

Letters to the Editor

Neuroendocrine carcinoma of the common bile duct

Key words: Neuroendocrine tumor. Extrahepatic bile ducts. Bile duct resection.

Dear Editor,

A 37-year-old female patient was admitted to the Emergency department complaining of itching, vomits, diarrhea, and abdominal discomfort for months. On examination the patient presented good general condition, with slight muco-cutaneous pallor, no jaundice, and mild discomfort in lower abdomen. Blood tests revealed an elevation of liver enzymes (GOT 118 UI/l, GPT 215 UI/l, GGT 187 UI/l, and alkaline phosphatase 214 UI/l) without any other alteration in biochemical parameters. Coagulation tests, blood count and tumor markers (alpha-fetoprotein 1.5 ng / ml) were within normal range. An abdominal ultrasound was performed showing a nodular image of 4x3 cm adjacent to the hepatic hilum that compressed the common bile duct at this level and caused dilatation of the intrahepatic bile ducts. The screening was completed with a computed tomography scan (CT) and a magnetic resonance imaging (MRI) (Fig. 1A), which showed an apparently benign mass compressing the common bile duct and hampering bile flow without causing a complete obstruction of the bile duct. The patient underwent surgical exploration, finding a large tumor solely dependent of the common bile duct (Fig. 1B), with no hepatic infiltration (proved with intraoperative ultrasound). A resection of the bile duct, from the upper pancreatic edge to the bile ducts confluence at the hepatic hilum, was performed

(Fig. 1C), with a lymphadenectomy and a Roux-en-Y hepaticojejunostomy. The patient had a favorable postoperative recovery and was discharged without complications. Histological study informed of a well differentiated neuroendocrine carcinoma of the common bile duct, TNE G2 (mitotic index: 3x10 HPF, Ki-67 3%), with free margins and no nodal infiltration. Typical immunohistochemical properties were positive: chromogranin A +, synaptophysin+, CD56 +, CK AE1/AE3 + (Fig. 1D).

Discussion

Biliary neuroendocrine tumors (NETs) are extremely rare, accounting for 0.1 %-2 % of all gastrointestinal neuroendocrine tumors (1). The most common anatomical locations are the common bile duct (58 %), perihilar (28 %), cyst duct (11 %) and common hepatic duct (3 %). They are more prevalent in women, during the fifth decade of life (2,4).

Although NETs may be hormonally active, most cases are not-functioning and the symptoms result from bile duct obstruction, like itching or biliary colic, which is difficult to be identified on imaging studies. The submucosal location of these tumors determines a large number of false negative results on brush biopsy, making it difficult to achieve a correct preoperative diagnosis. Differential diagnosis must be established with other hepatic and biliary tumors. The definitive diagnosis is established by histopathological and immunohistochemical analysis of the surgical specimen (3,5).

Radical resection of the tumor is the main prognostic factor in patients with biliary NETs. Complete resection of the extrahepatic bile ducts with a biliodigestive reconstruction (Roux-en-Y hepaticojejunostomy) with portal lymphadenectomy must be performed, that achieves a long-term high survival rate. Large tumors that infiltrate the proximal pancreas may require a cephalic duodenopancreatectomy.

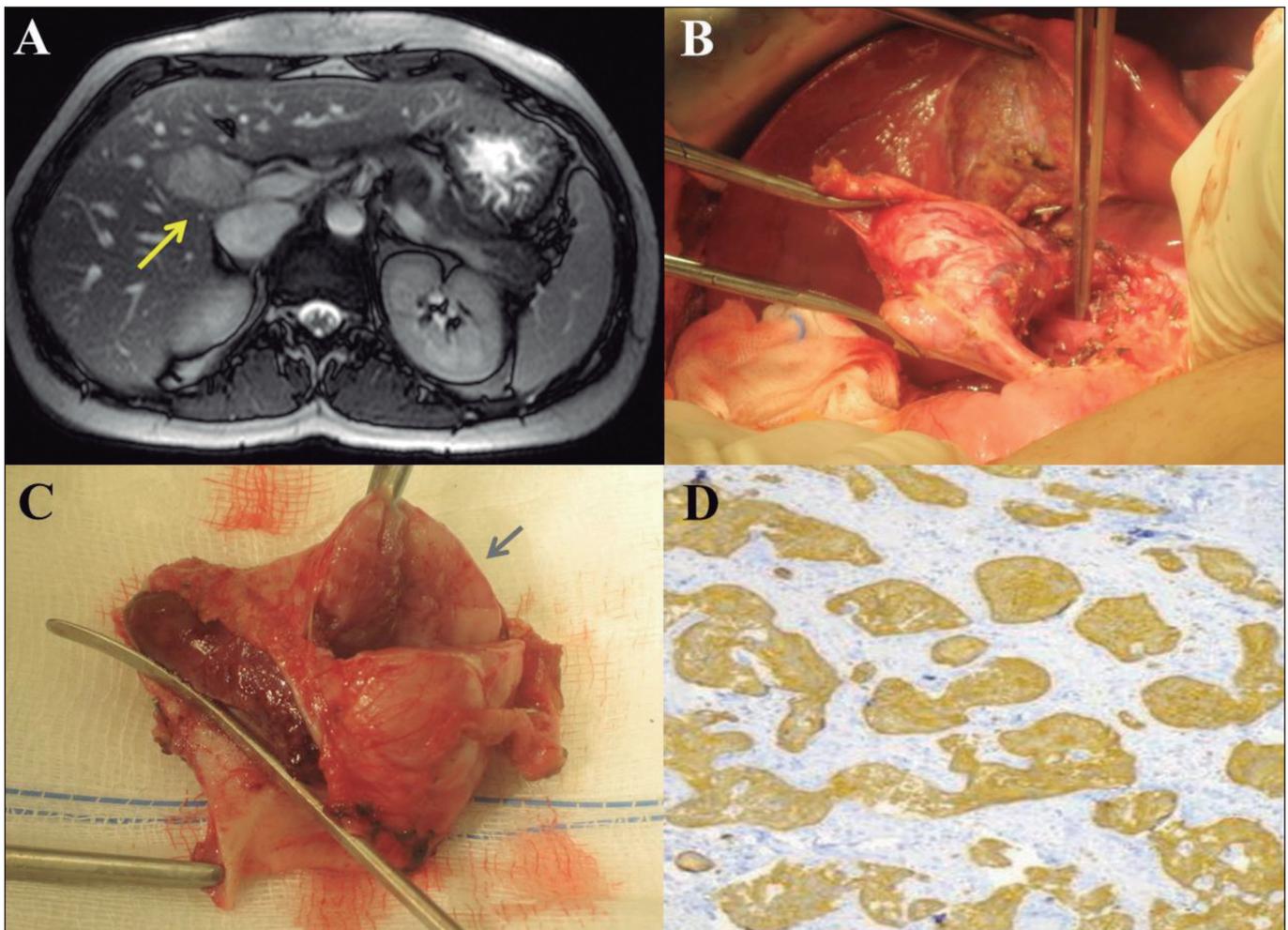


Fig. 1. A. Magnetic resonance imaging (MRI) showing the tumor (arrow). B. Intraoperative image of the common bile duct and the tumor. C. Surgical specimen. D. Immunohistochemical study: Cromogranine A +.

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