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Bile duct schwannoma, an uncommon cause of a bile duct tumor, diagnosis and therapeutic management

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Dear Editor,

Biliary schwannoma is an uncommon tumor in the digestive system and extremely rare in the bile duct. We report the case of a 48-year-old male who was admitted due to a COVID-19 infection. A chest computed tomography (CT) scan was performed due to the suspicion of a pulmonary embolism. An incidental tumor was identified in the hepatic hilum (3.2 cm), abutting the first portion of the duodenum, without common bile duct obstruction (Fig. 1A and B). The patient was asymptomatic and had normal tumor markers. An endoscopic ultrasound (EUS) demonstrated contact between the tumor, common bile duct and right hepatic artery and the biopsy was suggestive of a mesenchymal tumor. The tumor board recommended excision of the tumor due to the risk of malignancy and biliary obstruction. The tumor was resected via a laparoscopic approach, with a Roux-en-Y hepaticojejunostomy (Fig. 1C). The histological report

showed a biliary schwannoma (positivity for S100 and focal GFAP), which infiltrated the external wall of the bile duct, without affecting the cystic duct; the surgical margins were free of neoplasia (Fig. 1D).

Discussion

Schwannomas are benign neoplasms that originate from Schwann cells (1,2). The most frequent location is usually in the head and neck. Gastrointestinal involvement is unusual, and particularly rare in the bile duct (1). They are sometimes associated with hereditary syndromes (type 2 neurofibromatosis). These tumors affect middle age (40-45 years) females (1). The most common presenting symptoms are jaundice (78 %) and abdominal pain (< 50 %) (1-3).

The diagnosis is based on obstructive symptoms secondary to the space-occupying lesion (cholangitis, jaundice, abdominal pain, etc.) and radiological findings (common bile duct obstruction, malignant features). The best method of biopsy is via EUS, however, transpapillary biopsy is currently gaining great importance, since it obtains submucosal cells, aiding the differentiation from cholangiocarcinoma (4).

On histological examination, schwannomas show Schwann cells (fusiform cells), which are arranged alternating compact (Antoni A) and lax-hypocellular (Antoni B) (4) areas, positive for S-100 and vimentin (5). The differential diagnosis includes other causes of obstructive jaundice, either benign (lithiasis, autoimmune pancreatitis, choledochal cysts) or malignant (pancreatic adenocarcinoma, cholangiocarcinoma, ampullary cancer, duodenal gastrointestinal stromal tumors [GIST]) (5).

Regarding treatment, the standard of care is complete resection (5). In cases without a definitive preoperative diagnosis, resection might be more extensive, to ensure negative surgical margins. This often includes part of the bile duct, making it necessary to perform biliary reconstruction (2,5). Therefore, prior histological confirmation is essential to prevent unnecessary extensive surgical resection and its associated morbidity. Nevertheless, the prognosis after resection is excellent, with a low risk of recurrence (< 10 %) (1).

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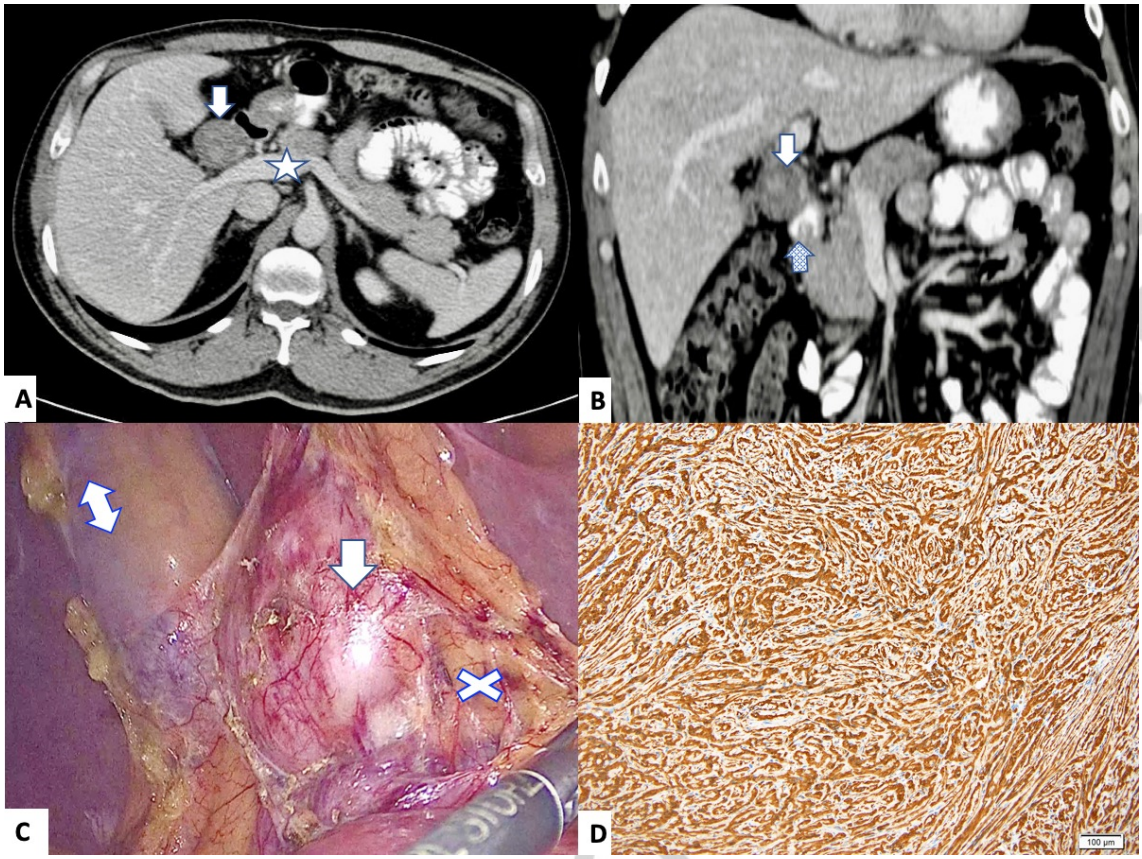

Fig. 1. Abdominal CT. A. Axial view showing a biliary tumor (arrow) with wide portal contact (star). B. Coronal view showing an intimate relationship between the tumor (arrow) and the first duodenal portion (dotted arrow). C. Laparoscopic view of the biliary tumor (arrow), in proximity to the gallbladder (double-headed arrow) and the main bile duct (cross). D. The neoplastic cells show immunohistochemical positivity for S100 (20x).