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IMAGE OF THE MONTH

IgG4-related disease mimicking pancreatic tumour☆

Enfermedad IgG4-mediada simulando neoformación de páncreas

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58-year old woman who consulted for pruritus and liver panel alterations: AST 153 U/l, ALT 266 U/l, GGT 979 U/l, alkaline phosphatase 807 U/l with Ca 19.9 15 U/mL.

The magnetic resonance suggested malignant neoformation (Fig. 1). The endoscopic ultrasound-guided FNA revealed cellular atypia. A cephalic pancreatectomy was performed without metastasis or vascular invasion being observed. The rest of the pancreatic parenchyma was atrophic and fibrous. The histopathological study was consistent with IgG4-related type 1 autoimmune pancreatitis (Fig. 2).

Type 1 autoimmune pancreatitis is the pancreatic manifestation of IgG4-related disease.[1] The histopathology[2] is what was found in our case. This process involves other organs[3]; the most common is association with sclerosing cholangitis (70% cases).[1]

The patient was followed up and 4 years later she presented with cholestasis GGT 290 U/l and alkaline phosphatase 130 U/l without cytolysis. An increase in IgG of 1850 mg/dl (normal: 600–1600 mg/dl) and ANA + 1/160 were detected in the lab tests. The endoscopic ultrasound did not show any biliary alterations. A liver biopsy was performed and was consistent with cholangitis attributed to IgG4, although in this case the staining for IgG4 plasma cells was not positive (Fig. 3).

Corticosteroids associated with azathioprine were prescribed, obtaining a good response.

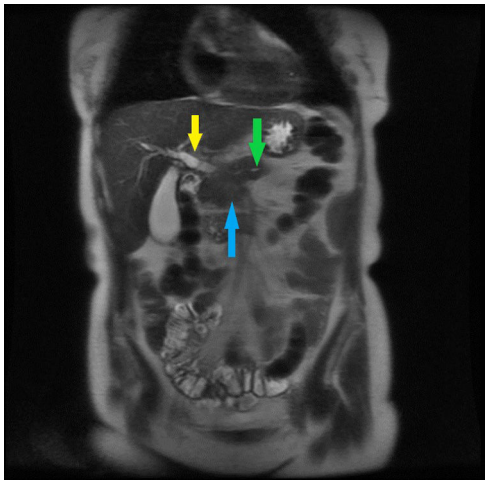

Figure 1 MRI: Irregular, hyperintense mass of 2.5 cm, poorly defined in head of the pancreas (blue arrow) with intra- and extrahepatic bile duct (yellow arrow) and duct of Wirsung (green arrow) dilatation.

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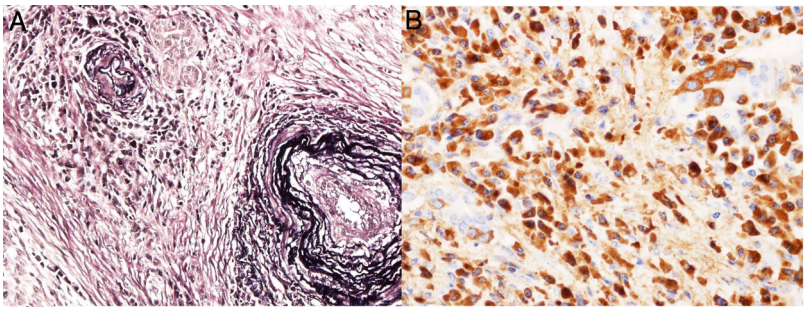

Figure 2 A) Microscopic image of the pancreatic surgical specimen showing diffuse storiform interstitial fibrosis, obliterative phlebitis and intense inflammatory infiltrate of T lymphocytes (A) together with high positivity for IgG4 plasma cells (B).

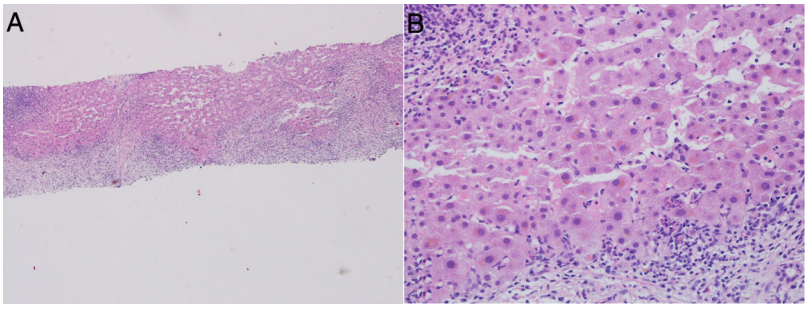

Figure 3 Liver biopsy showing a distorted parenchyma (A) with significant portal interphase infiltration, necrosis and fibrosis in bridges; the intrahepatic bile ducts showed intense obliterative fibrosis (B).

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