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EDUCATION AND IMAGING

Hepatobiliary and Pancreatic: An unusual cause of biliary obstruction

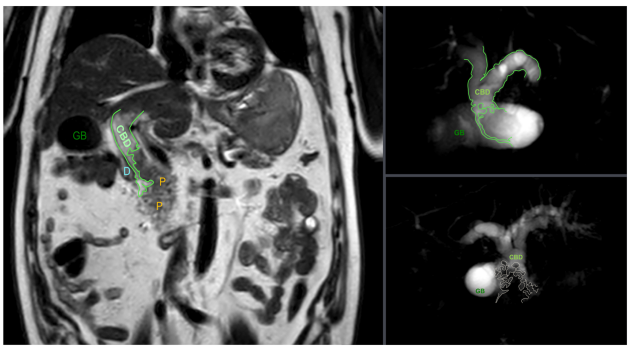

Figure 1 MRCP showed an irregular stenosis of the CBD with upstream dilation. CBD, common bile duct; D, duodenum; GB, gallbladder; P, pancreas.

A 41-year-old man, with no relevant personal history, presented with postprandial fullness, nausea, and weight loss in the last 2 years. On physical examination, he had mild jaundice. Laboratory examination revealed conjugated hyperbilirubinemia (total bilirubin 3.62 mg/dL, conjugated bilirubin 2.60 mg/dL) and cytocholestasis with cholestatic predominance (AST 102 U/L, ALT 144 U/L, FA 330 U/L, GGT 681 U/L).

An abdominal computed tomography followed by magnetic resonance cholangiopancreatography (MRCP) showed common bile duct (CBD) stenosis, without mural thickening or intraluminal mass, and intrahepatic and extrahepatic duct dilation. There were no lesions in the hepatic or pancreatic parenchyma (Fig. 1).

Endoscopic retrograde cholangiopancreatography (ERCP) showed a heterogeneous filling defect in the distal half of the CBD with upstream dilatation. Biliary sphincterotomy and exploration with a balloon were performed, and tumor tissue fragments with a villous appearance and necrotic areas were obtained. Cholangioscopy allowed the identification of a friable villous vegetative lesion in the middle third of the CBD leading to luminal narrowing (Fig. 2). Biopsies revealed an intraductal tubulopapillary neoplasm with high-grade dysplasia and focal transformation into adenocarcinoma (ADC).

Staging exams excluded lymph node or distant metastasis, and, after multidisciplinary discussion, the patient underwent Whipple's cephalic pancreatoduodenectomy with hepaticojejunostomy. The anatomopathological examination of the surgical specimen showed a moderately differentiated ADC in intestinal-type intraductal papillary neoplasm of the bile duct (IPNB) (pT1N0R0) (Fig. 2).

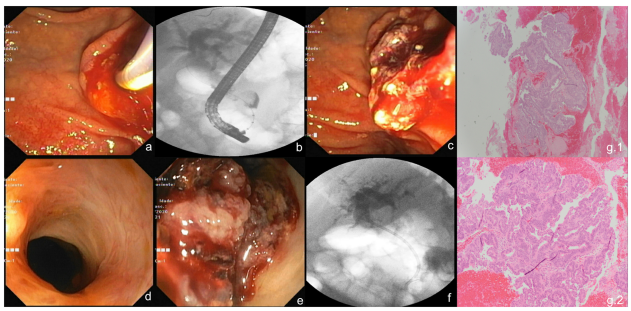

Figure 2 ERCP with sphincterotomy (a), showing a heterogeneous filling defect in the distal half of the CBD with upstream dilatation (b). Tumor fragments were obtained after CBP exploration with balloon (c). Cholangioscopy revealed normal distal CBD (d) with vegetative and stenotic lesion in the middle third (d). After biopsies, a plastic prosthesis was placed (f). Postoperative pathology showed a moderately differentiated adenocarcinoma in intestinal-type intraductal papillary neoplasm of the bile duct (CDx2+; MUC5AC focal; MUC1−; MUC2−) (g1-2).

There was no indication for adjuvant chemotherapy, but clinical, analytical, and imaging surveillance was recommended. After 1 year of follow-up, the patient remains asymptomatic.

IPNBs are rare neoplasms arising from the bile duct considered to be an important precursor of cholangiocarcinoma. They are commonly found in Far East Asia where hepatolithiasis and clonorchiasis are endemic. IPNBs can occur anywhere in the extrahepatic or intrahepatic bile ducts and microscopically are classified into four types: pancreaticobiliary, intestinal, gastric, and oncocytic. Most cases show at time of diagnosis high-grade intraepithelial neoplasia or invasive carcinoma.

IPNBs secrete mucin and induce biliary obstruction and dilation, but do not have a typical clinical or imaging presentation, making their early diagnosis a clinical challenge.

The major treatment is surgical resection with a better prognosis compared to cholangiocarcinoma.

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