Case of an extra-large biliary intraductal papillary neoplasia

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DESCRIPTION

Intraductal papillary neoplasia (IPNB) is a biliary epithelial premalignant tumour with a predominant papillary intraductal growth.¹ IPNBs arise from peribiliary glands, which are rich in biliary tree progenitor cells with potential to differentiate into hepatocytes, cholangiocytes and pancreatic islets.² In 2010, it was added to the WHO classification, encompassing from low-grade intraductal neoplasia to IPNB with invasive carcinoma (present in 40%–80% IPNBs) that represents intraductal growth-type cholangiocarcinoma.^{1,3}

Nearly one-third of IPNB show histological and immunophenotypic similarities with pancreatic intraductal papillary mucinous neoplasm (IPMN), with production of mucin, while IPNBs without mucin hypersecretion resemble nonpapillary cholangiocarcinomas.^{2,4}

IPNBs are endemic in Far Eastern countries, usually between fourth and sixth decades of life, associated with hepatolithiasis and clonorchiasis.³

We present a case of a Caucasian 65-year-old woman, with prior cholecystectomy, presented at the emergency department with intermittent jaundice, coluria, pruritus and abdominal pain. Blood investigations revealed raised hepatic transaminases, gamma-glutamyl transferase

(GGT) and alkaline phosphatase (ALP) and normal tumour markers.

The patient underwent ultrasound examination which revealed a non-specific extrahepatic intraductal lesion, combined with dilatation of the proximal biliary tree. MR with magnetic resonance cholangiopancreatography (MRCP) depicted a large intraductal solid lesion, occupying the distal two-thirds of the common bile duct (CBD) (figure 1A–D), with upstream dilatation of the biliary tract (figure 1E), compatible with IPNB.

After surgical resection, histopathology examination revealed a high-grade IPNB, with a pancreaticobiliary phenotype with an invasive adenocarcinoma component.

IPNBs are usually sessile or polypoid lesion and can have multifocal superficial spreading masses along the mucosal surface, in both intrahepatic and extrahepatic bile ducts, known as papillomatosis. These multiple lesions may detach spontaneously, mimicking biliary lithiasis.⁵

On imaging, IPNBs can have five patterns: diffuse duct ectasia with a grossly visible papillary mass (type 1); diffuse duct dilatation without a visible mass (type 2); localised ductal ectasia with an intraluminal polypoid mass (type 3); intraductal cast-like lesions (type 4) and mild ductal dilation proximal to a focal stricture-like lesion (type 5).^{3,6}

Only complete surgical resection allows a curative treatment. IPNBs have a better prognosis when compared with conventional cholangiocarcinoma,

Figure 1 Coronal T2-weighted (A), axial contrast-enhanced three-dimensional gradient echo image with fat suppression after contrast injection (B) and diffusion-weighted imaging (C) with apparent diffusion coefficient map (D) MR images show the intraductal papillary neoplasia (white arrows in A–D) as a large intraductal solid lesion, occupying the distal two-thirds of the CBD. MRCP (E) shows upstream dilatation of the biliary tract.

Patient's perspective

I am pleased that my case helps the scientific community and so can help other patients.

Learning points

- ▶ Intraductal papillary neoplasia (IPNB) is an uncommon form of epithelial biliary papillomatosis with a variable degree of malignancy that may be clinically present by jaundice and coluria.
- ▶ Imaging of IPNBs can range from a localised or diffuse biliary dilatation, with or without visible intraluminal mass to a focal stricture evoking cholangiocarcinoma.
- ▶ The only curative treatment is complete surgical resection.

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with 5-year survival rates between 60% and 82% in patients undergoing R0 resection.⁷

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