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Case Report

CASE REPORT

Autoimmune pancreatitis associated with retroperitoneal fibrosis mimicking cholangiocarcinoma

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Abstract

Autoimmune pancreatitis (AIP) is a rare systematic autoimmune disease that causes chronic pancreatitis. Type 1-AIP (IgG4-related disease) may involve other organs as well. In this report we are presenting a case of a 74-year-old man with obstructive abdominal pain jaundice, mild and a history of retroperitoneal fibrosis and hydronephrosis. Labs were remarkable for hyperbilirubinemia, high serum IgG4 levels, mildly elevated CA 19–9, elevated rheumatoid factor and new onset diabetes. MRI revealed pancreatic enlargement, dilated intrahepatic bile ducts and stricture of the distal common bile duct concerning for cholangiocarcinoma. EUS-FNA biopsy was negative for malignancy but showed findings of pancreatitis. The diagnosis of type 1-AIP was made and the patient was treated with steroids. After one month of treatment jaundice and MRI findings resolved. It is important to include AIP in the differential diagnosis of pancreatic conditions causing obstructive jaundice, especially in the presence of other autoimmune conditions like retroperitoneal fibrosis.

INTRODUCTION

Autoimmune pancreatitis (AIP) is inflammation of the pancreas caused by an autoimmune mechanism. There are two types of AIP. Type 1 AIP belongs in the broad category of IgG4-related diseases and can affect other organs like the bile duct, kidneys, salivary glands, lymph nodes and retroperitoneum. On the other hand, type 2 AIP affects only the pancreas and is not associated with high serum levels of IgG4 [1–3]. AIP usually presents with jaundice, mild abdominal pain and weight loss. Labs indicate elevated direct bilirubin, high levels of IgG4 (in type 1) and occasionally elevated carbohydrate antigen 19–9 (CA 19–9). AIP and pancreatic cancer can mimic each other and sometimes coexist [4]. Excluding cancer is always necessary before making the diagnosis of AIP [1]. The international consensus criteria are used to identify AIP, a benign disease which responds well to steroids and does not require surgery. In this case report we present a 74-year-old man that was initially considered to have pancreatic cancer and was eventually diagnosed with type 1 AIP associated with retroperitoneal fibrosis [5].

CASE REPORT

A 74-year-old man presented with jaundice and recurrent mild epigastric pain for 3 weeks. He had a history of idiopathic retroperitoneal fibrosis and right-sided hydronephrosis. Physical examination was remarkable for positive Courvoisier sign

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(palpable nontender gallbladder). Pancreatic cancer was initially high in the differential; however, AIP was also considered as a possibility given the history of retroperitoneal fibrosis. Labs showed evidence of cholestasis and mild transaminase elevation (ALP: 448 IU/L, GGT: 541 IU/L, Bili: 10.8 mg/dL, ALT: 93 IU/L, AST: 69 IU/L), hyperglycemia (new onset diabetes), high levels of CA 19–9 up to 303.4 U/mL (normal ref < 37), significantly elevated serum IgG4 at 3.100 g/L (normal ref < 2.0) and rheumatoid factor at 79.20 IU/mL (normal ref < 20). MRI of the abdomen revealed diffuse pancreatic enlargement with low density in T1 weighted images (Fig. 1), consistent with acute pancreatitis. It also showed dilatation of the extra-pancreatic common bile duct (CBD) (Fig. 2) and intrahepatic bile ducts and stricture of the intrapancreatic portion of CBD (starting 1.5 cm higher from the ampulla of Vater) with the characteristic 'beak sign' (Fig. 3). The pancreatic duct was mildly dilated. The gallbladder appeared normal with the presence of gallstones. The radiologist report suggested cholangiocarcinoma of the distal CBD as the most likely diagnosis, given the pathognomonic 'beak sign'. ERCP was attempted, which was unsuccessful due to significant stenosis of the intrapancreatic CBD not allowing the catheterization of CBD. Endoscopic ultrasound (EUS) demonstrated heterogenous-appearing parenchyma, lobularity and hyperechoic foci on the pancreas, severe stricture of the intrapancreatic portion of CBD and dilatation of extra-pancreatic CBD with wall thickening. EUS-FNA biopsy of the intrapancreatic CBD wall revealed fibrosis and lymphoplasmacytic infiltration without evidence of malignancy. According to the International consensus diagnostic criteria (ICDC) [6] for type 1 AIP the patient was diagnosed with type 1 AIP and started treatment with prednisolone 40 mg daily. A repeat MRI 1 month later showed resolution of the pancreatitis, CBD stricture and intrahepatic bile duct dilatation (Figs 4 and 5). There was no evidence of hydronephrosis. The patient's jaundice and abdominal pain resolved in the meantime. Serum IgG4 and CA 19–9 levels also normalized. The patient continued treatment with steroids for a total of 6 months.

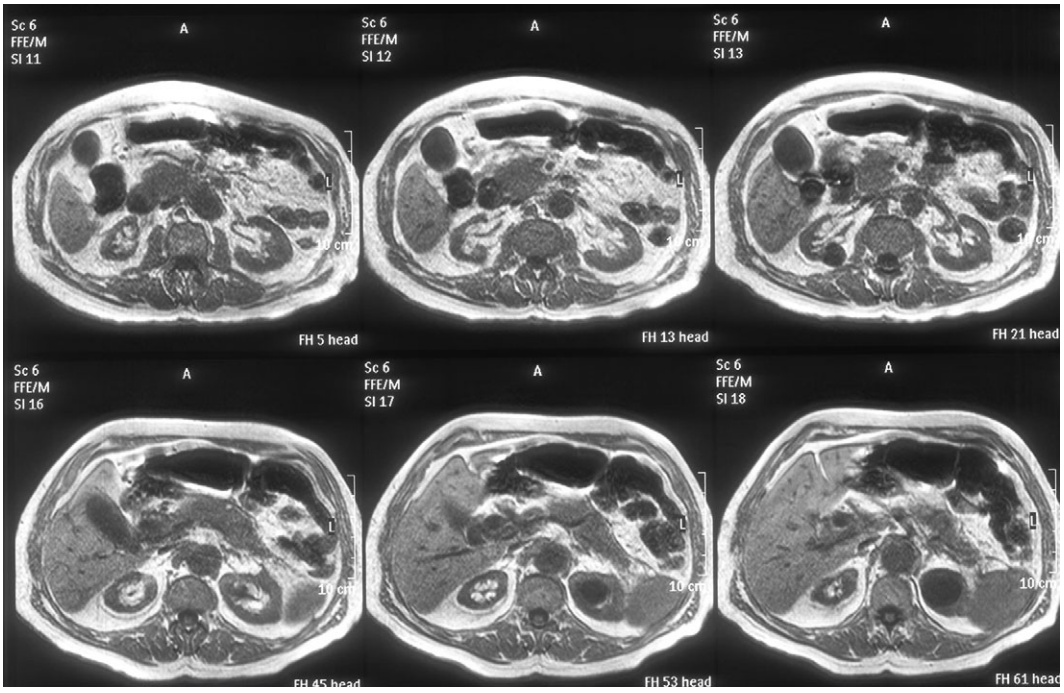

Figure 1: MRI showing diffuse pancreatic enlargement

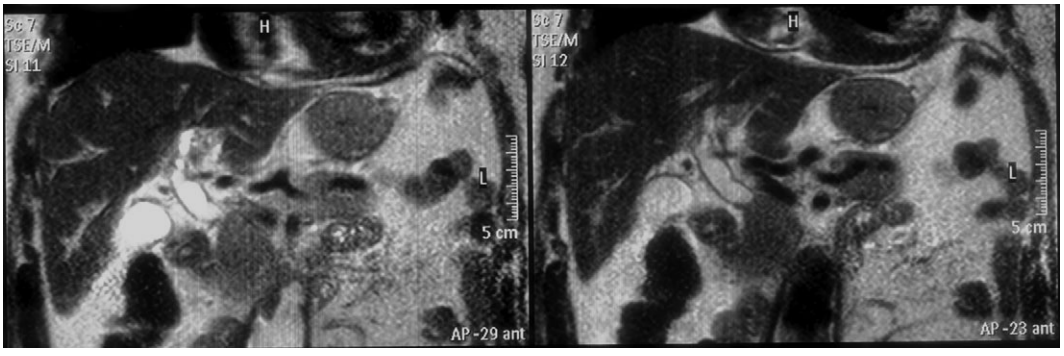

Figure 2: MRI showing dilatation of extra-pancreatic common bile duct

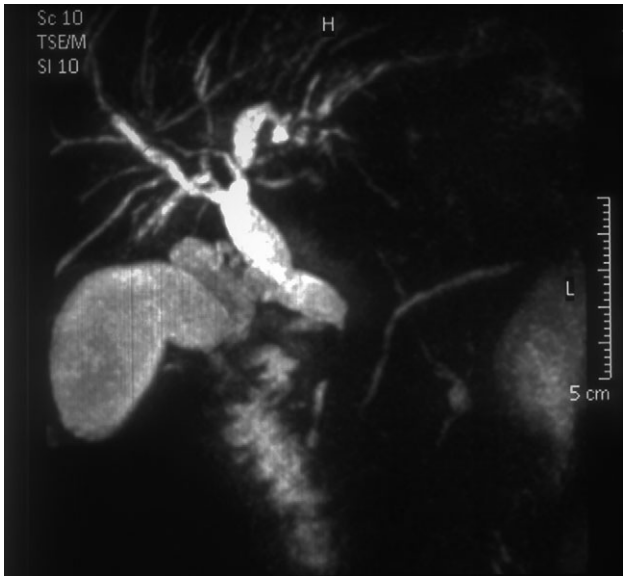

Figure 3: MRCP showing the characteristic 'beak sign' indicating cholangiocarcinoma

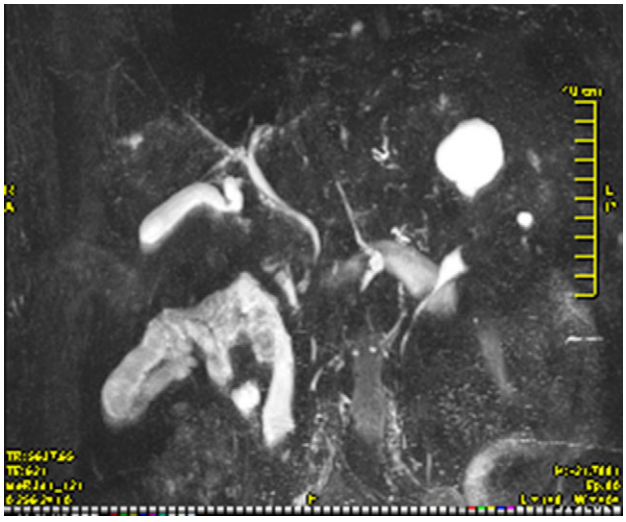

Figure 4: MRCP after 1 month of treatment with steroids showing resolution of common bile duct dilatation

DISCUSSION

AIP is a rare form of chronic pancreatitis caused by an autoimmune mechanism. There are two types of AIP, type 1 and type 2. Both types respond well to treatment with steroids. Type 2 AIP affects only the pancreas and is not associated with elevated serum IgG4. Type 1 AIP is an IgG4-related disease, characterized by high serum IgG4 levels (>135 mg/dL) and extra-pancreatic multiorgan involvement (retroperitoneum, bile duct, kidneys, lymph nodes, salivary glands). However, 20% of the patients in this group do not have elevated IgG4 (seronegative), which means that normal IgG4 levels do not exclude the diagnosis of type 1 AIP [7]. ICDC for AIP use clinical, serologic, pathology and imaging findings to accurately make the diagnosis of AIP. Pancreatic cancer should always be excluded, as sometimes AIP and malignancy may coexist [4]. Type 1 AIP presents clinically with obstructive jaundice, mild abdominal

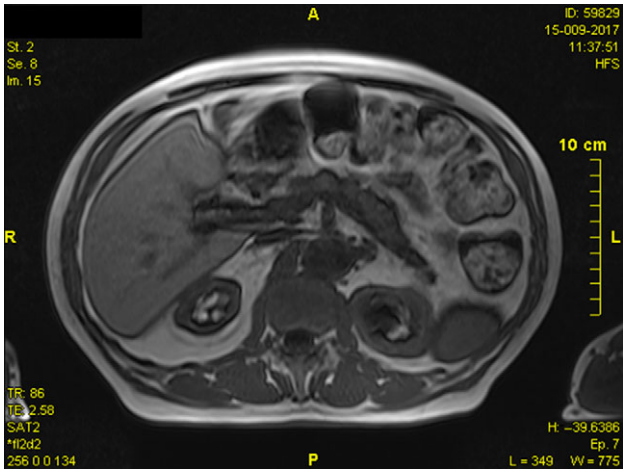

Figure 5: MRI after 1 month of treatment with steroids showing normal in size pancreatic parenchyma without signs of Inflammation

pain and weight loss. It can also present with symptoms from other organs, like retroperitoneal fibrosis, renal disease and cholangitis. New onset diabetes and positive rheumatoid factor may be present. Retroperitoneal fibrosis is the formation of fibrous tissue in the retroperitoneum that can lead to ureteral compression and hydronephrosis [5]. Obstructive jaundice is caused by infiltration of the CBD wall and pancreatic enlargement which causes stricture of the intrapancreatic portion of CBD, often giving the impression of cholangiocarcinoma on abdominal imaging (MRI). MRI findings in AIP include distal CBD stenosis, proximal CBD and biliary dilatation and diffuse pancreatic enlargement with low signal intensity in T1 images. EUS-FNA biopsy is helpful for the diagnosis of pancreatic cancer or cholangiocarcinoma of the CBD. The absence of malignant cells on the cytology does not always exclude the presence of malignancy as the samples obtain with the FNA are often inadequate [1, 2]. However, if there is a high suspicion for malignancy, a second EUS-FNA or EUS-guided core biopsy can be considered. In this case, the high serum levels of IgG4 and rheumatoid factor, extra-pancreatic manifestations (retroperitoneal fibrosis and hydronephrosis), EUS images and biopsy suggested the diagnosis of type 1 AIP. The patient received treatment with steroids with significant improvement in his clinical laboratory and imaging findings in less than a month. This also reinforced the initial diagnosis of type 1 AIP. Monitoring the IgG4 serum levels is helpful for evaluating the response to treatment and detecting possible recurrences. In case CA 19–9, jaundice and radiological features do not get back to normal, the patient should be re-examined for malignancy [3]. AIP is not a very common disease, but it should be included it in differential diagnosis of obstructive jaundice. High suspicion for AIP is required especially when the patient has extra-pancreatic involvement and pancreatic malignancy has been excluded. In cases when the diagnosis is not clear after the initial work-up, repeating biopsy prior to initiation of steroid therapy should be considered. When the patient meets all the diagnostic criteria of ICDC for AIP and malignancy is excluded, treatment with steroids should be initiated promptly. The patient should be monitored for appropriate response to treatment. If the clinical, laboratory and imaging findings do not improve, alternative diagnoses and/or treatments should be considered. It is very important to differentiate between pancreatic cancer and AIP in order to avoid unnecessary operations on patients without cancer.

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CONFLICT OF INTEREST STATEMENT

No conflicts of interest.

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CONSENT FOR PUBLICATION

Written consent for publication of the patients' details was obtained.

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