Primary non-functioning neuroendocrine tumor of the extrahepatic bile duct

Santiago Sánchez-Cabús, Gabriella Pittau, Mylène Sebagh and Daniel Cherqui

1HPB Surgery and Transplantation Department. Hospital Clínic de Barcelona. Barcelona, Spain. 2HPB Surgery and Transplantation Department. Centre Hépato-Biliaire Paul Brousse. Villejuif, France. 3Histopathology Department. Centre Hépato-Biliaire Paul Brousse. Villejuif, France

ABSTRACT

In contrast to the primary biliary carcinoma or cholangiocarcinoma, other tumors derived from the bile duct are difficult to diagnose preoperatively, mainly because of its low incidence and difficult diagnostic process. However, since cholangiocarcinomas account for about 80% of all primary biliary tumors, it is important to think about other options despite their low frequency when a patient presents with abnormal characteristics. We present a case of a primary neuroendocrine tumor of the bile duct, and a review of the literature on this rare disease.


CASE REPORT

A 38-year-old male patient consulted for a two-month history of abdominal discomfort associated with obstructive jaundice and asthenia in the last 20 days. CT scan and MRI cholangiogram showed a partially calcified 2 cm mass arising from the extrahepatic bile duct with intense contrast enhancement, causing retrograde dilatation of proximal biliary tree (Fig. 1 A and B), with no signs of local or distant extension. CEA, CA 19.9 and AFP levels were normal.

An en-bloc surgical resection of the tumor including the extrahepatic bile duct, cholecystectomy and regional lymphadenectomy was performed (Fig. 1C). The intraoperative examination of the surgical specimen suggested a neuroendocrine tumor (NET) of the common bile duct with clear margins. The postoperative course was uneventful, and the patient was discharged 6 days after surgery.

The final pathology report showed a 2 cm tumor with a fibrous stroma in its central part with small and homogeneous cells organized into pseudo-acinar formations. In the periphery, the tumor was composed of large cells with abundant microvacuolized cytoplasm showing rounded nucleus (Fig. 2 A and B). There was also perineural and lymphatic involvement. The final diagnosis was neuroendocrine carcinoma with high expression of CD56 and CK7 (Fig. 2 C and D, respectively), chromogranin and moderate expression synaptophysin, and with an index of Ki67 15% (G2), without lymph node involvement.

DISCUSSION

Primary biliary tree NETs represent 0.2-1% of all digestive NETs (1,2). While functioning NETs of gastro-
intestinal origin account for 6-10% (3), clinical symptoms resulting from hormonal hypersecretion of biliary NETs are rare, and only a few cases have been reported (4). The most common anatomical site of this neoplasm is the common bile duct, which represents about 60% of cases reported (5). Surgical resection is the primary treatment.

REFERENCES