Ectopic pancreas as a rare cause of obstructive jaundice

Dear Editor,

The authors describe a case of a 68-year-old man presenting in the emergency room with a one-week history of jaundice, pruritus and weight loss. Patient had history of hypertension (on therapy with telmisartan), alcohol (75 g/day), and tobacco consumption. On physical examination only jaundice was noticed. Blood tests showed elevation of total and conjugated bilirubin (21.1 and 13.3 mg/dL), ALT (493 IU/L), AST (522 IU/L), alkaline phosphatase (542 IU/L), gamma-glutamyl transpeptidase (2102 IU/L) and carbohydrate antigen 19-9 (229 IU/mL). Contrast-enhanced helicoidal computed tomography and magnetic resonance cholangiography demonstrated dilation of intrahepatic ducts and distal common bile duct (CBD) (16 mm) ending in an abrupt stenosis, without an apparent cause. Liver, pancreas and Wirsung were normal.

Endoscopic retrograde cholangiopancreatography revealed a slightly enlarged papilla and a stenosis in the CBP, where a 10 Fr plastic stent was placed; brush cytology of the CBD and biopsies of the ampulla were normal. In endoscopic ultrasonography (EUS), a hypoechoic submucosal nodule of 8 mm was seen in the area of the ampulla (Fig. 1A).

There was clinical and laboratory improvement and patient was discharged. He returned one month after with an episode of cholangitis.

The case was then discussed with surgeons and as a neoplastic cause could not be ruled out based on the imagiologic and endoscopic findings (repeated twice), the decision was to perform a pancreaticoduodenectomy.

Macroscopic examination revealed dilated bile ducts and normal pancreas. In section of the ampulla, there was an expansion of the submucosa composed of ectopic pancreatic tissue with features of chronic pancreatitis (Fig. 1B).

The patient remains asymptomatic after a year of follow-up.

Discussion

Ectopic pancreas is a rare condition defined as pancreatic tissue abnormally situated, without contact with normal pancreas and with its own ductal system and blood supply (1). The most common locations are duodenum, stomach and jejunum (2). Periampullary location has been described in less than 25 reports (3). Clinical presentation is usually asymptomatic or, in case of inflammation, obstructive jaundice can occur.

Accurate preoperative diagnosis using endoscopic or radiologic imaging is difficult because the findings are not pathognomonic. The presence of a central umbilication of the ampulla in endoscopy, although characteristic, is infrequent (4). EUS may be important at the diagnosis showing a hypoechoic and heterogeneous nodule with indistinct margins, generally in the submucosa (4,5).

Local excision may be the treatment of choice. However, as the correct diagnosis is sometimes difficult and this situation can mimic a neoplasia, pancreaticoduodenectomy is performed in the majority of cases (3,5,6).

We describe this case of periampullary ectopic pancreas, where the clinical doubt about etiology (malignant or not) ended in a radical surgery. Although the findings in EUS can be recognized as a feature of this disease, a fine needle aspiration or a biopsy of the nodule would not be conclusive because, in the presence of pancreatic tissue in this location, we would probably interpret it as an error sample.
We admit that the possibility of ectopic pancreas, although rare, should be included in the differential diagnosis of obstructive jaundice and that EUS can show a typical image and help in the diagnosis.

Rita Carvalho¹, Paulo Freire¹, Francisco Portela¹, Dário Gomes¹, Paulo Andrade¹, Ana Bento², Hamilton Batista², M. Augusta Cipriano³ and Carlos Sofia¹

¹Gastroenterology Department, ²General Surgery Department and ³Pathology Department. Coimbra University Hospital, Portugal

References


Fig. 1. A. Ultrasonography showing a submucosal nodule of 8 mm adjacent to the papilla. B. HE 20X (40X in the right upper quadrant): Section of the ampulla revealing ectopic pancreatic tissue in the submucosa of the duodenum wall, with atrophic acini and dilated ducts.