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Contribution of Imaging in the Diagnosis and Management of Type 1 Autoimmune Pancreatitis Associated with Main Bile Duct Cholangitis: A Case Report

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Abstract

Introduction: Autoimmune pancreatitis type 1 (AIP) is a rare condition characterized by inflammatory and fibrosing involvement of the pancreas, associated with elevated serum IgG4 and potential involvement of other organs. It often mimics a malignant tumor with which the differential diagnosis is often difficult. **Observation:** We report a case of AIP type 1, in a 70-year-old patient, with a heavy history of arterial hypertension, insulin-induced type II diabetes, overweight, gastric ulcer, ischemic heart disease with triple bypass, COPD, revealed by intense abdominal pain and acute evolution. The diagnosis was evoked on imaging, in front of an aspect of "sausage pancreas", with delayed contrast enhancement of the pancreas, loss of lobulations, and the presence of a fibrous peripancreatic shell. The biological and histological assessment confirmed the diagnosis and avoided inappropriate surgical treatment. **Conclusion:** AIP type 1 is a rare pathology whose diagnosis is difficult. Only a perfectly interpreted imagery can evoke it. The biological assessment, in particular, the elevation of the serum level of IgG4 and the histology make it possible to confirm the diagnosis. The challenge in its management is to avoid unnecessary surgery and/or inappropriate treatments. Imaging, in this context, plays a preponderant role, in particular, thanks to the dynamic injection sequences of gadolinium chelates on MRI which suggest the fibro-inflammatory nature of pancreatic lesions.

Keywords

Autoimmune Pancreatitis CT, MRI

1. Introduction

Autoimmune pancreatitis (AIP) is a condition that accounts for less than 2% of chronic pancreatitis cases [1]. The first cases of AIP were reported in 1961 by the Marseille team of Sarles *et al.* [2]. Two pitfalls should be avoided:

- Proposing pancreatic resection to a patient with pseudotumor AIP;
- Do not offer corticosteroid therapy to a patient suffering from pain related to AIP.

This is a diagnosis of elimination based on a number of arguments, among which imaging plays a key role.

Although the CT scan is currently the first-line examination for AIP, the problem posed by pseudo-tumoural forms simulating neoplastic involvement leads to additional investigations, the aim being to avoid inappropriate surgical resection and/or unnecessary treatments, as this benign condition is treated with simple corticosteroids [3] [4].

We report a case of IgG4 AIP associated with main biliary tract cholangitis, which demonstrates how imaging can suggest the diagnosis and illustrates the value of MRI in the lesion assessment, the demonstration of suggestive extra-pancreatic signs, and the post-treatment evaluation.

2. Observation

We report the case of a 70-year-old patient with a history of arterial hypertension, type II diabetes with insulin, overweight, gastric ulcer, ischemic heart disease with triple bypass surgery, and COPD. He was admitted to the emergency room for intense transfixing abdominal pain that had been evolving for a week, without any notion of reflux, without nausea or vomiting.

The clinical examination revealed a conscious and oriented patient, pain in the epigastrium, peri-umbilical and iliac fossae, radiating to the back. There was no defensiveness or contracture. Vitals showed a temperature of 36.5˚C, blood pressure 157/99mmhg, pulse 66 bpm, SpO_2 100%.

Biological findings included normocytic anaemia, normochromic 10 g/dL, hyperkalaemia 5.6 mmol/L, hyperglycaemia 20.4 mmol/L, minimal biological inflammatory syndrome with CRP 23 mg/L, lipasemia 98 IU/L. Tumour markers, CEA was normal and CA 19 - 9 slightly elevated. Liver function tests were normal.

Abdominal and pelvic CT scan (**Figure 1**) showed dilatation of the main bile duct to 12.5 mm with no visible obstruction, diffuse pancreatic hypertrophy with delayed contrast and loss of lobules, a hypodense peri-pancreatic sleeve and a 27 mm pseudocyst of the tail of the pancreas.

PET scan showed a hypertrophic, hypermetabolic pancreas.

MRI (Figure 2) subsequently showed a global T1 hyposignal pancreas, progressive enhancement of the infiltrated parenchyma, maximum and persistent in

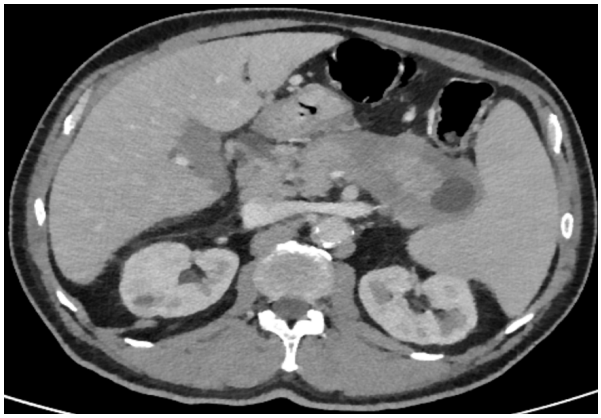

Figure 1. Abdominal CT scan with portal time acquisition, axial section, parenchymal window, showing diffuse pancreatic hypertrophy "sausage pancreas" with delayed contrast uptake and loss of lobulations, a hypodense peripancreatic sleeve and a 25 mm pseudocyst of the pancreatic tail.

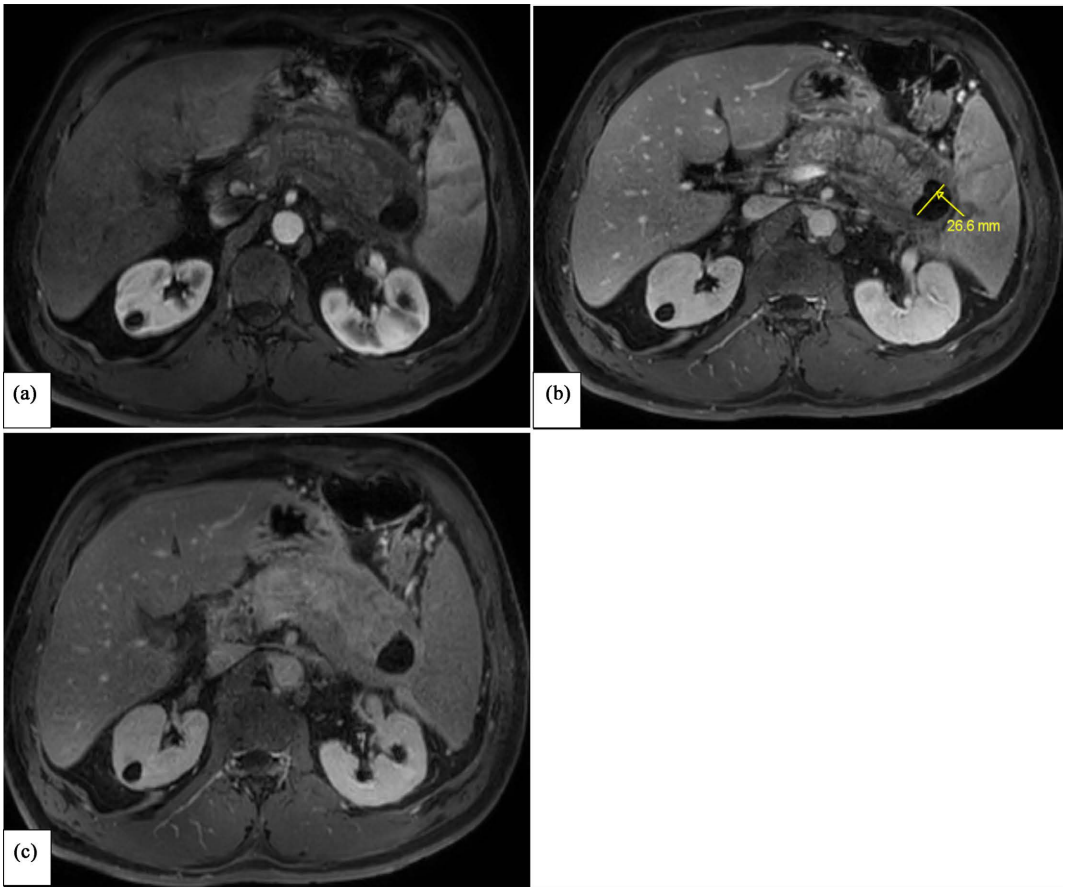

Figure 2. Pancreatic MRI in axial section. Dynamic T1-weighted sequences with suppression of the fat signal and injection of gadolinium chelates, showing a pancreas in global T1 hyposignal, a global "sausage pancreas" hypertrophy with loss of lobulations, a 27 mm pseudocyst of the tail, a fibrous peripancreatic shell aspect and a progressive enhancement of the infiltrated parenchyma, maximum and persistent at the late time, without any suspicious focal lesion. (a) Arterial time. (b) Portal time. (c) Late time.

the late period, without any suspicious focal lesions, loss of lobulations, global hypertrophy with a 27 mm pseudocyst of the tail and a fibrous peripancreatic shell appearance.

Autoimmune pancreatitis was suspected in view of these radiological findings. The diagnosis was therefore supported by a serum IgG4 assay which was higher than normal (195 mg/dL), echo-endoscopy which showed a major and homogeneous hypertrophy of the pancreas with marked peripheral oedema and cholangitis of the main biliary tract, and a biopsy which allowed histological confirmation and elimination of the differential diagnosis: tumour or lymphomatous infiltration. The diagnosis of AIP type 1 with cholangitis of the VBP was thus retained.

Corticosteroid therapy was initiated, and the evolution was marked by a rapid clinical improvement, progressive atrophy of the pancreatic gland on control MRI, disappearance of the peripancreatic fibrous shell and pseudocyst of the tail of the pancreas.

3. Discussion

Autoimmune pancreatitis type 1 is a rare cause of non-biliary non-alcoholic pancreatitis, especially in men between 50 and 70 years of age, typically presenting with jaundice or pancreatic pain, and accounts for 2% to 6% of chronic pancreatitis [5]. Described over the past 40 years, it was progressively individualised as an autoimmune entity in the 1990.

Clinical manifestations are often discreet or absent. Inaugural acute pancreatitis, as in our patient's case, is uncommon. AIP may be isolated or associated with one or more autoimmune diseases in 35% - 56% of cases. Among the most common are Goujerot-Sjögren's syndrome, primary sclerosing cholangitis, rheumatoid arthritis, retroperitoneal fibrosis and chronic inflammatory bowel disease. Type 2 diabetes may be present at diagnosis, as in our case [6]. Frequently, autoantibodies and increased serum gamma globulin levels, especially IgG4, are found but there is no clearly identified serological profile and the negative predictive value of autoantibodies is low [7] [8].

Imaging has an important place in the suspicion of type 1 AIP. As seen in our case, the typical parenchymal sign is the large sausage pancreas (head of pancreas > 3 cm, tail of pancreas > 2 cm thick) with delayed contrast. The existence of a hypodense or hypointense peripheral ring under the capsular is almost pathognomonic [9] [10]. The sensitivity and specificity of CT scans have been estimated at 86% and 95% respectively [11]. Abnormalities of the main bile duct (parietal irregularities, thickening, strictures and wall enhancement) may indicate associated cholangitis [12]. Hypodense infiltration may be seen around the intrahepatic bile ducts [4]. The radiological presentation is not always typical. Calcifications (during the course of the disease), peripancreatic nodes, venous abnormalities and pseudocysts have been described [13] [14]. A recent publication confirms the difficulty of imaging diagnosis by showing a non-negligible

proportion of pseudo-tumour AIP wrongly operated on patients followed for suspected degenerative chronic pancreatitis [15]. MRI is a very useful complement to CT. The excellent contrast resolution allows precise study of the infiltrated pancreatic parenchyma, which appears homogeneous, in T1 hyposignal. The boundaries with the healthy pancreas are often clear in pseudo-tumour forms. The peri-pancreatic border, if present, is hyposignal on all sequences. Rarely, a pseudo-capsule is found which enhances late after injection, indicating the existence of a fibrous shell developed around the pancreas, which was observed in our patient.

Positron emission tomography (PET), possibly coupled with a CT scan, is now part of the diagnostic arsenal. A recent study shows qualitative and semi-quantitative differences in 18-fluorodeoxyglucose uptake between AIP and pancreatic adenocarcinoma. However, the specificity of this method is only good if there is concomitant extra-pancreatic tracer uptake at specific sites (lacrimal and salivary glands, retroperitoneum, biliary tract, etc.) suggestive of an associated autoimmune disease [13]. Here again, access to this imaging modality remains difficult in routine. However, this imaging modality was performed in our case and revealed a large hypermetabolic pancreatic gland but did not reveal any extra-pancreatic involvement.

Histologically, AIP type 1 is characterised by the following criteria: dense lymphoplasmacytic infiltration of the parenchyma, periductal and interlobular fibrosis, arranged at least focally in a storiform manner and obliterating venulitis. The infiltrate is positive for anti-IgG4 antibody (>10 IgG4/HPF on biopsy or >50 IgG4/HPF on surgical specimen). The presence of these lesions indicates a diagnosis of AIP type 1. The pancreatic ducts may be stenotic but their epithelia are usually preserved. Indeed, granulocytic epithelial lesions (inflammatory infiltrate with epithelial destruction) are rather specific to AIP type 2 and were not found in our case. When associated, IgG4 cholangitis is characterised by fibrosis and infiltration of the bile ducts by IgG4 plasma cells. The IgG4+ plasma cell infiltrate is accompanied by an increase in serum IgG4 levels (>1.35 g/L) in patients with type 1 AIP. This increase is not specific and can be seen in pancreatic cancer. Elevation of the tumour marker CA19-9 is also possible in this condition [6], which was slightly elevated in our case. When histological criteria are present, the diagnosis of AIP is certain. However, taking samples of pancreatic tissue during echo-endoscopy is not always easy [7], but it was possible in our case to demonstrate cholangitis of the main biliary tract and to confirm histologically the type 1 AIP.

The evolution of AIP is generally favourable with total or partial regression (depending on the extent of fibrosis) of pancreatic and extra-pancreatic lesions under corticosteroid therapy. If an early relapse occurs, a new treatment with a higher dose is prescribed. Biliary drainage may be necessary if inflammatory involvement of the main bile duct is severe. Spontaneous remissions are possible.

In our case, the patient was treated with corticosteroids, the evolution was

very favourable with the complete regression of the clinical signs and the disappearance of the abnormalities observed on the different imaging modalities on the remote control MRI.

4. Conclusion

In summary, type 1 AIP is a rare pathology that can simulate a resectable pancreatic adenocarcinoma. Its diagnosis is difficult. Only a perfectly interpreted imagery can evoke it. The challenge in its management is to avoid unnecessary surgery and/or inappropriate treatments. Imaging plays a major role in this context, particularly thanks to the dynamic injection sequences of gadolinium chelates on MRI, which suggest the fibro-inflammatory nature of the pancreatic lesions. AIP is therefore a disease that deserves to be known by all, both by the clinician and by the surgeon and radiologist, as the diagnosis is often based on a combination of clinical and radiological evidence

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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