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Case report

Autoimmune pancreatitis presenting as a pancreatic head mass [☆]

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ABSTRACT

The diagnosis of autoimmune pancreatitis (AIP) is often difficult as the main differential diagnosis is pancreatic head adenocarcinoma. Some clinical, radiological, serological and histological criteria help in guiding the diagnosis. The serum gamma-globulin IgG4 dosage is the most sensitive and specific marker. The shape of the ductal stenosis in AIP is characteristic. The treatment is based on corticosteroids. We report the case of a 53-year-old woman, presenting with pancreatic head mass mimicking an adenocarcinoma.

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Introduction

Autoimmune pancreatitis (AIP) is a rare entity. The differential diagnosis remains with pancreatic adenocarcinoma. Immunological markers or even biopsy of the pancreas help in making the diagnosis. Imaging, especially Magnetic resonance cholangiopancreatography (MRCP) plays an important role in the diagnosis of AIP, as it gives detailed information about pancreatic parenchyma and ducts, highlighting some characteristic signs of AIP. MRCP typically shows multiple skipped narrowings of the main pancreatic ducts (MPD) without dilation of the upstream MPD. A fibrous halo can also be seen around the pseudo mass [1,2].

Case report

A 53-year-old woman, with no history of alcohol abuse or other predisposing factors for chronic pancreatitis, presenting with transfixing epigastric pain evolving for 10 days associated with the progressive onset of mucous and cutaneous jaundice with pale stools and dark urine. She had moderate deterioration in general condition and weight loss (3kg).

Clinical examination found apyretic patient with mucocutaneous icterus and epigastric tenderness. The rest of the physical examination was normal.

The biological analysis showed bilirubin 97 $\mu\text{mol} / \text{L}$ alanine aminotransferase (ALT) 300 IU / L, aspartate aminotrans-

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Fig. 1 – Contrast enhanced abdominal CT scan at the pancreatic phase showing a hypodense pancreatic head mass, little enhanced respecting the vascular structures with infiltration of peri-pancreatic fat.

ferase (AST) 155 IU / L, γ -glutamyl transpeptidase (GGT) 500 IU / L, Tumor marker values: CA19-9 = 90 U / ml.

A contrast enhanced abdominal CT scan at the pancreatic phase was performed showing a hypodense pancreatic head mass, little enhanced respecting the vascular structures with infiltration of peri-pancreatic fat, measuring 23 × 18mm (Fig. 1), suggesting a resectable adenocarcinoma of the head of the pancreas.

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Further imaging with MRCP showed a pancreatic head mass hypointense on T1 and T2 weighted sequences, with diffusion coefficient restriction, surrounded by a capsule of hypointensity on T1 and T2 weighted sequences with mild enhancement after injection of gadolinium, measuring 23 × 18mm. There was irregular narrowing of the main pancreatic duct which was not clearly visualized in the pancreatic head (Fig. 2).

The features identified after the MRCP were in keeping with the diagnosis of focal autoimmune pancreatitis. An IgG4 assay was performed showing a high rate of 1.65 g/l. The patient underwent corticosteroid therapy with a good clinical evolution and regression of the morphological abnormalities observed during a CT scan control (Fig. 3).

Discussion

Autoimmune pancreatitis (AIP) is a rare clinical entity. The diagnosis is based on clinical, radiological, serological and histological criterias. In the majority of cases, risk factors for chronic pancreatitis or alcohol dependence are missing [1].

It can manifest clinically as acute epigastric pain associated with the progressive onset of weight loss with cutaneous mucosal jaundice [1,2].

There are 2 types of AIP: Type 1 AIPs corresponding to IgG4 sclerosing disease, which is more frequent in persons over 50-60 years old, with male predominance. Its histological peculiarities are a dense per-ductal lymphoplasmacytic infiltrate, pancreatic fibrosis and obliterating venulitis. Serum immunoglobulins are greatly elevated and cause multi-organ damage (mainly biliary damage). Type 2 AIP, which is more common in 40-year-olds, is often associated with other autoimmune diseases such as sclerosing cholangitis, chronic inflammatory bowel disease, Gougerot-Sjögren Syndrome, thyroiditis, diabetes and retroperitoneal fibrosis. In this type, serum immunoglobulin levels are normal [1,3].

There are many and varied biological markers of AIP: rheumatoid factor antinuclear antibody (ANA), anticarbonic anhydrase antibody-II, antilactoferrin antibody, but the serum gamma-globulin IgG4 dosage remains the most sensitive and specific marker (> 1.35g / l or > 2 times normal). However, these markers have a poor negative predictive value and their absence should not rule out the diagnosis if there is a strong clinical and morphological presumption [4,5].

In imaging, AIP causes parenchymal and ductal changes. The pancreas becomes delobulated, swollen, with a hypodense appearance on CT and hypo intense on MRI in T1-weighted sequence with late enhancement after injection of contrast product.

A fibrous halo can also be seen around the pseudo mass. The presence of calcifications and pseudocysts remain rare. Ductal changes are best studied by cholangiopancreato-MRI (CP MRI), which typically shows multiple skipped narrowings (skip lesions) of the main pancreatic ducts (MPD) without dilation of the upstream MPD (no MPD dilatation) [3,5].

The shape of the ductal stenosis helps guide the diagnosis. The main pancreatic duct becomes threadlike, irregular with stepped strictures, centered without dilation upstream, as well demonstrated on the CP MRI performed in our patient.

Endoscopic ultrasound may show diffuse enlargement of the pancreas, hypoechoic, heterogeneous or a hypoechoic mass with irregular contours. The Endoscopic retrograde cholangiopancreatography (ERCP) shows an irregular and narrowed aspect of the wirsung duct with stepped strictures [3,5,6].

Histological and anatomic-pathological analysis are rarely available, after resection of pseudo-tumor forms suggesting carcinoma, especially when the involvement predominates in the head of the pancreas. The main differential diagnosis is adenocarcinoma of the head of the pancreas and it is often difficult to separate it. According to the literature, 5 to 10% of pancreatic resections for adenocarcinoma of the head of the pancreas show inflammatory masses [1,2].

The clinical and biological criteria in favor of tumor involvement of the head of the pancreas are high age, persistent jaundice, weight loss, and an increase in the level of CA19-9 greater than 300 U/ml against an increase in serum IgG4 level in AIP.

The shape of the ductal stenosis on imaging also helps guide the diagnosis. In the case of adenocarcinoma of the head of the pancreas, there is ductal dilation with sudden stenosis,

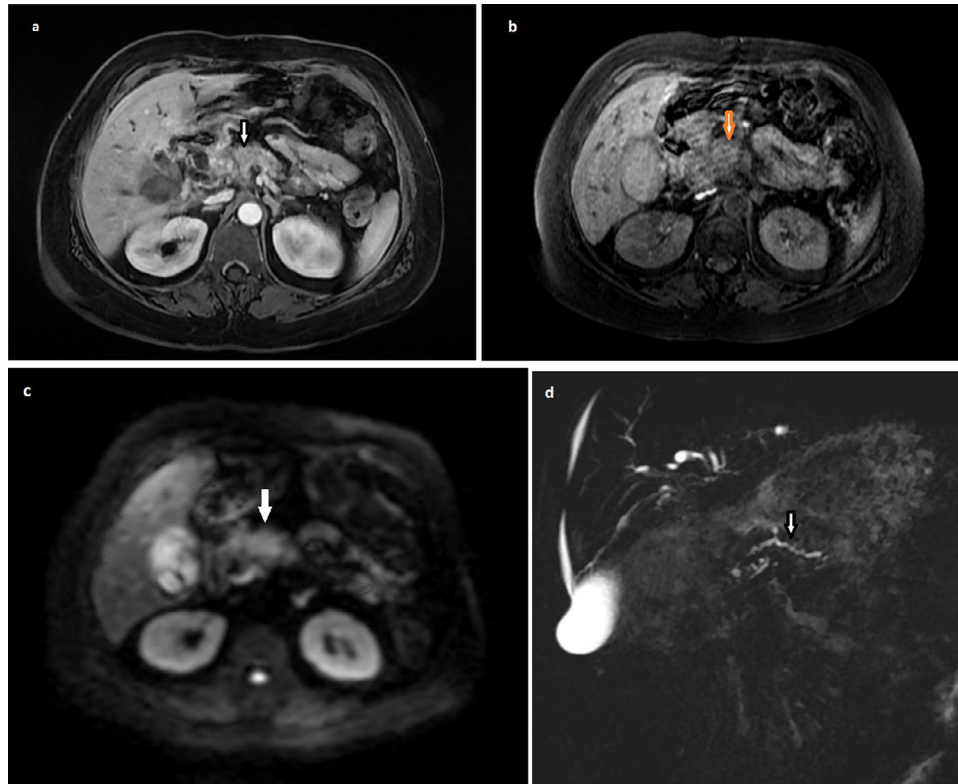


Fig. 2 – (a): T1 weighted MR On a pancreatic-phase of dynamic contrast-enhanced image showing the main pancreatic duct which is partially unclear in the head of the pancreas (arrow). (b): T1-weighted MR images showing a nodular lesion in the pancreatic head surrounded by hypointense capsule-like rim (arrow). (c): diffusion weighted image ($b = 800$) showing a nodular hyperintense lesion in the pancreatic head (arrow). (d): MRCP showing irregular narrowing of the main pancreatic duct (arrow).



Fig. 3 – Contrast enhanced abdominal CT scan at the pancreatic phase showing a normal pancreas.

while in the case of AIP, the stenosis is irregular, thread-like without clear dilation of the pancreatic duct [2,3,6].

The treatment is with corticosteroids, resulting in clinical improvement in a few days and a reduction in morphological abnormalities [5,6].

Conclusion

The diagnosis of AIP is based on clinical, biological and radiological arguments. It must be evoked when there is a pancreatic mass without suggestive context of pancreatic adenocarcinoma in order to avoid unnecessary pancreatic resection. If the doubt persists between AIP and an adenocarcinoma, it is recommended to do a corticosteroid test and observe the evolution.

Patient consent

Written informed consent for publication was obtained from the patient.

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