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Extrahepatic biliary obstrution secondary to neuroendocrine tumor of the common hepatic duct

Faraz A. Khan*, Anastasia Stevens-Chase, Rahman Chaudhry, Asra Hashmi, David Edelman, Donald Weaver

Department of Surgery, Wayne State University/Detroit Medical Center, 6C UHC, 4201 St. Antoine, Detroit, MI 48226, USA

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ABSTRACT

INTRODUCTION: Primary neuroendocrine tumors (NET) of the extrahepatic biliary tree are a rare entity with less than 100 reported cases in the literature.
PRESENTATION OF CASE: Here, we report a case of NET of the extrahepatic bile duct in a 64-year-old male patient presenting with painless jaundice, direct hyperbilirubinemia, and mildly elevated transaminases. Diagnostic workup with an ultrasound revealed dilation of the intrahepatic biliary ducts, without cholelithiasis or choledocholithiasis. Additional cross sectional imaging identified a stricture at the confluence of the common hepatic and cystic duct junction. Given lack of additional findings presumptive diagnosis of localized klatskin's tumor was made. The patient subsequently underwent resection of the common bile duct and roux-en-y hepaticojejunostomy reconstruction. Final pathologic diagnosis showed G2 well-differentiated NET of the extrahepatic bile duct, measuring $1.3 \times 1.1 \times 1$ cm.
DISCUSSION: When a patient is evaluated for a primary bile duct neoplasm, differentiation between cholangiocarcinoma and an unusual bile duct tumor, such as a NET is very difficult before surgical resection and histologic review.
CONCLUSION: NET of the extrahepatic biliary tree are a rare entity. Typical presentation is with painless jaundice and other symptoms related to obstruction of the biliary tree and the diagnosis is usually made post-operatively.

1. Introduction

Neuroendocrine tumors (NETs) are thought to originate from enterochromaffin, or Kulchitsky cells, of the gastroenteropancreatic tract. These cells presumably undergo a pre-malignant intestinal metaplasia from inflammation. Enterochromaffin cells are found in highest proportion in the small intestine, and rarely within the biliary ducts. This low proportion of precursor cells corresponds with the low incidence of extrahepatic biliary neuroendocrine tumors (NET) [1,2,9]. The presenting signs and symptoms of extrahepatic NETs are secondary to the obstruction of the normal flow of bile, thereby causing hyperbilirubinemia and jaundice. Here we report case of a patient with extrahepatic NET in line with the SCARE criteria [3].

2. Presentation of case

64-year-old male presented with 2-week history of jaundice, pruritus, acholic stools, and tea colored urine. He also noted poor oral intake and approximately 16-pound weight loss during that time. He denied any abdominal pain, nausea, or vomiting and had no family history of hepatopancreatobiliary malignancy. On examination, the patient's skin was jaundiced, his abdomen was soft, non-tender, non-distended and no masses were palpable. Laboratory test were suggestive of obstructive jaundice with Total and Direct Bilirubin elevated at 1.6 and 1.1 mg/dL respectively. Right upper quadrant ultrasound showed dilation of the intrahepatic biliary ducts, without cholelithiasis or choledocholithiasis. The patient subsequently underwent computerized tomography (CT) scan of the abdomen and pelvis with intravenous and oral contrast, which showed diffuse mild to moderate intrahepatic biliary ductal dilatation, with a distended gallbladder. The common bile duct (CBD) at the level of the gallbladder measured 1.4 cm, with a distal abrupt caliber change and a soft tissue density mass measuring 1.3 cm was noted in the hepatic duct (Fig. 1). These findings were considered concerning for a Klatskin tumor/cholangiocarcinoma, without

* Corresponding author.
E-mail address: dr.farazali@gmail.com (F.A. Khan).

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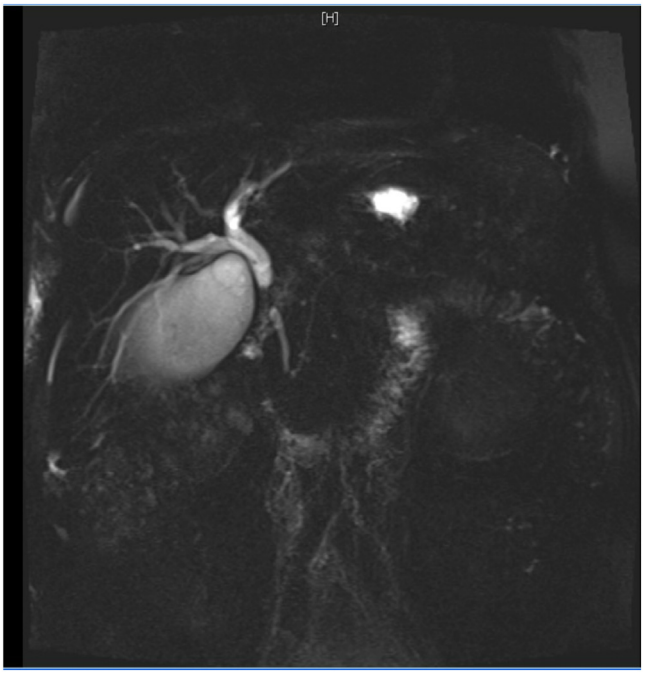

Fig. 1. CT scan demonstrating distal abrupt caliber change and a soft tissue density mass measuring 1.3 cm was noted in the hepatic duct.

evidence of lymphadenopathy or metastasis. Magnetic resonance cholangiopancreatography (MRCP) validated the same findings. Moreover Carbohydrate antigen 19-9 (CA 19-9) was checked and found to be elevated at 50 U/mL.

The patient was then referred to the Department of Surgical Oncology at a teaching hospital and offered surgical resection. The patient underwent resection of the CBD, cholecystectomy, end to side roux-en-y hepaticojejunostomy, and portal lymphadenectomy, with a presumed pre-operative diagnosis of hilar cholangiocarcinoma. Intraoperative frozen sections of resection margins were negative for tumor. Dissected lymph nodes in the porta hepatis were normal appearing. The patient's immediate postoperative course was uncomplicated and he was discharged home on POD 4. The final pathologic results showed a G2 neuroendocrine tumor of the extrahepatic CBD, measuring 1.3 × 1.1 × 1 cm (Fig. 3). Histologically, the tumors cells were well differentiated (Fig. 2A) with 13–15 mitoses per 10 high power fields (Fig. 2B). The tumor showed perineural invasion but was negative for lymphovascular invasion. Immunohistochemical staining showed all the tumor cells were strongly positive for chromogranin, synaptophysin (Fig. 2C), and CD 56. Immunohistochemistry for Ki-67 showed a proliferation index of approximately 5 (Fig. 2D). The histologic findings and the immunoprofile of the neoplasm were consistent with neuroendocrine tumor, well differentiated, G2 (grade 2). Given the rarity of the clinical diagnosis verbal informed consent was obtained from the patient to publish this case in a scientific journal.

3. Discussion

Less than 100 cases of extrahepatic biliary NET have been reported in the literature to date. A review of the recent literature

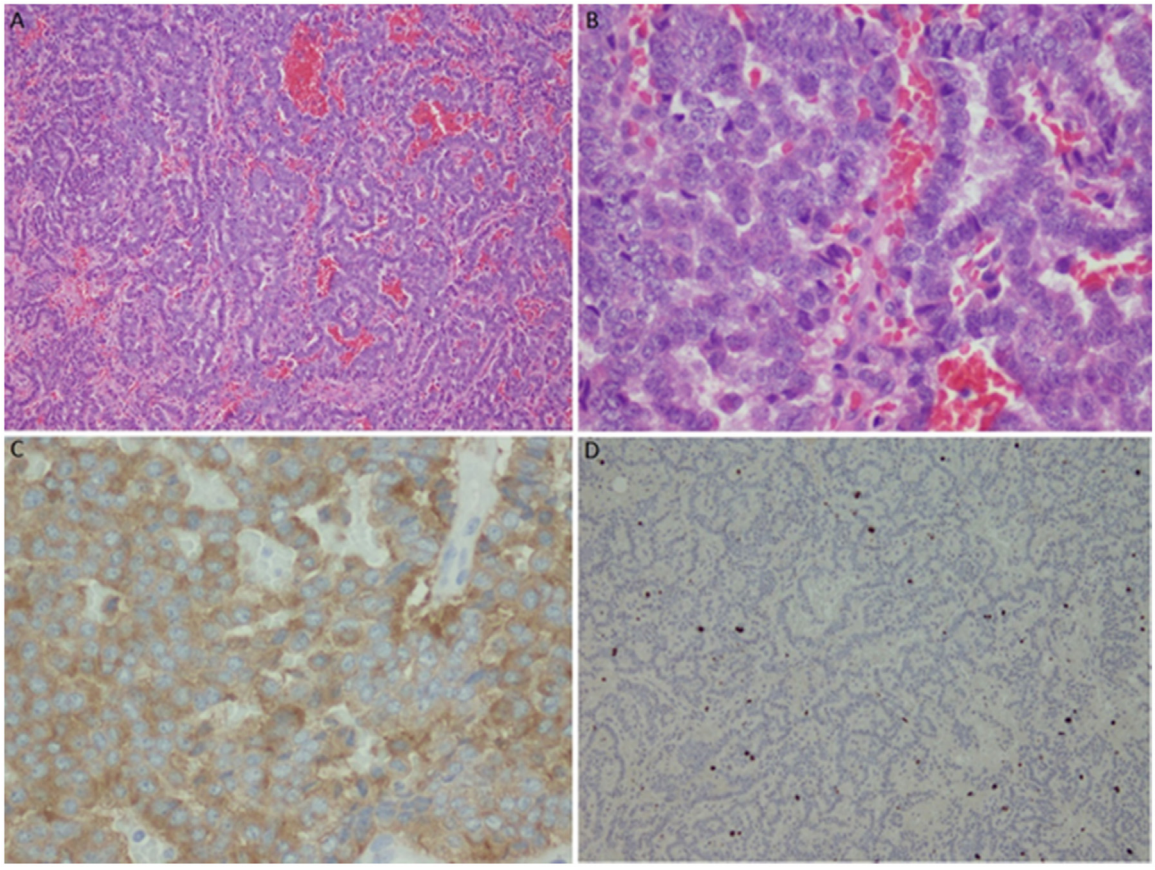

Fig. 2. A: Histologic findings of the gallbladder neoplasm: Tumor cells arranged in a trabecular pattern with scant intervening fibrovascular stroma (Hematoxylin and eosin (HE), ×10); B. The tumors cells showed medium to large hyperchromatic nuclei with coarse granular chromatin, occasional nucleoli, small to moderate amphophillic cytoplasm, and scattered mitotic figures (HE, ×40); C: The tumor cells were strongly positive for synaptophysin (×40); D: 5% of the tumor cells were positive for Ki-67 (×10).