LETTER / Gastrointestinal imaging

Granular cell tumour of the common bile duct: A condition to be aware of

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KEYWORDS
Abrikossoff’s tumour; Granular cell tumour; Common bile duct; Imaging

A granular cell tumour (GCT), also known as Abrikossoff’s tumour, is an infrequent benign neurogenic tumour, occurring ubiquitously but with a predilection for cutaneous and buccopharyngeal sites [1,2]. It appears at between 40 and 50 years of age and seems to be more frequent in women [1]. It is diagnosed exclusively from histological/immunohistochemical analyses which show an origin in the Schwann cells with positive S100 protein labelling [3]. A biliary location is exceptional and mimics biliary cholangiocarcinoma in all aspects. Only 79 cases have been described in the literature [1–4]. The authors report an additional case of a granular cell tumour of the proximal common bile duct.

Clinical case

A 40-year-old man with no medical history was hospitalised for cholestatic jaundice which had developed over 45 days, with dark urine and normally coloured stools. His general state of health had remained unaffected. Laboratory liver tests were disturbed with alkaline phosphatase at 254 U/l (normal value 30–130 U/l) and total bilirubin at 50 mg/l (normal value 3–10 mg/l), with predominance of conjugated bilirubin. A CT scan showed a tissue process in the proximal common bile duct, with a long axis measuring 15 mm, that was enhanced after contrast injection and occupied the lumen of the common bile duct (Fig. 1). MRCP confirmed the biliary wall tumour mass, by revealing a T2-weighted hypointense zone enhanced after injection of gadolinium chelates, which was responsible for proximal bile duct stenosis with a preceding dilatation (Fig. 2). Cholangiocarcinoma was diagnosed and the patient underwent resection of the proximal common bile duct with a Roux-en-Y biliodigestive anastomosis and cholecystectomy. Histological examination revealed the presence of tumour cells with eosinophilic granular cytoplasm and regular nuclei, with positive immunostaining for S100 protein, on immunohistochemical examination, indicating a granular cell tumour of the common bile duct. There were no postoperative complications and the clinical outcome after 18 months was good.

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http://dx.doi.org/10.1016/j.diii.2012.10.019
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Discussion

Extrahepatic bile duct granular cell tumours are exceptional (less than 1%). They generally occur singly, usually in the common bile duct (54% of cases) [4]. Clinically, they are revealed predominantly as cholestatic jaundice and/or abdominal pain. This jaundice is related to obstruction of the bile duct and may be complicated by secondary cirrhosis [1]. In imaging, the morphological appearance is of endoluminal obstruction or extrinsic compression. The mass is of soft tissue, isodense in a CT scan, while in T1- and T2-weighted images it is isointense relative to the liver parenchyma, and enhances after injection of a contrast agent to produce an image of regular segmental stenosis in MRCP sequences. Endoscopic ultrasonography shows a well-defined, hypoechoic tumour, with regular contours, developing in the submucosa of the common bile duct. The main differential diagnoses are most frequently with cholangiocarcinoma and sclerosing cholangitis and less frequently with other benign tumours (adenoma). Only the results of histology of a cytological sample or endobiliary biopsy can discriminate between them. Imaging also provides information on both the extent and resectability of the tumour and thus provides guidance for the surgical procedure. The appearance in a young patient of cholestatic jaundice that fluctuates initially and slowly but progressively worsens should suggest a benign tumour as the origin of the obstruction [2].

Treatment is surgical and requires complete excision with clear margins. The risk of recurrence is increased where excision is incomplete, and long-term follow-up is required to watch for recurrence. This surgery obviously depends on the location and extent of the tumour.

Histological diagnosis is always made retrospectively and the definitive results are only obtained from the surgical specimen. The Schwann cell origin is now recognised and confirmed by the expression of S100 protein on immunohistochemical examination. Signs of malignancy are systematically sought.

Figure 1. Abdominal axial CT before (a) then after injection of a contrast agent (b) and frontal reconstruction (c). Proximal common bile duct tissue mass measuring 15 mm, occupying the lumen of the duct with a preceding dilatation.

Granular cell tumours of the common bile duct are rare but they do exist. They are responsible for obstructive jaundice and can simulate malignant biliopancreatic disease in all clinical, biological and radiological aspects. Diagnosis is essentially histological. It is important to be aware of this condition: it can sometimes help avoid major, and above all, unnecessary surgery.

Disclosure of interest
The authors declare that they have no conflicts of interest concerning this article.

References