



Neuroendocrine Tumor in the Biliary Tract Presenting With Cholangitis

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Introduction

Neuroendocrine tumors (NETs) of the biliary tract account for less than 2% of all gastrointestinal cancers with an incidence of 0.32%. Extrahepatic biliary NETs are mostly found incidentally in the distal common bile duct (CBD) but in rare cases, they may present with obstructive jaundice. We present a case of an elderly male with symptoms of biliary obstruction and acute cholangitis secondary to a NET in the proximal bile duct.

Case Presentation

HPI: 64-year-old male presented with epigastric abdominal pain, vomiting and fever. The patient reported an unintentional 50-pound weight loss over the past year.

Past Medical History:

- Hypertension

Review of Systems:

- Abdominal Pain, Nausea, Vomiting, Fever

Physical Exam:

- Fever 100.9F, Tachycardia to 118 beats/min.
- Tender to palpation in Epigastric/RUQ

Diagnostic Labs & Biochemistry

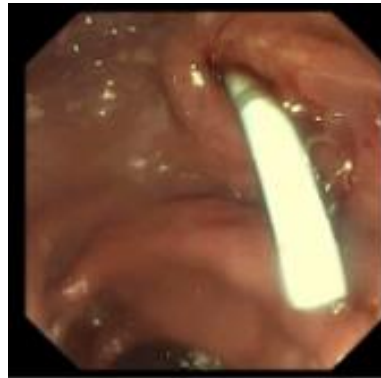
white blood cell count of 21,000 u/L, AST 303 U/L, ALT 563 U/L, alkaline phosphatase 300 U/L, total bilirubin 1.9 mg/dL.

Diagnostic Imaging & Intervention

- CT abdomen showed intrahepatic ductal dilatation to 1.1 cm, and a 1.2 x 1.1 cm low attenuating lesion within the left lobe of the liver and wall thickening of the proximal jejunal loops. Piperacillin-tazobactam was started for acute cholangitis.



- Endoscopic ultrasound showed duct dilation in the left hepatic lobe and ERCP showed ulceration of major papillae. Following biliary sphincterotomy, exploration revealed severe stenosis of the main bile duct and a 12 mm polypoid lesion in the upper third of the CBD (Figure 1).



- A 10fr x 9cm stent with a single external and internal flap was placed. Following a 7-day course of antibiotics and symptom resolution, the patient was discharged with a plan for elective surgery. Left hepatectomy, CBD resection, and Roux-en-Y hepaticojejunostomy were performed 6 months after hospitalization. At 2 months follow up in clinic, the patient was asymptomatic.

- Histology showed well differentiated NET with trabecular and organoid pattern, positive chromogranin and synaptophysin with Ki-67 index of 3%. Special staining was also positive for reticulin and trichrome. 6 months after hospitalization. At 2 months follow up in clinic, the patient was asymptomatic.

Discussion

- Biliary NETs are found in the common hepatic duct, CBD and the cystic duct.
- Biliary NETs are twice more common in females, with a mean age of 47 years old. Although most cases are incidental, presenting symptoms are jaundice and pruritis.
- Diagnosis requires histologic evidence and positive expression of chromogranin A, synaptophysin, CD56, or neural cell adhesion molecule.
- Treatment is surgical excision with Roux-en-Y hepaticojejunostomy.
- Since biliary NETs uncommonly present with obstructive jaundice, this case presentation highlights the need for a broad differential diagnosis in evaluating patients with obstructive jaundice.