumors tend to produce neither hormonal symptoms nor a detectable serum marker, and there are no known hallmark radiographic characteristics.[2] Endoscopic biopsy can provide a diagnosis preoperatively; however, the vast majority of published cases have been diagnosed only after surgical resection. Although extrahepatic biliary NETs tend to grow slowly, surgical resection is the only curative treatment.[3]

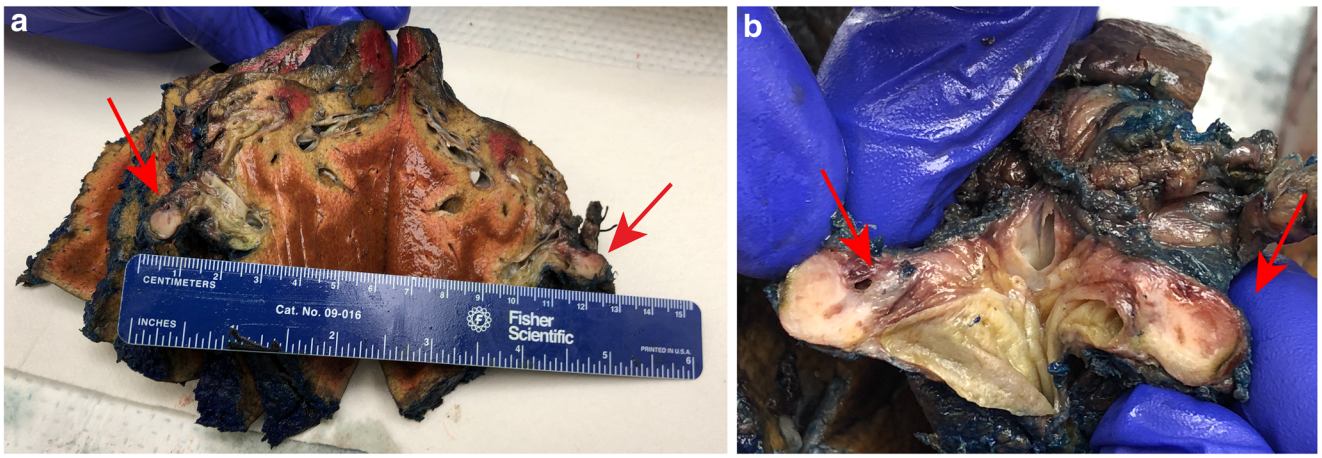

Fig. 2 Gross pathology of the surgical specimen, consisting of the right hepatic lobe, common bile duct, and hepatic confluence (**a**), demonstrates a 1-cm mass (arrows) emanating from the right hepatic duct at the ductal confluence (**b**)

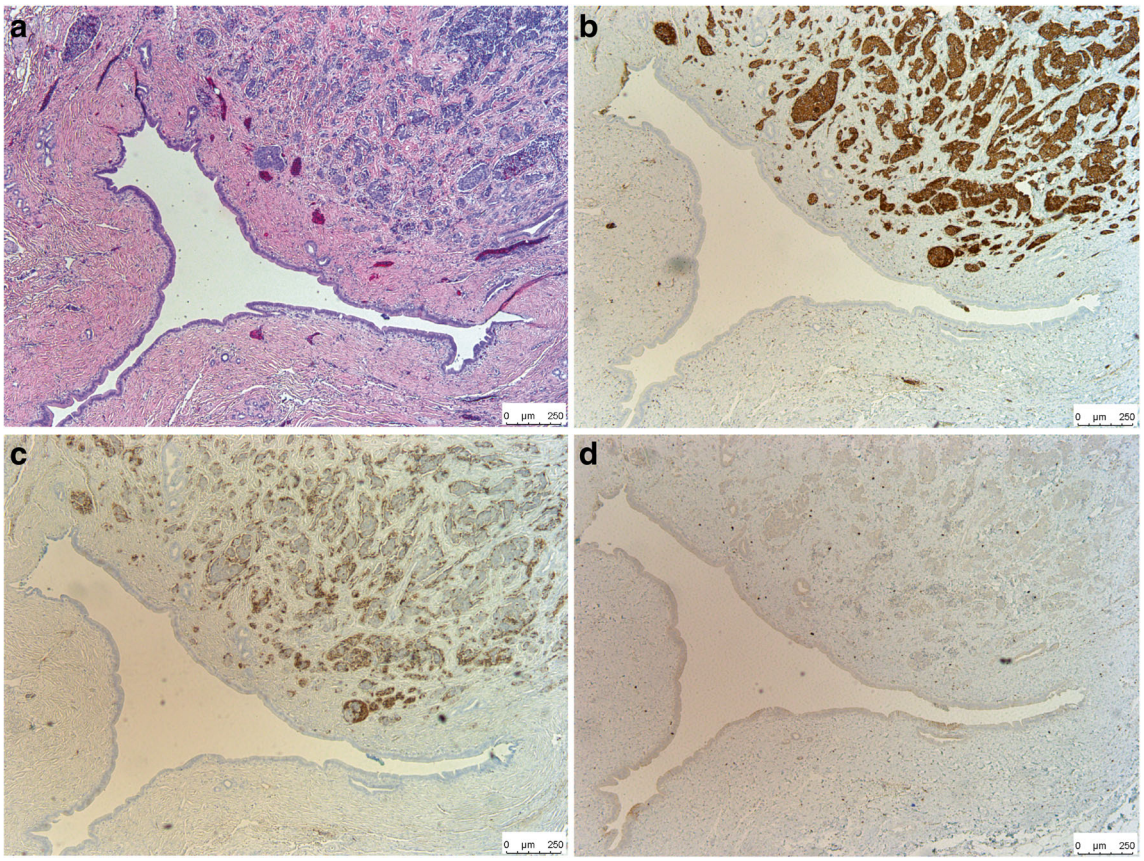

Fig. 3 Hematoxylin and eosin (**a**) staining demonstrates luminal epithelial erosion and muscularis propria involvement by the tumor. Strong synaptophysin (**b**) and chromogranin (**c**) staining provide the diagnosis of a neuroendocrine tumor, while very weak Ki-67 (**d**) staining indicates its low-grade nature

Radiographic surveillance is of uncertain utility because of the limited data regarding the natural history of biliary NETs. This stands in contrast to the management of small, non-functional pancreatic NETs, where serial imaging is an accepted practice.

This case illustrates an uncommon cause of a very common clinical presentation that surgeons encounter. Because of their extreme rarity, extrahepatic biliary NETs are poorly understood, and their optimal treatment is not well-defined. Awareness and reporting of biliary NETs may contribute to a better understanding and development of more individualized treatment sequencing strategies for this rare tumor.

Author Contribution Both Drs. Thomas and Rehfuss contributed substantially to the conception, drafting, and revision of the submitted work, and both authors approve the final version and agree to be accountable for all aspects of this work.

References

1. Michalopoulos N, Papavramidis TS, Karayannopoulou G, Pliakos I, Papavramidis ST, Kanellos I: Neuroendocrine tumors of extrahepatic biliary tract. *Pathol Oncol Res* 2014, 20:765–775.
2. Modlin IM, Sandor A: An analysis of 8305 cases of carcinoid tumors. *Cancer* 1997, 79:813–829.
3. Hubert C, Sempoux C, Berquin A, Deprez P, Jamar F, Gigot JF: Bile duct carcinoid tumors: an uncommon disease but with a good prognosis? *Hepatogastroenterology* 2005, 52:1042–1047.