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A rare cause of extrahepatic biliary tract stricture: a neuroendocrine carcinoma

Roberto Oleas, Juan I. Olmos, Juan Alcívar-Vásquez and Carlos Robles-Medranda

Gastroenterology and Endoscopy Division. Instituto Ecuatoriano de Enfermedades Digestivas (IECED). Guayaquil, Ecuador

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Correspondence: Carlos Robles-Medranda

e-mail: carlosoakm@yahoo.es

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The case was an 82-year-old Hispanic female who complained of painless jaundice and weight loss. Endoscopic retrograde cholangiopancreatography (ERCP) showed a distal common bile duct (CBD) stricture with dilatation of the intra and extra-hepatic bile ducts (Fig. 1). Endoscopic ultrasound (EUS) detected two mediastinal lymphadenopathies, pleural effusion, ascites and peritoneal carcinomatosis. The CBD and intrahepatic ducts were dilated with a distal stricture associated with a 30 mm hypoechoic lesion with portosplenic vein involvement. EUS-fine-needle aspiration of the lymph nodes and peritoneal nodules was performed. Per-oral cholangioscopy (SpyGlass™, Boston Scientific®, Malborough, USA) detected an irregular, ulcerated, distal biliary lesion with signs of neovascularity (Fig. 2A). Probe-based confocal laser endomicroscopy (pCLE) of the lesion was suggestive of neoplasia (Fig. 2B). A partially covered metal stent was deployed and the patient died one month-later. Histopathology described a m ixed-neuroendocrine carcinoma of the CBD with a positive immunohistochemical analysis for chromogranin, cytoketarins and Ki-67 (Fig. 3).

Neuroendocrine carcinomas (NECs) in the extrahepatic biliary tract (EHBT) are a rare condition with a poor prognosis (1). Preoperative diagnosis of NECs in the EHBT is challenging and should be suspected as a Klatskin-mimicking lesion in order to avoid a misdiagnosis and provide appropriate treatment (2,3). MRCP and EUS fail to distinguish NECs from other bile duct tumors. Cholangioscopy with pCLE-guided biopsy and immunohistochemical analysis provided a definitive diagnosis of NECs.

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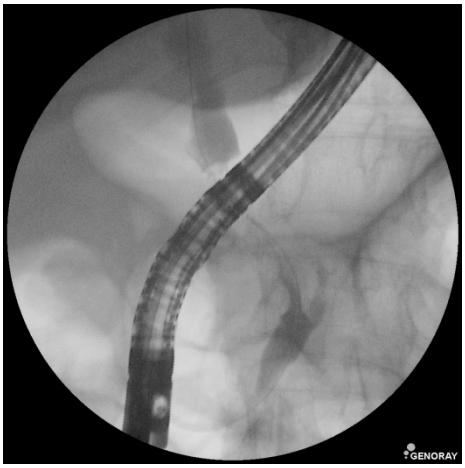

Fig. 1. Endoscopic retrograde cholangiopancreatography (ERCP) showing a distal common bile duct (CBD) stricture with dilatation of the intra and extra-hepatic bile ducts.

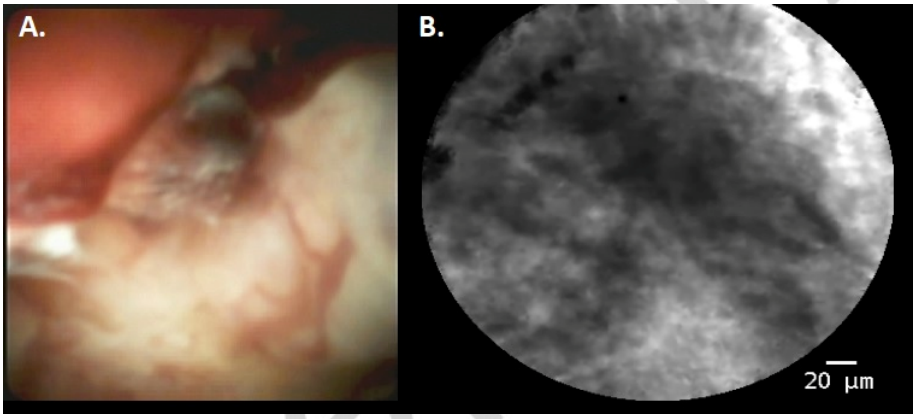

Fig. 2. A. Per-oral cholangioscopy of the bile duct showing an ulcerated mucosa with signs of neovascularity. **B.** *In vivo* probe-based confocal laser endomicroscopy showing a trabecular epithelial pattern (circumscribed clusters of cells in a trabecular growth pattern separated by vascular or fibrous cords).

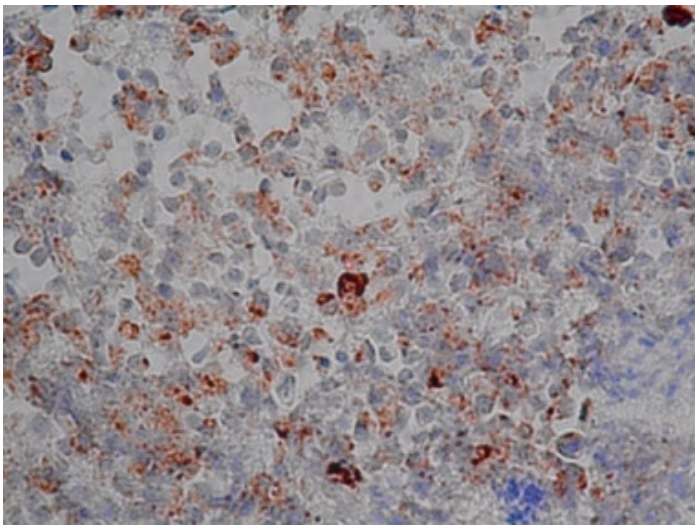

Fig. 3. Immunohistochemical analysis of the biliary stricture showing positivity for chromogranin.

COMMENTARY

Mixed adenoneuroendocrine carcinoma (MANEC) is a rare subtype of neuroendocrine neoplasm consisting of both adenocarcinomatous and neuroendocrine cells. Each component must account for at least 30 % of the lesion (1). MANEC in the EHBTs is extremely rare. Currently, the diagnosis relies on histopathological examination and requires confirmation with immunohistochemical methods (synaptophysin, chromogranin and CD56) and the determination of proliferation rates (Ki-67 > 20 % implies poor prognosis). Echoendoscopic diagnosis of EHBT-MANEC has been described (2). Cholangioscopy with pCLE-guided biopsy may help to achieve a diagnosis.

Maite Betés

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