Autoimmune Pancreatitis Mimicking Pancreatic Cancer: A Case Report

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Abstract

Autoimmune pancreatitis is a rare condition, accounting for less than 5% of chronic pancreatitis, characterized by clinical, histopathological, radiographic, serologic, and therapeutic features. This benign disease resembles a pancreatic tumor both clinically and radiographically. We report the case of a 53-year-old man with no history who presented cholestatic jaundice and an image suggestive of the pancreatic tumor. A cephalic duodenopancreatectomy (Whipple’s operation) was performed and an anamnepathological examination of the operative specimen confirmed that it was type II autoimmune pancreatitis. The evolution was marked by the remission of clinical and biological signs. Accurate and timely diagnosis of autoimmune pancreatitis is particularly important because steroid therapy is effective and pancreatic resection is not necessary.

Keywords: Pancreatitis; Radiograph; Jaundice; Cancer; Plasma cells

Introduction

Autoimmune pancreatitis (AIP) is a particular form of recently individualized chronic pancreatitis that was sometimes confused with the diagnosis of pancreatic cancer. It was from single cases reporting cases of pancreatitis associated with hypergammaglobulinemia, primary biliary cirrhosis, or Sjögren’s syndrome [1-3] that the hypothesis of an autoimmune process was born. In the pathogenesis of certain pancreatitis [4,5]. The term PAI was introduced for the first time in 1995 by Yoshida et al. [6,7]. Since then, PAI has been individualized as a separate disease entity. It is very important for clinicians to distinguish between AIP and pancreatic cancer because of the treatments and prognoses are significantly different. Although the diagnosis of AIP has been improved, it remains a practical strategy to differentiate between AIP and pancreatic cancer.

Case Reports

It is a patient of 53 years, chronic smoking, admitted for a cholestatic jaundice, without fever nor digestive haemorrhage, which evolves for 15 days in a context of apyrexia and conservation of the general state. The biological assessment showed: Hb: 14.4 g/dl; TP: 100%; Total bilirubin: 18 mg/l; Direct bilirubin: 10 mg/l; Indirect bilirubin: 8 mg/l ASAT: 177 IU/l; ALT: 312 IU/l; PAL: 336 IU/l; GGT: 511 IU/l; ACE: 3.26 ng/ml; Ca19-9: 54.3 IU/ml. Abdominal ultrasound and BillIRM showed a tumoral process of the pancreatic head 30 × 30 mm in diameter with dilatation upstream of the main bile duct and intrahepatic bile ducts, a discreet dilatation of the Wirsung without secondary locations (Figure 1). Given the malignancy of the lesion, an exploratory laparotomy was performed, revealing a mass of the pancreas that is resectable and a Cephalic duodenopancreatectomy (Whipple’s operation) was performed and an anatomopathological examination of the operative specimen confirmed that it was type II autoimmune pancreatitis. The disease most often affects adults over the age of 50 [21], but the age range of PAI is broad (16-83 years) [22-24]. There are two types of autoimmune pancreatitis (PAI) identified to date [7]:

- Autoimmune type I pancreatitis, which corresponds to the pancreatic localization of an autoimmune systemic inflammatory disease recognized in the early 2000s. It represents at least 80% of PAI cases. In 2012, the name IgG4-related disease was adopted by the 1st international consensus on this systemic disease [8-10].

- Type II autoimmune pancreatitis, less common, of more recent discovery, which is an autoimmune disease more classical in its pathophysiology and which is associated in 20 to 30% of cases with chronic inflammatory bowel disease (MlCi) [7].

Histologically, it is chronic inflammatory pancreatitis with precise criteria that must all be observed to confirm the diagnosis [25-28]:

Four criteria for AIP type I

- Marked lympho-plasmocyte infiltration without neutrophil infiltration.

Discussion

PAI is currently defined as a pancreatic manifestation of an inflammatory and fibrotic systemic disease that may affect not only the pancreas but also various other viscera, particularly the bile ducts, salivary glands, retroperitoneum and lymph nodes [8-10]. All organs affected by the disease have a dense lymphoplasmocytic infiltrate rich in cells expressing IgG4 [11,12]. PAI is a rare disease. Its prevalence remains poorly specified. A national Japanese study estimated in 2002 the annual number of PAIs at 900, a calculated prevalence of 0.82 cases per 100,000 individuals [13-15]. This prevalence, derived from a Mayo Clinic study [16,17], did not represent an absolute prevalence of PAI. It was only its diffuse form and the prevalence was estimated from pancreatectomies performed for benign pancreatic disease, a rare condition in chronic pancreatitis [18-20].

PAI is at least twice as common in men than in women [18,21]. The disease most often affects adults over the age of 50 [21], but the age range of PAI is broad (16-83 years) [22-24]. There are two types of autoimmune pancreatitis (PAI) identified to date [7]:

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Storiform fibrosis (arciform).

Phlebitis obliterans.

Abundance of IgG4 plasma cells (>10 IgG4 plasma cells per large field). This lesion called LPSP for Lympho-Plasmocytic Sclerosing Pancreatitis is the signature of the type I PAI.

Two criteria for type II AIP

- Destruction of the interlobular and intralobular channels by neutrophils (called GEL for granulocytic epithelial lesions), lesions that are surrounded by lymphoplasmocytic infiltration and less abundant fibrosis than in the LPSP.

- No or few IgG4 plasma cells (<10 IgG4 plasma cells per large field). This IDCP lesion for Idiopathic Duct-centric Chronic Pancreatitis is the signature of type II autoimmune pancreatitis.

The clinical presentations of AIP are very variable. The most common acute clinical presentation is painful obstructive jaundice. It was observed in 65% of the cases of the study by Kim et al. [15], in 86% of cases that of Takvama et al. [29]. This presentation may closely resemble that of pancreatic cancer with, in particular, a tumor aspect of the pancreas head, as the case of our patient other symptoms have been described during IBD, such as abdominal pain. [15] On the other hand, true acute pancreatitis is exceptional [18]. This table would be more common in young people [24]. Elsewhere, it may be recurrent vomiting or weight loss [21,30]. Occasionally, extra-pancreatic manifestations may be present, such as salivary gland hypertrophy, hydrenephrosis associated with peritoneal fibrosis, and lymphadenopathy [31]. Lipasemia is moderately or very moderately elevated in 50% of cases.

It is very rarely >3N. Cholestasis (sometimes fluctuating, which must then attract attention) is in the foreground, observed in 60 to 85% of cases. It is rarely due to pancreatic disease itself (by bile compression). It almost always reports the sclerosing cholangitis associated with IgG4, and 15% of asymptomatic patients all free from cholestasis. Diabetes is observed in 65% of cases. It precedes the diagnosis of autoimmune pancreatitis in 1/3 of cases, is synchronous every other time and appears under corticosteroid therapy in 15% of cases [32,33]. The elevation of serum IgG4 levels is the key to diagnosis. At the threshold of 2.70 g/l, the criterion retained by the international consensus, the specificity is 99% but the sensitivity is only 53% [34]. In other words, half of type I AIPs are seronegative or not associated with IgG4 levels high enough to rule out the main differential diagnosis of pancreatic adenocarcinoma. No imaging technique makes it possible to carry out the diagnosis of AIP in an absolute way, however each makes it possible to bring additional arguments. The combination of several types of imaging (including tomodensitometry [CT] and MRI) is essential. The most typical abnormalities visualized in CT are an overall increase of the entire pancreatic gland associated with loss of lobulation. The smooth appearance of the contours gives an image in saussicce “sausage pancreas”. We can also note:

- A decrease in peripheral contrast enhancement at the origin of a halo or a peripheral ring;
- An involution of the tail of the pancreas;
- A contrast enhancement of the wall of the bile ducts thickened in the form of cockade;
- Tiered and suspended stenoses of the Wirsung canal without upstream dilatation;
- Focal pseudo-tumor forms not enhancing after injection of contrast enhancement (hypodense mass).

In the case of PAI that has been evolving for many years, calcifications and vascular abnormalities are possible [30,36]. MRI provides essential additional data. There is also a loss of intensity in the T1 phase and T2 hyperintense of the parenchyma correlated with inflammation of the gland [37]. The interest of the Petscan has been evaluated in special cases: response to treatment with corticosteroids and diagnostic tool in case of suspicion of cancer and it can detect the involvement of other organs (indirect argument) and it would “monitor” the activity of the disease [38,39]. There is no effective diagnostic tool to differentiate PAI from cancer with a sensitivity of 100% apart from a biopsy showing carcinomatous cells. A negative puncture does not formally eliminate the diagnosis of cancer and it is essential to know how to
repeat the biopsy puncture once or twice in case of strong suspicion [40–42]. In practice, in case of diagnostic doubt in the presence of a pancreatic mass associated with a dilation of the bile ducts, it is possible to propose an assay of serum IgG4, an abdominopelvic CT with thin sections centered on the pancreas, an MRI pancreatic with Wirsungo-MRI sequences and diffusion sequences, endoscopic ultrasound with puncture-biopsy of the mass. The value of serum Ca 19-9 is limited because a non-specific elevation of the marker is noted in cases of uncompensated diabetes or cholestasis.

Corticotherapy is the treatment of choice, consensual of the AIP. The evolution of symptoms may be so dramatic after a few days of treatment that the response to corticosteroids is an integral part of the Asian diagnostic criteria and HISORt [43]. Clinical and morphologic remission was obtained in 98% of patients treated versus 74% of untreated patients (p<0.001) [44]. The initial dose of oral prednisolone is 40 mg/day for 4 weeks and a decrease of 5 mg/week is recommended from the beginning of the improvement of symptoms. This equates to a total treatment duration of 12 weeks on average [29,35,45]. Because of the high rate of recurrence, especially in cases of biliary disease, monitoring is recommended: hepatic assessment (transaminases, alkaline phosphatase and total/conjugated bilirubin) and serum IgG4 assay every 3 months for 2 years and achievement of Pancreatic and biliary MRI every year for 2 years. In our case, the patient had certain clinical and radiological features that could impose a malignant tumor process. According to the diagnostic algorithms of Shimosegawa et al. [46], the absence of a characteristic characteristic of the PAI with CT or MRI in our patient would have required the search for another organ affected, the realization of an IgG4 serology, even a pancreatic biopsy whose results might have to guide diagnosis and therapeutic management.

Conclusion

Autoimmune pancreatitis is a rare but not exceptional disease. It is a real challenge for gastroenterologists and surgeons, because we must both avoid unnecessary pancreatic resections, and conversely do not delay a salvaging surgery with sterile corticotherapy, whose inefficiency must both avoid unnecessary pancreatectomies, and conversely do not impose a malignant tumor management.

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References