

REVIEW ARTICLE

CURRENT CONCEPTS

Autoimmune Pancreatitis

Dmitry L. Finkelberg, M.D., Dushyant Sahani, M.D., Vikram Deshpande, M.D.,
and William R. Brugge, M.D.

From the Departments of Medicine (Gastrointestinal Unit) (D.L.F., W.R.B.), Radiology (D.S.), and Pathology (V.D.), Massachusetts General Hospital, Boston. Address reprint requests to Dr. Brugge at the Gastrointestinal Unit, Blake 452c, Massachusetts General Hospital, 55 Fruit St., Boston, MA 02114, or at wbrugge@partners.org.

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THE TERM “AUTOIMMUNE PANCREATITIS” (OR “AUTOIMMUNE-RELATED pancreatitis”) was introduced by Yoshida et al. in 1995 to describe the form of chronic pancreatitis that is associated with autoimmune manifestations revealed on laboratory, histologic, and clinical testing.¹ In the past 10 years, interest in autoimmune pancreatitis has grown because of an increasing ability to diagnose it with the use of new markers of disease and pancreatic biopsy. Treatment of autoimmune pancreatitis with corticosteroids leads to the rapid and sustained resolution of pancreatic mass lesions, biliary obstruction, and pancreatic-duct strictures, which has stimulated widespread interest in this condition from gastroenterologists, endoscopists, pathologists, and surgeons.

DEFINITION

Autoimmune pancreatitis is a type of chronic pancreatitis characterized by an autoimmune inflammatory process in which prominent lymphocyte infiltration with associated fibrosis of the pancreas causes organ dysfunction. For the past four decades, various morphologic descriptions have been proposed to characterize this disease: nonalcoholic duct-destructive chronic pancreatitis,² lymphoplasmacytic sclerosing pancreatitis with cholangitis,³ chronic sclerosing pancreatitis,⁴ pseudotumorous pancreatitis,⁵ and duct-narrowing chronic pancreatitis.⁶ Recently, the term “autoimmune pancreatitis” has become widely accepted, although it is apparent that autoimmune pancreatitis is a heterogeneous disease.^{7,8}

EPIDEMIOLOGY

Autoimmune pancreatitis is rare. Although there has been an increase in the number of reports of autoimmune pancreatitis in the medical literature in the past 10 years, the overall number of patients is still relatively small. The overall prevalence and incidence of the disease have yet to be determined. Three series have reported the prevalence of autoimmune pancreatitis as between 5 and 6% of all patients with chronic pancreatitis.⁹ According to the only series from the United States, 11% of patients (27 of 254) with chronic pancreatitis received a diagnosis of autoimmune pancreatitis based on histologic findings.¹⁰ Clinical or biochemical autoimmune stigmata are present in 40% of patients with idiopathic pancreatitis.¹¹ Autoimmune pancreatitis occurs in both sexes, but it is at least twice as common in men as in women. Patients vary widely in age; most are older than 50 years.^{9,12}

CAUSE AND PATHOGENESIS

Although the cause of autoimmune pancreatitis is unknown, current evidence strongly suggests an autoimmune basis for this disease.¹²⁻¹⁵ Like other autoimmune

diseases, autoimmune pancreatitis is frequently associated with rheumatoid arthritis, Sjögren's syndrome, and inflammatory bowel disease. Kawa et al. found an association of the DRB1*0405–DQB1*0401 HLA haplotype with autoimmune pancreatitis in the Japanese population.¹⁶ Immunologic abnormalities including hypergammaglobulinemia, elevated serum IgG4 levels, and the presence of autoantibodies against carbonic anhydrase and lactoferrin are important markers of the disease. However, little is known about its actual pathogenesis. Although elevated serum IgG4 levels have been linked to autoimmune pancreatitis, this bispecific and functionally monovalent antibody may merely represent a secondary response to a yet-unidentified primary trigger of the inflammatory process.

Autoantibodies against lactoferrin and carbonic anhydrase II have been identified as potential serologic markers of autoimmune pancreatitis.^{12,13} Most of the organs involved in autoimmune pancreatitis (including the lung, biliary tree, and renal tubules) contain intracytoplasmic carbonic anhydrase.¹⁷ Pancreatitis with histologic features similar to those associated with autoimmune pancreatitis in humans has been shown to develop in neonatal mice that have undergone thymectomy and have been immunized with lactoferrin or carbonic anhydrase II.¹⁴ Although these antibodies are frequently identified in patients with autoimmune pancreatitis, it is unlikely that they are the primary mechanism of this disease. As compared with patients with other forms of chronic pancreatitis, those with autoimmune pancreatitis have greater numbers of peripheral-blood CD4+ T lymphocytes that secrete greater amounts of interferon gamma. The administration of amylase-sensitized CD4+ T lymphocytes in rats can produce a model of autoimmune pancreatitis.¹⁵

PATHOLOGY

A diffusely indurated and firm pancreas on gross examination characterizes autoimmune pancreatitis. In a subgroup of patients, a focal mass is apparent in the pancreas. The histologic hallmark of this disease is a collar-like periductal infiltrate composed of lymphocytes and plasma cells (Fig. 1).^{7,18,19} Occasionally, periductal nonnecrotizing epithelioid-cell granulomas are also seen. Most lymphocytes in the infiltrate are CD8+ and CD4+ T lymphocytes, with fewer B lymphocytes. The interlobular septa are thickened by a proliferation

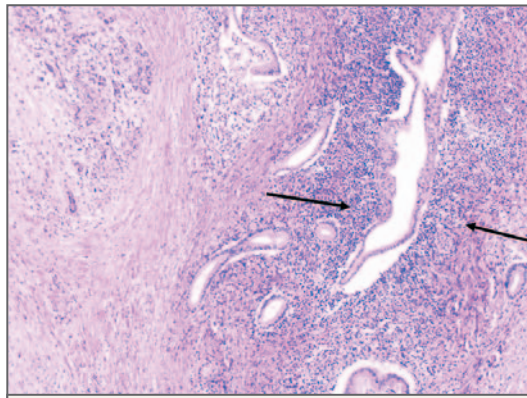


Figure 1. Histologic Characteristics of Autoimmune Pancreatitis, Including a Periductal Collar of Lymphoplasmacytic Inflammation (Arrows) (Hematoxylin and Eosin).

of myofibroblasts and infiltrated by lymphocytes and plasma cells.

In systemic autoimmune pancreatitis, the gallbladder, bile ducts, kidney, lung, and salivary glands are often involved with a dense lymphoplasmacytic infiltrate, accompanied by proliferation of myofibroblasts. IgG4-positive plasma cells have been found both within the pancreas and at the extrapancreatic sites of involvement.²⁰ Focal infiltration of the stomach, duodenum, and colon has been detected endoscopically and confirmed by histologic analysis.²¹

CLINICAL FEATURES

Patients with autoimmune pancreatitis present with a wide variety of symptoms, but severe abdominal pain or acute pancreatitis is unusual. In a large series, 63% of patients had jaundice and 35% of patients had abdominal pain.⁹ Most symptoms associated with autoimmune pancreatitis are responsive to corticosteroid therapy.

Occasionally, patients present with symptoms related to extrapancreatic organ involvement. Inflammatory bowel disease is frequently associated with autoimmune pancreatitis. In one series, 9 of 53 patients (17%) with autoimmune pancreatitis had either ulcerative colitis or, less commonly, Crohn's disease.⁷ However, the presence of diffuse monomorphic or long segmental strictures of the intrapancreatic bile duct is suggestive of an autoimmune process. Band-like strictures or focal lesions causing a beaded or pruned-tree appearance are more frequent in patients with primary sclerosing cholangitis.²² These distinctions have

important clinical implications, since biliary-duct strictures associated with autoimmune pancreatitis can respond dramatically to steroid treatment, in contrast to the poor response of strictures associated with primary sclerosing cholangitis.

Extrapancreatic manifestations of autoimmune pancreatitis may involve the lungs and kidneys. Pulmonary involvement may result in discrete or diffuse nodules, infiltrates, and adenopathy in the lung. Renal manifestations of autoimmune pancreatitis are mild renal insufficiency or multiple low-attenuation renal lesions on computed tomography (CT).^{23,24} Microscopy of these lesions reveals a destructive lymphoplasmacytic infiltrate containing IgG4-positive plasma cells.

DIAGNOSTIC IMAGING

The classic appearance of the pancreas on abdominal CT in patients with diffuse pancreatic involvement is sausage-shaped enlargement with homogeneous attenuation, moderate enhancement, and the peripheral rim of a hypoattenuation “halo” (Fig. 2). A loss of lobularity is commonly seen. Peripancreatic fat stranding is usually minimal. In long-standing autoimmune pancreatitis, involution of the pancreatic tail is almost always evident. Mild enlargement of the regional lymph nodes is also common. Focal pancreatic involvement is encountered more often in the head of the pancreas and typically appears as a low-attenuation or an isoattenuation mass. Therefore, the differential diagnosis of the focal form of autoimmune pancreatitis rather than pancreatic carcinoma can be very difficult on the basis of CT imaging only. However, the finding of diffuse pancreatic-ductal narrowing is highly diagnostic of autoimmune pancreatitis.²⁵ Focal lesions in the lungs, kidneys, or soft tissue around the aorta, described as “inflammatory pseudotumors,” will resolve after treatment with corticosteroids.²⁵ Response to corticosteroids appears on CT as resolution of the pancreatic enlargement and improvement or absence of the peripancreatic rim of the hypoattenuation “halo.” Likewise, pancreatic and biliary strictures may also resolve partially or completely. Morphologic changes can occur as early as 1 to 2 weeks after initiation of therapy.

The hallmark finding on endoscopic retrograde cholangiopancreatography (ERCP) in patients with autoimmune pancreatitis is a focal, diffuse or segmental attenuation of the main pancreatic duct and the disappearance of right-angled branches



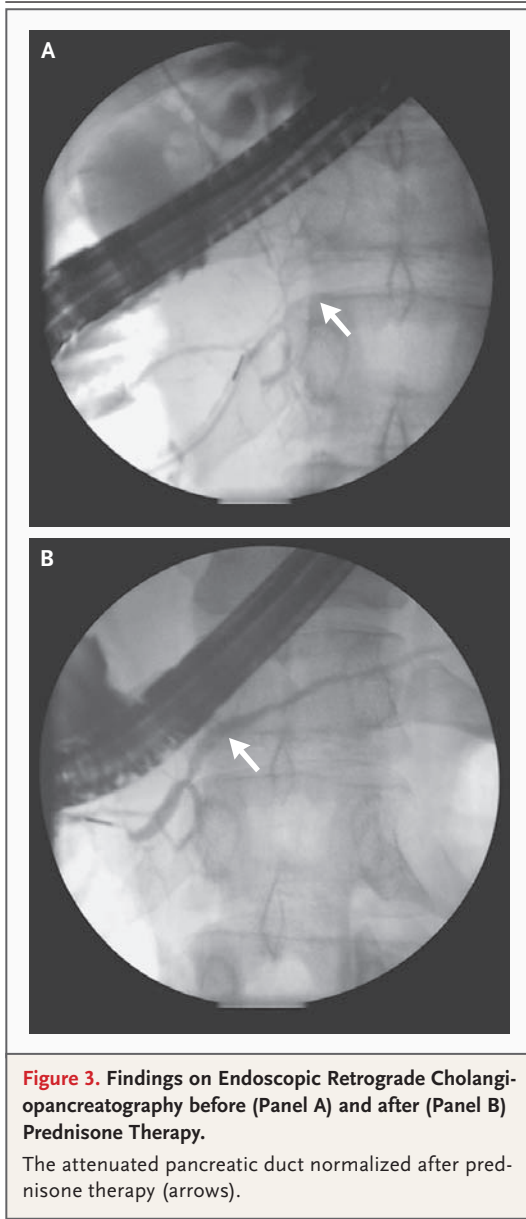
Figure 2. CT Scan Showing Typical Features of Autoimmune Pancreatitis: Diffuse Enlargement of the Pancreas with Homogeneous Attenuation and the Peripheral Rim of a Hypoattenuation “Halo” (Arrow).

An attenuated pancreatic duct was also present (not shown).

(Fig. 3). The main pancreatic duct adjacent to or upstream of the strictures is minimally dilated. The other common findings on ERCP are narrowing of the intrapancreatic portion of the common bile duct, irregular narrowing of extrahepatic bile ducts, and less frequently, enlarged intrahepatic bile ducts.^{9,25}

Because of its ability to show the gland in detail and to direct fine-needle aspiration, endoscopic ultrasonography is an important tool in the diagnosis of autoimmune pancreatitis and its differentiation from other pancreatic diseases. The most common finding on endoscopic ultrasonography is diffuse or focal pancreatic enlargement along with a diffusely hypoechoic parenchyma, similar to findings on transabdominal ultrasonography.^{26,27} Fine-needle aspiration guided by endoscopic ultrasonography or core biopsy of the pancreas may result in the cytologic or histologic diagnosis of autoimmune pancreatitis, but this approach to tissue acquisition has not been evaluated in large trials.²⁷

The role of transabdominal ultrasonography and magnetic resonance imaging (MRI) in the diagnosis of autoimmune pancreatitis is not well established. Ultrasonographic images of the pancreas, obtained transabdominally, are rarely diagnostic of autoimmune pancreatitis. Furthermore, findings on ultrasonography may be similar for autoimmune pancreatitis and for other forms of acute and chronic pancreatitis. The use of corticosteroid therapy results in improvement in diffuse gland enlargement and abnormal vascular-



ity.²⁸ Findings on MRI and magnetic resonance cholangiopancreatography (MRCP) in patients with typical autoimmune pancreatitis include a diffusely enlarged gland. As on CT, a peripheral rim of the hypointense “halo” can be seen on MRI. In addition, diffuse attenuation of the main pancreatic duct or a small, strictured main pancreatic duct may be present on MRCP.²⁹

DIAGNOSTIC CRITERIA

A set of diagnostic criteria has been proposed by the Japan Pancreas Society in an attempt to dif-

ferentiate autoimmune pancreatitis from other forms of chronic pancreatitis and pancreatic cancer.³⁰ The criteria are based on a combination of the findings of imaging, laboratory testing, and histologic analysis. The criteria have been slightly modified, and the required findings for each method are listed in Table 1. In an appropriate clinical setting, the typical findings on imaging along with supportive abnormal laboratory or histologic findings are sufficient for a diagnosis of autoimmune pancreatitis. These findings often form the basis of the decision to initiate corticosteroid therapy.

Several other investigators have proposed diagnostic criteria for autoimmune pancreatitis. Frulloni et al. have proposed the use of a combination of histologic and cytologic findings, the association with other autoimmune diseases, and the response to steroid therapy.³¹ Aparisi et al. have proposed a scoring system based on clinical presentation, morphologic findings, and laboratory results.¹³

The pathological features of lymphoplasmacytic infiltration and fibrosis are often used as the gold standard for the diagnosis of autoimmune pancreatitis (Fig. 1).¹⁰ However, these findings are also seen in patients with alcohol-induced chronic pancreatitis.^{8,32} The criteria of the Japan Pancreas Society do not require histologic evidence for a diagnosis of autoimmune pancreatitis.³⁰ Without a laparotomy, obtaining a pancreatic specimen can be difficult. Since the manifestations of the disease have a patchy distribution, such sampling may yield false negative results. Therefore, the role of a histopathological examination of the pancreas in patients with suspected autoimmune pancreatitis may be to exclude other diseases such as cancer rather than to provide definitive evidence for a diagnosis of autoimmune pancreatitis.⁹ However, given the error inherent in fine-needle aspiration of the pancreas, clinicians should remain vigilant for the possibility of cancer. Investigators at the Mayo Clinic have recently proposed the expansion of the diagnostic criteria for autoimmune pancreatitis to include a response to steroids and less typical findings on radiography supported by serologic or histologic evidence.³³

DIAGNOSTIC AND THERAPEUTIC APPROACHES

The diagnosis of autoimmune pancreatitis is most commonly considered when findings on imaging are highly suggestive of the disease (Fig. 4). In pa-

Table 1. Diagnostic Criteria for Autoimmune Pancreatitis.*

Findings on Imaging Radiography (One Required)		Serologic and Histologic Findings (One Required)		
Cross-Sectional Imaging	ERCP or MRCP	Serologic Analysis	Pancreatic–Biliary Histologic Analysis	Nongastrointestinal Histologic Analysis
Diffusely enlarged pancreas	Segmental pancreatic ductal narrowing	Elevated serum IgG4 level	Periductal lympho- plasmacytic infil- tration or fibrosis	Tubulointerstitial nephri- tis with immune de- posits within tubular basement membranes
Enhanced peripheral rim of hypoatten- uation “halo”	Focal pancreatic duc- tal narrowing	Elevated serum IgG or gamma globulin level	Obliterative phlebitis	Pulmonary interstitial lymphoplasmacytic infiltration with IgG4- positive plasma cells†
Low-attenuation mass in head of pancreas	Diffuse pancreatic ductal narrowing	Presence of ALA, ACA II, ASMA, or ANA	IgG4-positive plasma cells in tissue†	Chronic sialadenitis with IgG4-positive plasma cells†

* Criteria were modified from those of the Japan Pancreas Society.³⁰ ERCP denotes endoscopic retrograde cholangiopancreatography, MRCP magnetic resonance cholangiopancreatography, ALA antilactoferrin antibody, ACA II anti-carbonic anhydrase II antibody, ASMA anti-smooth-muscle antibody, and ANA antinuclear antibody.

† The presence of tissue IgG4-positive cells is not necessarily abnormal, but an increased number of infiltrating IgG4-positive plasma cells is abnormal.

tients whose findings on imaging are suggestive of autoimmune pancreatitis, alcohol-induced chronic pancreatitis or pancreatic cancer are the most common conditions that should be differentiated from autoimmune pancreatitis. However, when cross-sectional imaging shows diffuse gland enlargement and ERCP reveals a long attenuated segment of the pancreatic duct, autoimmune pancreatitis should be strongly considered.

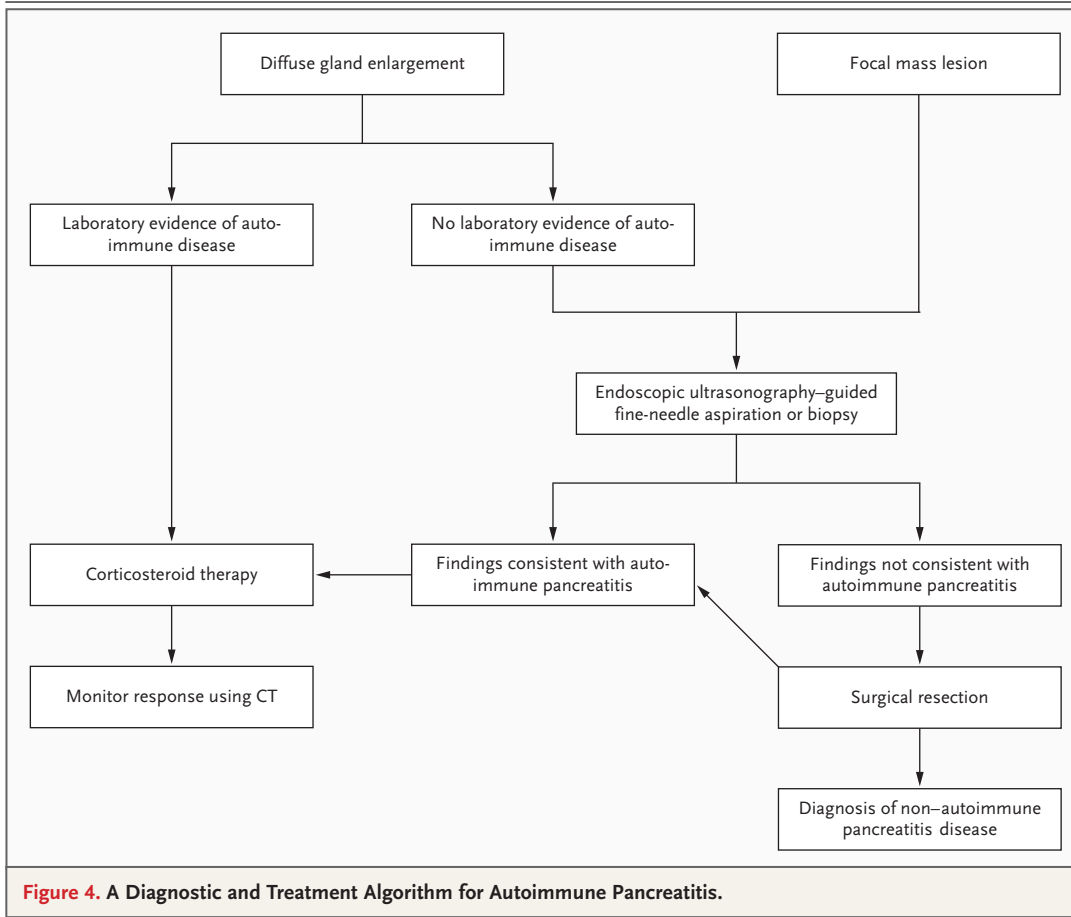
In the next stage of evaluation, patients should undergo laboratory testing. The finding of increased serum IgG levels or the presence of autoantibodies is supportive of the diagnosis, whereas an elevated serum IgG4 level is nearly diagnostic. The presence of diagnostic findings on imaging, coupled with supportive laboratory results, is sufficient to indicate a course of treatment with corticosteroids.

If the imaging studies reveal a discrete mass, the most important disease to consider in the care of these patients is pancreatic cancer. Autoimmune pancreatitis can produce a mass in the head of the pancreas that can mimic pancreatic adenocarcinoma. The diagnosis of autoimmune pancreatitis should be considered particularly in patients with laboratory findings suggestive of the disease and with a well-established history of autoimmune disorders. In these patients, an early fine-needle aspiration of the mass may be help-

ful, particularly in those for whom operative treatment is an option. Since pathological confirmation of autoimmune pancreatitis is difficult, corticosteroid therapy can be used as a diagnostic tool in patients whose clinical and laboratory findings are strongly suggestive of autoimmune pancreatitis. In these patients, short-interval imaging at 2 to 4 weeks after the initiation of therapy must be used to confirm whether there is resolution of the mass. Since malignant masses can also respond to corticosteroids, complete resolution of the pancreatic mass is critical for the diagnosis of autoimmune pancreatitis. If the clinical history and laboratory and pathological findings do not provide support for the diagnosis of autoimmune pancreatitis, or if corticosteroid therapy fails to resolve the pancreatic mass, surgical biopsy of the mass should be considered.

A tissue diagnosis of autoimmune pancreatitis may be made during surgical exploration of the pancreas or other involved organs, such as the liver, bile ducts, and gallbladder. If histologic findings of an autoimmune process are obtained, a diagnosis of autoimmune pancreatitis can be confirmed in the absence of diagnostic laboratory findings. Operative intervention will also rule out the possibility of pancreatic cancer.³⁴

Once a diagnosis of autoimmune pancreatitis has been made through a combination of imag-



ing, laboratory testing, and histologic analysis, a treatment course with corticosteroids should be considered. However, the use of corticosteroid therapy is not mandatory, since there have been reports of the spontaneous resolution of a pancreatic mass, stricture, and jaundice.^{35,36} The initial dose of corticosteroids (prednisone) should be 40 mg daily for 1 week, followed by a taper of the daily dose by 5 mg per week. The response to corticosteroids is often dramatic.³⁷⁻³⁹ During the first 2 to 4 weeks of therapy, CT should be used to monitor the response.^{40,41} Laboratory abnormalities (such as elevated IgG4 levels, hypergammaglobulinemia, and presence of autoantibodies) may also show improvement with corticosteroid therapy. Extrahepatic biliary obstruction has also proved to be responsive to corticosteroid therapy.^{40,42} A poor response to corticosteroids should raise the question of pancreatic cancer or other forms of chronic pancreatitis. Although the vast

majority of patients with autoimmune pancreatitis will readily respond to corticosteroids within a few weeks, a small subgroup may require maintenance therapy with prednisone at a dose of 5 to 10 mg per day.⁶ The role of immunosuppressive agents has not been defined.

In summary, autoimmune pancreatitis is an immune-based systemic disease that should be diagnosed on the basis of imaging, histologic, and serologic criteria. Its diagnosis is important, since autoimmune pancreatitis can mimic pancreatic cancer (as a pancreatic mass obstructing pancreatic–biliary ducts) and its lesions respond so readily to corticosteroids. More widespread use of pancreatic biopsy will aid in the diagnosis of autoimmune pancreatitis and provide a secure basis for the treatment with corticosteroids.

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