

Autoimmune Pancreatitis Presenting with Mass and Biliary Obstruction

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We report a case of autoimmune pancreatitis in a 31-year-old man with signs and symptoms of biliary tract obstruction. Evaluation with sonography, computed tomography, and ultimately endoscopic retrograde cholangiopancreatography, demonstrated a 5 by 2 cm hypoechoic, hypodense mass near the head of the pancreas that extended into the porta hepatis. Common bile duct obstruction with proximal dilatation was present. Following fine needle aspiration of the mass, cytology showed findings consistent with autoimmune pancreatitis. Treatment with common bile duct stent was followed by resolution of the patient's symptoms.

Introduction

The term autoimmune pancreatitis is used to describe a heterogeneous set of pancreatic conditions which are associated with characteristic laboratory and histologic findings [1]. The prevalence of autoimmune pancreatitis has been estimated to constitute approximately 5-10% of patients diagnosed with chronic pancreatitis [2]. The mean age at diagnosis of 55 years, and while autoimmune pancreatitis occurs in both sexes,

it is twice as common in men as in women [3]. Imaging of patients with autoimmune pancreatitis commonly reveals an enlarged homogeneously disorganized pancreas and regional lymphadenopathy. When confirmed with serum IgG4 levels or response to treatment, the diagnosis of autoimmune pancreatitis can be made.

Case Report

A 31-year-old man originally from Brazil presented to the Emergency Department with a chief complaint of one week of moderate epigastric pain accompanied by nausea and vomiting. The pain was of moderate severity and was burning in nature, with radiation to the right and left upper quadrants but not his back. He also noted that his vomiting occurred after every meal, and his wife confirmed that he had lost weight (exact amount was unknown) in the previous week in association with his inadequate nutritional intake. Upon further questioning, he also reported a one-week history of jaundice and dark brown urine as well as a four-day history of acholic diarrhea. He noted that he had never experienced symp-

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Abbreviations: CT, computed tomography; ERCP, endoscopic retrograde cholangiopancreatography; MRI, magnetic resonance imaging

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toms like this on any previous occasions. He denied any history of known hepatitis, intravenous drug use, tattoos, infectious exposure, or any recent use of medications or illicit drugs. The patient was further questioned about risk factors for acute pancreatitis, pancreatic cancer, and lymphoma. He denied any significant history of alcohol use, stating that he limited his consumption to approximately one beer per month, and he also denied any history of colicky abdominal pain or known gallstones. He had no smoking history or family history of gastrointestinal cancer or any other gastrointestinal disease. He denied any night sweats, fevers, lymphadenopathy, fatigue, or weight loss over the preceding months. He had previously been in excellent health and had an unremarkable past medical history.

On physical exam in the Emergency Department, the patient was a thin, jaundiced man who appeared his stated age. Vital signs were all within normal ranges. Cardiac and pulmonary exams were both normal, and the patient had no palpable lymphadenopathy of the neck or supraclavicular region. The abdominal exam revealed tenderness to palpation throughout the upper abdomen and right lower quadrant; however, the remainder of the examination was unremarkable, with normal bowel sounds, no palpable masses or organomegaly, no fluid wave, no Murphy's sign, and no evidence of prior trauma or incisions. Labs drawn in the Emergency Department showed elevated levels of alkaline phosphatase, total and direct bilirubin, AST and ALT; urinalysis also showed dark urine containing

elevated levels of bilirubin. Lipase and amylase were not significantly elevated. CBC and differential were normal except for a hematocrit of 41.4.

Based on the physical exam and initial laboratory data, the patient was presumed to have a conjugated hyperbilirubinemia caused by obstruction of the common bile duct; he was therefore sent for ultrasound imaging (Fig. 1). Abdominal ultrasound showed a 5 x 2 cm hypoechoic mass in the region of the pancreatic head as well as a 1 x 2 cm hypoechoic area in the porta hepatis. A 9 mm dilatation of the common bile duct was also visualized, but there was no evidence of intrahepatic biliary ductal dilatation. No pericholecystic fluid, gallbladder wall thickening, or cholelithiasis was apparent. Based on this imaging, the patient was suspected to have a pancreatic neoplasm constricting the common bile duct, with possible lymphadenopathy of the porta hepatis.

A pancreatic CT with oral and intravenous contrast was performed on the following day to evaluate this possibility. The CT showed a 5 x 3 cm enhancing, heterogeneous mass located in the region of the pancreatic head, with extension into the porta hepatis (Fig. 2). Slight intrahepatic duct dilatation and distension of the gallbladder were also noted, and the possibilities of enlarged lymph nodes and lymphoma were raised.

The patient was scheduled for ERCP (Fig. 3) and stenting of the common bile duct two days following his presentation to the Emergency Department. ERCP showed a normal pancreatic duct, non-beaded narrow-

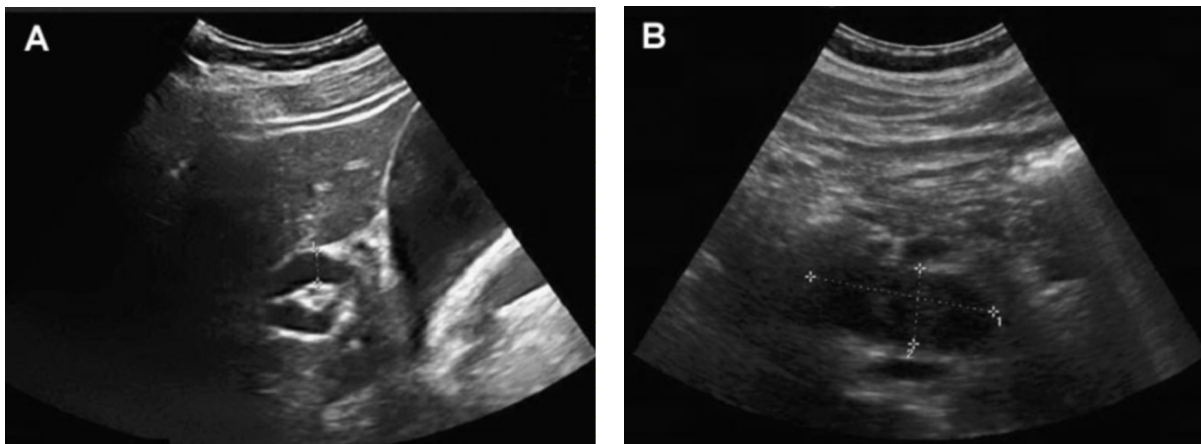


Figure 1A-B. 31-year-old man with autoimmune pancreatitis. Sonography shows (A) common bile duct dilatation without intrahepatic biliary ductal dilatation, and (B) hypoechoic mass in the region of the pancreatic head.

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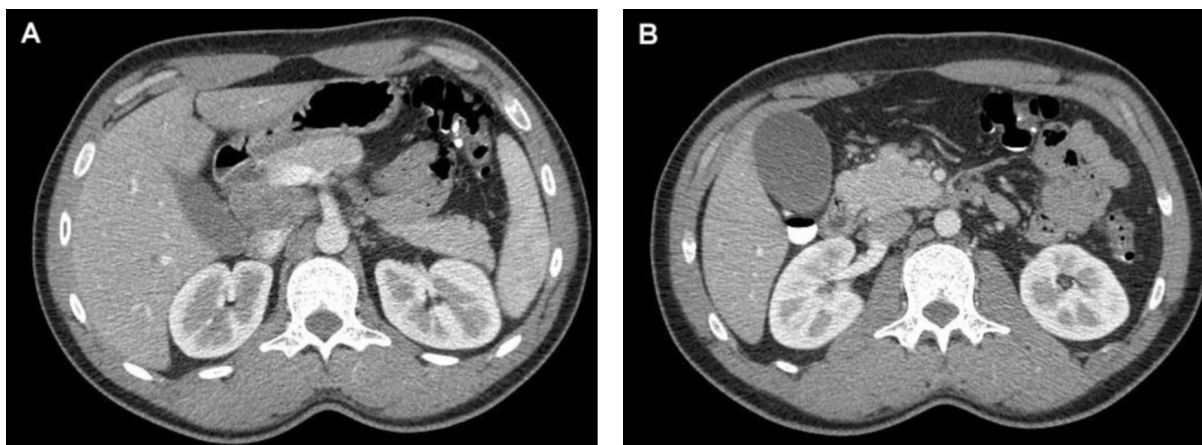


Figure 2A-B. CT of the abdomen shows (A) enhancing, heterogeneous pancreatic head mass, and (B) slight intrahepatic duct dilatation and gallbladder distension

ing of the distal common bile duct, and dilatation of the common bile duct proximal to the area of stricture. A stent was placed across the site of narrowing.

Ultrasound-guided fine needle aspiration of the pancreatic head mass was performed to assess further the nature of the lesion. Cytology showed benign glandular cells, neutrophils, lymphocytes, plasma cells, and fibrosis, consistent with either chronic pancreatitis or autoimmune pancreatitis. Based on the constellation of imaging findings, cytology, and clinical history, a diagnosis of autoimmune pancreatitis was proposed. Despite experiencing a brief post-ERCP pancreatitis, the patient's symptoms largely resolved in the week following the stenting; the patient's hyperbilirubinemia had also decreased. Additional laboratory tests including CEA, Ca-19-9, IgG4, ANA, strongyloides and schistosomiasis serologies were all negative.

Discussion

Autoimmune pancreatitis is a rare disorder of unknown etiology that can easily masquerade as a pancreatic or biliary neoplasm. Clinically, patients with autoimmune pancreatitis do not usually present with severe abdominal pain or acute pancreatitis, but jaundice is common (63% in one large series) [2]. Imaging in cases of autoimmune pancreatitis is often non-specific, but nevertheless serves as one of the cornerstones of diagno-

sis of the disorder. Classic CT findings include a focal mass in the head of the pancreas [4] or sausage-shaped enlargement of the pancreas with homogeneous attenuation, moderate enhancement, and a peripheral rim of hypoattenuating 'halo' [1]. Normal lobular architecture of the gland is often lost, but fat stranding is generally limited, and some enlargement of regional lymph nodes is typical. Distinguishing autoimmune pancreatitis from pancreatic carcinoma on CT is often not possible, and patients can present on CT with extra-pancreatic findings, including focal lesions in the lung, kidneys, and peri-aortic soft tissue. Neither MRI nor ultrasound has been found to provide more definitive diagnostic assistance [1].

An increase in serum IgG4 levels is common in autoimmune pancreatitis, and autoantibodies against lactoferrin and carbonic anhydrase II have also been reported. Additionally, autoimmune pancreatitis is frequently associated with other autoimmune conditions, including rheumatoid arthritis, Sjogren's syndrome, and inflammatory conditions [1, 5]. Still, the etiology of autoimmune pancreatitis is not well understood. On pathology, autoimmune pancreatitis has a typical histologic appearance, with a "periductal collar" that consists of an infiltrate of lymphocytes and plasma cells [6]. Some experts believe that the gold standard for diagnosis of autoimmune pancreatitis is pathology that shows such an infiltration and periductal fibrosis. However, such pathology can also be seen in chronic pancreatitis related

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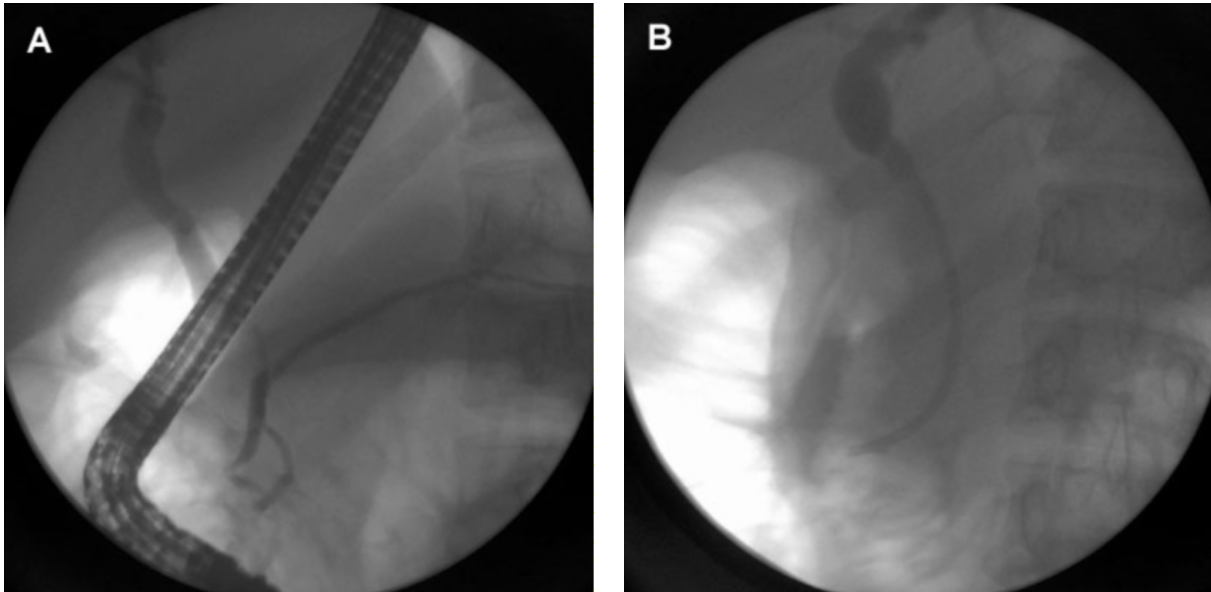


Figure 3A-B. ERCP shows (A) normal pancreatic duct, non-beaded narrowing of the distal common bile duct, and (B) proximal dilatation of the common bile duct.

to alcohol use [1]. Such pathologic specimens can be acquired by laparotomy, though fine needle aspiration and core biopsy have also been used.

There is a lack of consensus on the diagnosis of autoimmune pancreatitis, but a recent review suggested an algorithm for establishing the diagnosis. If diffuse gland enlargement is seen, laboratory tests should be performed to search for evidence of an autoimmune process, to include IgG4, IgG, antilactoferrin antibody, anti-carbonic anhydrase II antibody, anti-smooth-muscle antibody, and antinuclear antibody. If there is laboratory evidence of autoimmune disease-or if there is a lack of laboratory evidence but a presence of suggestive pathology (either through biopsy or fine needle aspirate)-then a trial of corticosteroid treatment is indicated. If this therapy produces a characteristically quick and sustained response, then the diagnosis of autoimmune pancreatitis is confirmed. If there is a paucity of laboratory or pathologic evidence, then surgical resection of the area of focal mass is warranted, and if the pathology of the surgical resection does show findings consistent with autoimmune pancreatitis, then a trial of corticosteroid therapy is reasonable [1]. In each of the above steps, a quick and sustained response to corticoid

therapy is considered diagnostic for autoimmune pancreatitis [7,8]. Corticosteroids also produce a resolution of any associated focal lesions, including associated lung and kidney lesions [1].

Our case likely represents a presentation of autoimmune pancreatitis due to the characteristic imaging (large, diffusely disorganized mass in the area of the pancreas and regional lymphadenopathy) and the results of the fine needle aspirate (lymphocytic infiltrate and plasma cell infiltrate). As discussed, pancreatic cancer and lymphoma were originally considered in the differential but were ruled out based on cytology. Primary sclerosing cholangitis was considered, but this diagnosis is less likely because in this disorder there is often a beaded narrowing of the distal common bile duct, whereas our patient presented with non-beaded narrowing. Alcoholic pancreatitis was also considered, but patient and his family report no history of alcohol use. This case demonstrates typical clinical features of autoimmune pancreatitis including moderate pain, jaundice, normal pancreatic enzymes, and imaging that was initially suggestive of pancreatic cancer.

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