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Autoimmune Pancreatitis, Pseudo-Tumor Form Mimicking a Pancreatic Adenocarcinoma

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ABSTRACT

Autoimmune pancreatitis is a rare form of chronic pancreatitis. There are two diffuse and focal forms that mimic the appearance of a pseudo-mass or pancreatic adenocarcinoma, which the differential diagnosis may be difficult, The classic CT scan appearance of AIP is that of diffuse hypertrophy of the sausage shaped pancreas and a pathognomonic hypodense peripheral ring with a delayed contrast enhancement, Focal pancreatic involvement is often seen in the head of the pancreas and appears in the form of a mass mimicking pancreatic adenocarcinoma.

Introduction

Autoimmune pancreatitis is a rare form of chronic pancreatitis. There are two diffuse and focal forms that mimic the appearance of a pseudo-mass or pancreatic adenocarcinoma, which the differential diagnosis may be difficult. This article aims to recall the imaging aspect of this pathology and how to orient the diagnosis.

Case report

This is a 62-year-old patient with no particular history or toxic habits. He was hospitalized for the exploration of cholestatic jaundice on progressive installation for six months, associated with epigastralgia of low intensity, vomiting, and alteration of general condition. The clinical examination shows a BMI calculated at 20, a body temperature measured at 37 °C. Mucosal skin examination reveals a yellowish coloration consistent with jaundice. Abdominal palpation finds a firm, immobile epigastric mass. The laboratory workup revealsthe presence of an inflammatory syndrome (increased C-Reactive Protein and hyperleukocytosis),cholestasis syndrome (increased alkaline phosphatases, Gamma-glutamyl-transferases and total bilirubin), and a moderate increase of lipasemia.

Due to these clinical and biological results, a tumor obstruction of the bile ducts, in particular a process of the head of the pancreas is suspected. The abdominal ultrasound did not show any dilation of the intra or extra-hepatic bile ducts. Computed tomography (CT) with an injection of contrast product, was performed which upstream: diffuse hypertrophy of the pancreas with loss of its lobulations producing the appearance of the large sausage pancreas, with individualization of a characteristic hypodense sub-capsular peripheral ring (red arrows), it is associated with a bodily cystic lesion related to a retentional pseudo-cyst (white arrow, Fig. 1). The corporeo-caudal portion being more hypodense with a defect of enhancement contrasting with the cephalic portion, the whole producing a pseudo-tumoral aspect (orange arrows, Fig. 2), absence of corporeo-caudal atrophy or dilation of the pancreatic duct, with the presence of bilateral hilo-mediastinal adenopathies (blue arrows, Fig. 3).

On basis of this radiological semiology, the diagnosis of autoimmune pancreatitis has been proposed. The second-line laboratory test found normal neoplastic markers, positive antinuclear antibodies with a serum IgG 4 level twice the normal.

Faced with this array of biological and radiological arguments, the diagnosis retained is an autoimmune pancreatitis type II, treated with corticosteroid therapy with good clinical outcome.

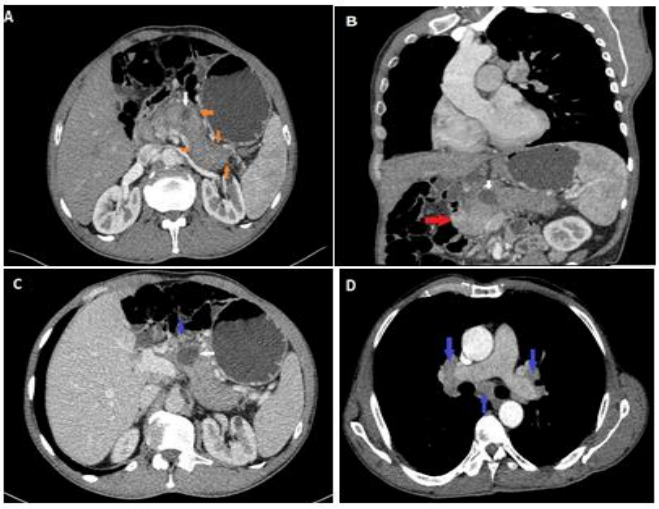

Figure. Thoracic and abdominal CT-Scan with an injection of contrast in Axial (A, C) and coronal (B) abdominal section, and axial thoracic section.

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Figure shows an hypertrophy of the pancreas making the sausage shaped pancreas, with individualization of a characteristic hypodense sub-capsular peripheral ring (red arrows), associated with a bodily cystic lesion related to a retentional pseudo-cyst (white arrow).The corporeo-caudal portion being more hypodense with a defect of enhancement contrasting with the cephalic portion, the whole producing a pseudo-tumoral aspect (orange arrows), associated with a peripancreatic and bilateral hilo-mediastinal adenopathies (blue arrows).

Discussion

Autoimmune pancreatitis (AIP)is a rare form of chronic pancreatitis characterized by an autoimmune-inflammatory process causing fibrosis and destruction of exocrine and possibly endocrine tissue in the pancreas. The term autoimmune pancreatitis wasintroduced by Yoshida et al in 1995 to describe the form of chronic pancreatitis associated with autoimmune manifestations. In 2001,Hamano et al.,first described sclerosing pancreatitis associated with elevation of serum IgG4levels [1,2].

There are 2 types of AIP: Type 1 (lymphocytic sclerosing pancreatitis), the most common in Asia, is characterized by a high serum IgG4 level. It is often associated with multi-visceral involvement (sclerosing cholangitis, tubulo-interstitial nephritis, retroperitoneal fibrosis, interstitial lung disease with mediatisnal lymphadenopathy). Type 2 (idiopathic duct-centric pancreatitis): common in Europe and the United States characterized by destructive polynuclear neutrophil infiltration of inter-lobular pancreatic ducts without elevation of serum IgG4 levels, chronic inflammatory bowel disease is often associated. [3, 4]

Because it is a rare entity its overall prevalence and incidence in the general population is not known, while three series have reported a prevalence of AIP between 5 and 6% of all patients with chronic pancreatitis, it affects both sexes with a male predominance, twice more frequent in men than in women. The dominant age group is that over 50 years. [5, 6]

The Clinical presentations are variable and nonspecific include prominent jaundice and abdominal pain abdominal, sometimes weight loss linked to exocrine pancreatic insufficiency. The classic CT scan appearance of AIP is that of diffuse hypertrophy of the sausage shaped pancreas and a pathognomonic hypodense peripheral ring with a delayed contrast enhancement, sometimes retentional pseudo-cysts, and slighten largement of the regional lymphnodes are associated. Ductal signs in the form of multiple stenoses of the main pancreatic duct without upstream dilations will be better appreciated by Bili-MRI. Ultrasound-endoscopy allows a more precisestudy of the parenchyma and pancreatic ducts, and performing a puncture-aspiration and ruling out with certainty a neoplastic origin. [7, 8]

Focal pancreatic involvement is often seen in the head of the pancreas and appears in the form of a mass mimicking pancreatic adenocarcinoma, hence the interest in establishing precise diagnostic criteria which are the subject of a consensus. The Japanese Society of Pancreas proposed a international classification, in 2002, revised in 2006, and then in 2009, based on the combination of radiological criteria, namely parenchymal and ductal signs. Serological criteria based on the level of IgG 4 and the presence of auto-antibodies such as antinuclear antibodies and rheumatoid factor. Histological criteria in particular lymphoplasmatic cell infiltration with abundant positive IgG4 cells and inter-fibrosis -lobular marked for AIP type II, The presence of extra-pancreatic lesions, and the response to corticosteroids. [4]

The treatment is based on systemic steroidal corticosteroid therapy; the initial dose of prednisone is 30-40 mg per day. The response is often spectacular with the improvement of clinical and biological signs and reduction of radiological abnormalities. It is recommended to carry out a radiological control at the end of the fourth week after initiation of the treatment. The absence of response to the treatment should suggest a pancreatic neoplastic origin or another form of chronic pancreatitis, in the event of relapse during withdrawal or maintenance treatment is recommended when stopping steroids. [9, 10]

Conclusion

An autoimmune pancreatitis is a rare form of chronic pancreatitis, its diagnosis is important because it is often difficult to differentiate it from pancreatic carcinoma leading to heavy and unnecessary surgeries, hence the interest of an international consensus including that of the Japanese Society of Pancreas. Its treatment is medically based on corticosteroid therapy with major efficiency.

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