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SCIENTIFIC LETTER

The value of preoperative histological confirmation in inconclusive biliary strictures: Biliary neurofibroma as a rare cause of icteric obstructive syndrome

La importancia de la confirmación histológica preoperatoria en las estenosis biliares sin diagnóstico concluyente. Neurofibroma biliar como causa rara de síndrome ictérico-obstructivo

Extrahepatic bile duct stricture is a common condition in clinical practice, with malignant tumours being the most frequent cause.[1] Benign tumours account for only 6% and the most common are intestinal adenoma and intraductal papillary neoplasm of the bile duct. Neurofibromas are rare in this location.[2] Neurofibroma is a benign neoplastic proliferation arising from the peripheral nerves and is made up of all the elements which form part of these nerves: Schwann cells, axons, fibroblasts and perineural cells. Characteristically it may be associated with neurofibromatosis type 1 (NF1), which causes gastrointestinal tract involvement in 10%–25% of patients.[2]

We present a case of common hepatic duct neurofibroma in a 76-year-old woman who had a cholecystectomy for an episode of acute calculous cholecystitis 20 years ago, with no subsequent episodes of biliary obstruction, who developed abdominal pain and fever. Laboratory tests showed hyperbilirubinaemia (direct fraction) and imaging tests (multidetector computed tomography [CT] and MR cholangiogram) showed dilation of the intrahepatic bile duct with stricture of the common hepatic duct, not affecting the common bile duct (Fig. 1A). An endoscopic ultrasound scan was then performed, with fine needle aspiration (FNA) on two lymph nodes identified in the biliary hilum, with no atypical cells observed. Subsequently, endoscopic retrograde cholangiopancreatography (ERCP) was performed, with balloon dilation and stent placement, leading to an improvement in the patient's condition (Fig. 1B). With all the accumulated clinical, radiological and cytological data and the clinical diagnosis of cholangiocarcinoma, and as oral cholangioscopy with biopsies was not available at our centre, we decided to perform surgery involving resection of the bile duct at the level of the stricture with Roux-en-Y anastomosis. The patient's postoperative recovery was uneventful and she was discharged six days after surgery.

Pathology examination of the bile duct showed the presence of a stricture-forming lesion within the thickness of the wall, 20 mm in diameter and plexiform in outline. Microscopically it consisted of spindle cells, some (Schwann cells) without significant nuclear atypia or mitotic activity and positive for S100 and others (fibroblasts) arranged in a more scattered manner, also without frank atypia and positive for CD34 (Fig. 1C, D). The definitive diagnosis was plexiform neurofibroma. Genetic testing subsequently ruled out NF1 in the patient.

Obstructive lesions of the bile duct give rise to similar clinical signs and symptoms, but can be due to different aetiologies and each has a different therapeutic approach and prognosis.[1,3–5] Clinical diagnosis of the obstructive condition and identification of the culprit lesion is usually achieved using non-invasive imaging tests.[3] Currently, the combination of magnetic resonance imaging (MRI) and magnetic resonance (MR) cholangiography is the technique of choice.[1,3,5] However, the aetiological diagnosis of the obstructive lesion requires pathology examination and the methods required for this pose major difficulties. The sensitivity and specificity of endoscopic and/or ultrasound studies with cytology and/or biopsy sampling are simply too low.[3–5] This situation has been improved by single-operator direct cholangioscopy with multiple biopsies taken under direct vision, but there are still unresolved cases, and it is a technique not available at all hospitals.[3–5] The limitations of all these preoperative diagnostic methods explain the continued reliance on intraoperative histological study to confirm the aetiology of the lesion causing the biliary obstruction and suitably adjusting the treatment to the histological diagnosis.

Neurofibromas in the common hepatic duct are rare (fewer than 40 cases reported)[1] and difficult to diagnose, as the clinical and radiological findings are similar to those of other obstructive bile duct lesions, such as cholangiocarcinoma. We have also seen how the various preoperative diagnostic methods available do not adequately solve the problem, particularly in the case of benign lesions, as they have sensitivity rates of only up to 80%, and in centres where oral cholangioscopy is available. We believe the case we

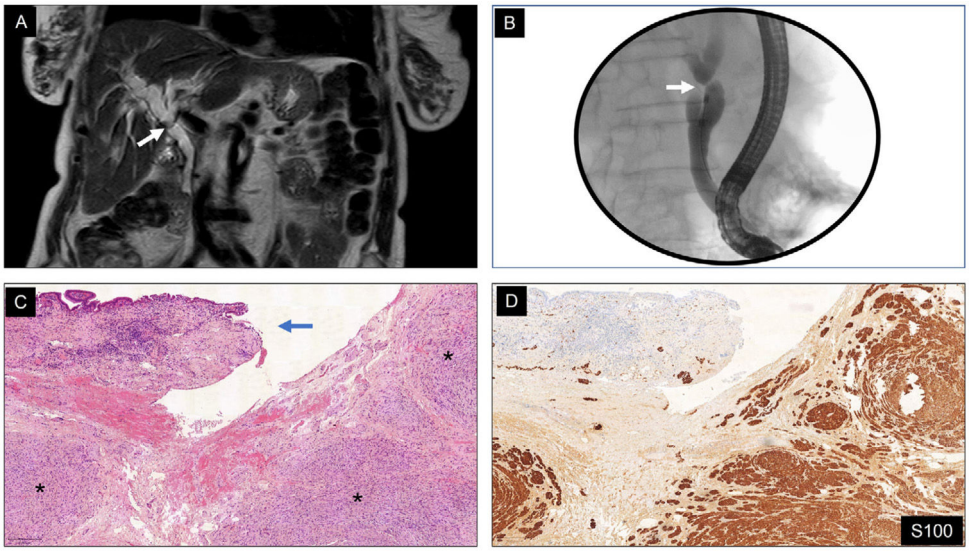

Figure 1 Marked dilation of the intrahepatic bile ducts with short stricture at the level of the common hepatic duct (arrow). A) MR-cholangiogram. B) ERCP. Bile duct mucosa with non-specific inflammatory infiltrate (arrow) and spindle cell proliferation in a plexiform configuration within the thickness of the wall (asterisks). C) H&E, 40×. D) Immunohistochemistry against S100 protein, 40×.

have presented here is a perfect example of the importance of intraoperative histological study in obstructive biliary lesions without a preoperative confirmatory diagnosis, as we believe that this is the only way to avoid excessively aggressive surgical treatment in cases of benign aetiology.

Conflicts of interest

The authors have no conflicts of interest.

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