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Case Report **Hepato-Gastro-Enterology**

Autoimmune Pancreatitis Mimicking Pancreatic Cancer: Case Report

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Abstract

The positive diagnosis of autoimmune pancreatitis (AIP) can be challenging and poses a problem of differential diagnosis with pancreatic cancer. It's most sensitive and specific biological marker is the serum IgG4 level. Endoscopic ultrasound-guided fine-needle aspiration (EUS-FNA) plays a key role in the positive and differential diagnosis. Treatment is based on corticosteroid therapy. We report the case of a 43-year-old female patient admitted for cholestatic jaundice with a cephalic pancreatic mass on imaging simulating pancreatic cancer.

Keywords: autoimmune pancreatitis (AIP), corticosteroid therapy, cholestatic jaundice, pancreatic cancer.

INTRODUCTION

Autoimmune pancreatitis (AIP) is a rare form of chronic pancreatitis. Two types have been identified: type 1 which is the digestive manifestation of a systemic fibroinflammatory disease called IgG4-related disease, and type 2, which is a pancreas-specific disorder [1]. AIP often mimics pancreatic cancer and if diagnosed incorrectly leads to unnecessary pancreatectomy in 25% of cases [2]. The serum IgG4 level is its most sensitive and specific biological marker [3]. EUS-FNA plays an important role in the differential diagnosis of pancreatic cancer [4]. The treatment of choice is corticosteroids [5, 6].

CASE REPORT

A 43-year-old woman with a history of hypothyroidism under no current treatment and with no history of alcohol abuse presented with cholestatic jaundice with sudden onset in February 2021 which spontaneously resolved associated with moderate epigastric pain. There was no deterioration of the general condition.

On clinical examination, the patient was apyretic and subicteric. She had no hepatomegaly or splenomegaly. There were no lymph nodes. The biological analysis showed elevated levels of bilirubin 20 mg/l, alkaline phosphatase 549 UI/l, gamma-glutamyl transferase (GGT) 413 UI/l, alanine aminotransferase (ALT) 349 UI/l, aspartate aminotransferase (AST) 266 UI/l. There was no hyperlipasemia. The level of tumor markers (CAE and CA19-9) was normal. Abdominal CT scan showed dilation of the intrahepatic and extrahepatic biliary ducts as well as of the Wirsung upstream of an enlarged pancreatic head, supplemented by MRI which showed a cephalic pancreatic tumor with dilation of intrahepatic and extrahepatic ducts and the Wirsung. The patient was hospitalized in the surgical department for cephalic duodenopancreatectomy, but given the spontaneous regression of jaundice, an EUS was performed before and revealed a heterogeneous lesion of the head of the pancreas measuring (26 x 30 mm) without vascular invasion with a slight dilation of the intrahepatic ducts and the choledochal duct to 11 mm (Fig 1). The body and tail of the pancreas were heterogeneous and lobulated with a sinuous Wirsung: appearance that may suggest chronic pancreatitis of the autoimmune origin or other (Fig 2).

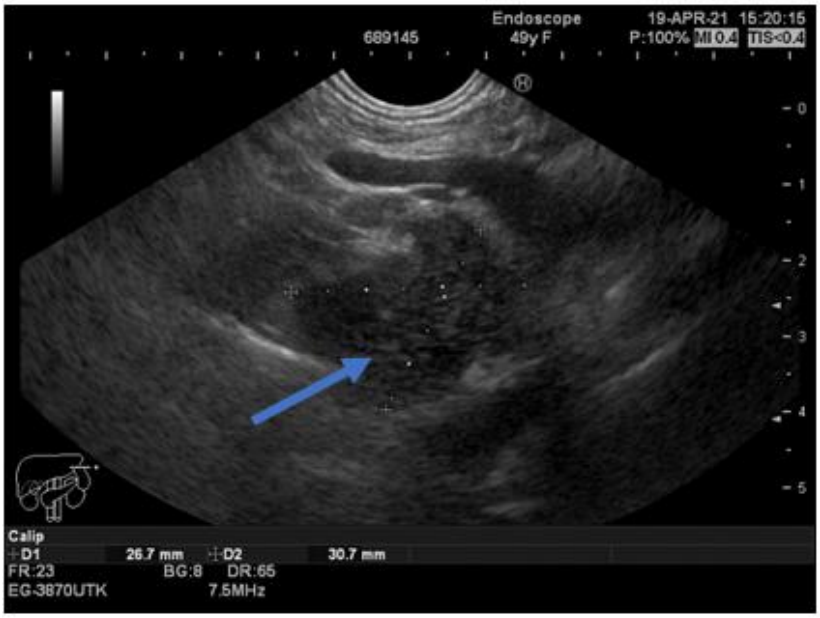

Figure 1: Ultrasound endoscopic appearance of the pancreatic masse (blue arow)

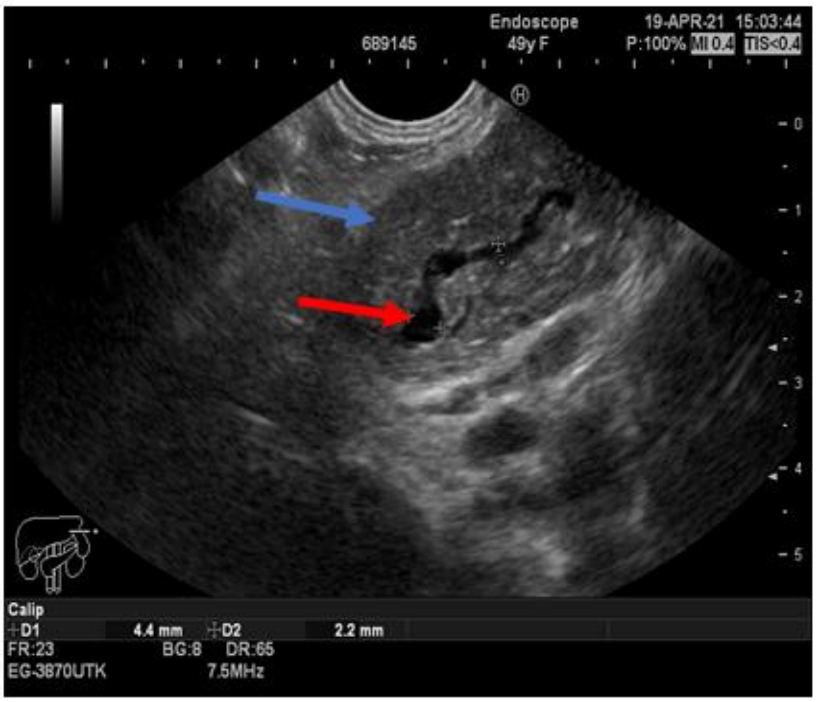

Figure 2: Ultrasound endoscopic appearance of the sinuous aspect of the Wirsung (red arrow) and the lobulated aspect of the pancreas (blue arrow)

The fine needle aspiration was done with a 22G needle and showed normal pancreatic tissue and no histological signs of malignancy (Fig 3). Given the spontaneous regression of jaundice and the preservation of the general condition, the presence of fluctuating biological cholestasis ++ with the negativity of tumor markers, as well as the presence of certain morphological signs (a lobulated aspect of the pancreas, thickening of the choledochal duct "Cholangitis", sinuous aspect of the Wirsung with moderate dilation upstream (<5 mm), and the absence of formal signs of malignancy (distant metastasis, vascular invasion), the diagnosis of AIP was strongly suspected.

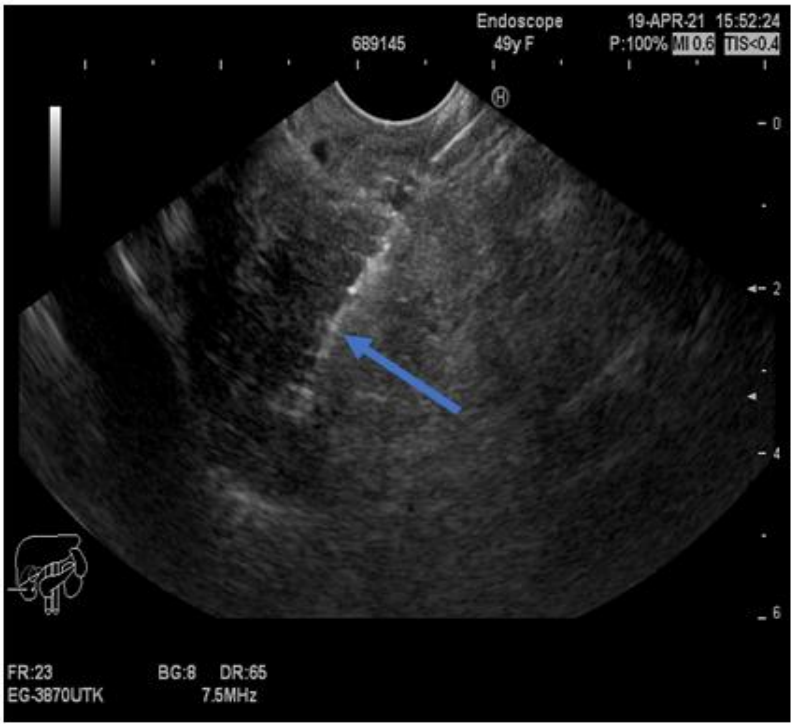

Figure 3: Ultrasound endoscopic guided fine-needle aspiration of the pancreatic masse (blue arrow)

Serum IgG4 was requested and returned positive at 4.48 g/l (5 times the normal value). The corticosteroid treatment was started in the dose of 0.6 mg/kg/day of prednisolone for 4 weeks and then reduced by 10 mg every week, up to half the dose, then reduced by 5 mg every week until stopping, with calcium supplements and salt and sugar-free diet. The evolution was good; there was no jaundice, no postprandial epigastric pain, with progressive normalization of the hepatic assessment and a decrease of the serum IgG4 level to 2.44 g/l (2.8 times the normal value) after one month of treatment, then to 1.63 g/L (1.8 times the normal value) at the second month. The MRI performed one month from the start of treatment showed a clear regression of the hypertrophy of the pancreatic head and the dilation of the intrahepatic duct.

DISCUSSION

AIP was first described by the team of Yoshida in 1995 as a form of chronic pancreatitis associated with autoimmune manifestations with a response to corticosteroid therapy (7).

Two forms of AIP have been described: type 1 and type 2. Type 1 is more common in Asia and constitutes 90% of the forms of AIP. It affects subjects over the age of 50 years old with a predominance of men. It is the most frequent digestive manifestation of IgG4 disease (12). Type 2 AIP affects younger subjects (30-50 years old), without gender predominance. It is often associated with IBD (12).

The clinical signs are variable and nonspecific. The most frequent symptom is cholestatic jaundice, which can pose a problem of differential diagnosis with pancreatic cancer (8), as was the case with our patient. It can also be manifested by abdominal pain and rarely by acute pancreatitis (9). In 15% of cases, patients are asymptomatic, and the discovery is then accidental on incident abdominal imaging (2).

There is no biological marker or circulating autoantibody specific for type 2 AIP whereas, in type 1 AIP, the only biological marker described in the literature is the serum level of IgG4. Above 1.35 g/l, the sensitivity is 75% and the specificity is 93% to differentiate it from pancreatic cancer. At the threshold of 2.70 g/l, the specificity is 99% but the sensitivity is only 53% (10). The combined serum assay of IgG4 and CA19-9 appears to be useful for the differential diagnosis of pancreatic cancer (11). In our observation, the initial serum IgG4 level was 5 times the normal value and the CA19-9 level was negative.

Histologically type 1 AIP is characterized by: storiform fibrosis, obliterating phlebitis, marked plasmacytic cell infiltration without neutrophils infiltration, and an abundance of IgG4 plasma cells (> 10 per large field) (2, 8). While type 2 AIP is characterized by two main criteria: infiltration by neutrophil leucocytes with the destruction of ductal cells and moderate or absent infiltration by IgG4 plasma cells (<10 per large field) [2, 13]. In our patient, the histological study ruled out the diagnosis of adenocarcinoma but did not show the specific lesions caused by AIP.

Radiological signs are common to both types of AIP, whether on CT scan or MRI. Typically, it is diffuse hypertrophy of the pancreas *"large sausage pancreas"* (40% cases) and characterized by its delayed contrast enhancement with a peripheral subcapsular ring (Hypodense or hypointense) which is almost pathognomonic. Less typical signs may be observed such as focal pancreatic hypertrophy (<50% of cases) or focal cephalic mass with dilation of biliary and pancreatic ducts upstream mimicking adenocarcinoma. Thrombosis of the splenic vein or mesenteric-portal confluence is possible. The ductal signs, it is typically long stenosis of the main pancreatic duct without upstream dilation or with moderate dilation (<5 mm) or multiple stenosis [2]. EUS-FNA plays an important role in the differential diagnosis of AIP with adenocarcinoma [14]. It can show ductitis as a change in the caliber of the Wirsung with alternating stenosis/dilation (> 3 mm and <5 mm) with thickening (hypo or hyperechoic of its wall), or cholangitis which, beyond 2 mm becomes very suggestive of type 1 AIP. The only way to get a histologic diagnosis of AIP is to get a "core biopsy". Until recently, only 19G needles and especially sharp 19G needles were able to provide this type of collection, but it was difficult if not impossible to use them through the duodenum, which greatly limited their value. Currently, there are needles capable of providing fragments for histopathological study (20G flexible cutting, 19G with flexible metal sheath, and 2 kinds of 22G cutting needles) [2]. In our patient, both abdominal CT scans and MRI showed a focal mass of the pancreatic head with upstream bi-ductal dilation mimicking adenocarcinoma. In our observation, EUS-FNA made it possible to rectify the diagnosis by revealing the lobulated aspect of the pancreas, the sinuous aspect of the Wirsung, and the cholangitis as well as the absence of histological signs of malignancy, thus preventing our patient from having an unnecessary pancreatectomy.

Treatment is based on corticosteroids and immunosuppressants. Corticosteroids can be prescribed using two protocols. One was used in Europe and the USA at a dose of 30-40 mg/day of prednisolone for 2-4 weeks. The dose is then reduced by 5 mg every 1–2 weeks to a maintenance dose of 5.0–7.5 mg/day. The maintenance treatment period varies, usually 12 weeks to 6 months. The second scheme is used in Asia where a dose of 0.6-1 mg/kg/day of prednisolone is administered for 2-4 weeks. The dose is then reduced by 5 mg at intervals of 1-2 weeks. However, the recommendation about the maintenance treatment period is different: 6 months to 3 years [15]. Relapses are common, especially in type 1 AIP. Maintenance treatment helps reduce this risk [16]. If corticosteroid therapy fails, AZP, 6-MP, and mofetil mycophenolate can be used [17]. Rituximab is a serious alternative to corticosteroid therapy allowing complete remission in 83% of cases in the event of relapse or intolerance to corticosteroid treatment [2, 18].

CONCLUSION

The diagnosis of AIP is difficult and relies on a body of clinical, biological, radiological, and histological arguments. AIP constitutes a real challenge for clinicians as it is necessary to avoid erroneous pancreatectomy whilst ensuring not to delay life-saving surgery with unnecessary corticosteroid therapy.

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