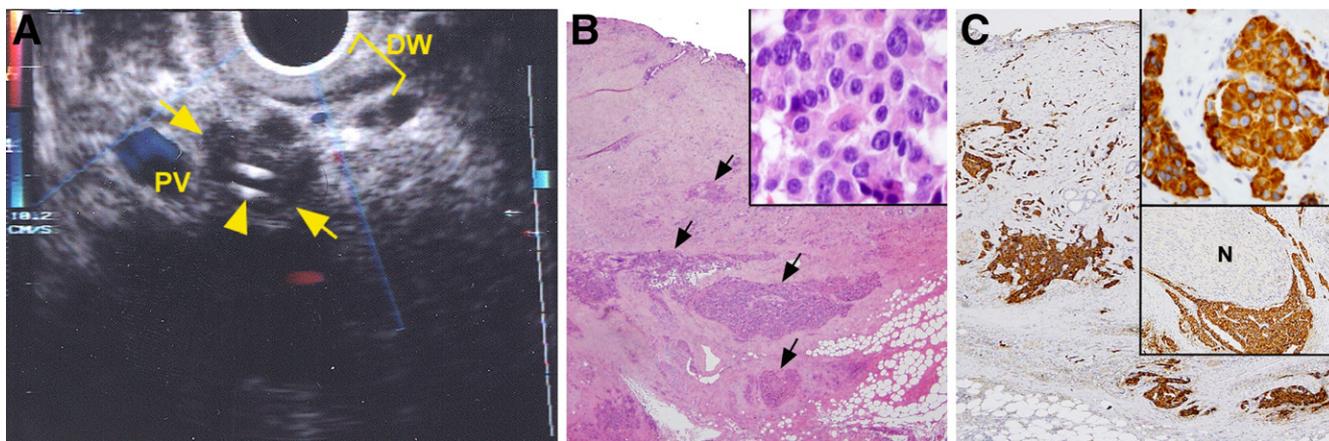


Image of the Month

Common Bile Duct Carcinoid Mimicking the Clinical, EUS, and ERCP Findings of Cholangiocarcinoma: A Rare but Potentially Curable Cause of Obstructive Jaundice

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A 42-year-old man presented with right upper quadrant pain and tenderness, progressive jaundice, and 20-kg weight loss, without episodic flushing, wheezing, or diarrhea, for 2 months. Serum levels: lipase, 328 U/L; aspartate aminotransferase, 54 U/L; alanine aminotransferase, 119 U/L; alkaline phosphatase, 528 U/L; and total bilirubin, 3.9 mg/dL. Abdominal ultrasound revealed a 1.0-cm wide choledochus, without intrahepatic lesions. Endoscopic retrograde cholangiopancreatography (ERCP) revealed mid-distal choledochal stricture, with mild proximal choledochal dilatation. The stricture was dilated endoscopically, and a 7 French stent was deployed. Cross-sectional endoscopic ultrasonography (EUS) with Doppler revealed a 1.5 × 1.2 cm, irregularly spherical, hypoechoic mass (Figure A; arrows) surround choledochal mass near ampulla; arrowhead, bright, hyperechoic, parallel lines from choledochal stent within dilated choledochal stricture; PV, portal vein; DW, duodenal wall). Cytologic examinations of ductal brushings obtained at ERCP and needle biopsies obtained at EUS were nondiagnostic. Serum carbohydrate antigen (CA) 19-9 level was 50 U/mL (minimally elevated, normal <35 U/mL).

Laparotomy, for presumed cholangiocarcinoma, revealed a 1.8-cm long, firm, mid-distal choledochal mass with choledochal thickening. A 2.5-cm area surrounding the mass was resected. Frozen sections of the resected specimen revealed locally invasive cancer. The patient then underwent pancreaticoduodenectomy. Microscopic examination of permanent sections of resected specimen revealed low-grade, circumferential,

choledochal carcinoid (World Health Organization nomenclature: choledochal neuroendocrine tumor-carcinoid), without tumor at resected margins or in resected lymph nodes (stage 2-T2N0M0) (Figure B, low-power photomicrograph shows multiple islands with prominent, darkly staining tumor cells [arrows] representing carcinoid lesions extending deeply into choledochal wall; high power inset shows cluster of tumor cells with large, eccentric nuclei and eosinophilic cytoplasm; H&E). The tumor stained strongly, diffusely positive for chromogranin and synaptophysin, but negative for epithelial membrane antigen (EMA), S100, Mart 1, CD31, and CD34, an immunohistochemical pattern characteristic for carcinoid (Figure C, immunohistochemical synaptophysin stain reveals

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Figure A graciously provided by Edward Yousif, MD, Division of Gastroenterology, William Beaumont Hospital, Royal Oak, Michigan, who performed and interpreted the endoscopic ultrasound on this patient.

Figures B and C graciously supplied by Mitul Amin, MD, Department of Pathology, William Beaumont Hospital, Royal Oak, Michigan.

Conflicts of interest

The authors disclose no conflicts.

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extensive synaptophysin positivity [*brown pigmentation*]; high power [*upper inset*] reveals individual synaptophysin-staining carcinoid cells). Tumor exhibited striking perineural invasion (Figure C, *lower inset*, carcinoid enveloping nerve [N]). The patient did well, was discharged 9 days postoperatively, and has been asymptomatic without recurrent tumor during 9 years of follow-up.

Primary extrahepatic biliary duct carcinoids are rare. Literature review identified about 30 cases, including 20 choledochal cases.¹⁻³ Rarity is attributed to a paucity of argentaffin/Kulchitsky cells, presumed carcinoid cell progenitor, within biliary tissue. Patients may present incidentally, with jaundice and abdominal pain^{1,2} or with generalized elevations of liver function tests,¹ as presently reported.

Biliary carcinoid is typically unsuspected preoperatively and diagnosed by histology of resected specimen. As currently occurred, cytologic examinations of brushings or superficial biopsies obtained at ERCP, or even cholangioscopy, are generally

nondiagnostic due to submucosal tumor location. Pancreatoduodenectomy is the surgical procedure of choice.¹ Five-year survival was 41% in 1 study.³ This case illustrates that choledochal carcinoid may mimic clinical, EUS, and ERCP findings of cholangiocarcinoma; that preoperative brushings or biopsies are relatively insensitive; and that aggressive pursuit of surgery can sometimes lead to long-term survival or cure of this malignancy.

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