Imaging in Autoimmune Pancreatitis and Immunoglobulin G4–Related Disease of the Abdomen

Kumaresan Sandrasegaran, MD[a,*], Christine O. Menias, MD[b]

KEYWORDS

• Autoimmune pancreatitis • Systemic IgG4 disease • IgG4 sclerosing cholangitis
• Pancreas cancer • Cholangiocarcinoma • Primary sclerosing cholangitis

KEY POINTS

- Autoimmune pancreatitis (AIP) is classified into type I, which is associated with systemic immunoglobulin (Ig)G4 disease, and type 2, which is localized to the pancreas.
- AIP may present as diffuse, segmental, or focal disease of pancreas.
- Fecal AIP may mimic pancreas cancer, although in many cases imaging features help to differentiate the 2 entities.
- IgG4 sclerosing cholangitis usually presents with a tapering or funnel-shaped biliary ductal stricture, whereas cholangiocarcinoma typically presents with an abrupt stricture.

INTRODUCTION

Autoimmune pancreatitis (AIP) is a benign fibroinflammatory disease. This disease was initially reported in 1995[1] and its association with immunoglobulin G subclass 4 (IgG4)-related autoimmunity was recognized between 2001 and 2003 by Japanese researchers.[2,3] Since then, IgG4 disease has been reported to affect almost all organ systems, including the liver, biliary tree, kidneys, prostate gland, testicles, the peritoneum and the retroperitoneum, salivary and lacrimal glands, orbital tissues, pituitary gland, thyroid gland, lungs, lymph nodes, breasts, and vascular structures. In 2004, a new version of AIP (AIP type 2) without IgG4 disease, but with histologic features of a granulocytic epithelial lesion, was described.[4] Diagnosis of AIP requires the combination of clinical, serologic, imaging, and pathologic findings, as well as a good

Disclosure: The authors have nothing to disclose.

[a] Department of Radiology, Indiana University School of Medicine, 550 North University Boulevard, UH0279, Indianapolis, IN 46202, USA; [b] Department of Radiology, Mayo Clinic School of Medicine, Mayo Clinic Hospital, 5777 East Mayo Boulevard, Phoenix, AZ 85054, USA
* Corresponding author.
E-mail address: ksandras@iupui.edu

Gastroenterol Clin N Am ■ (2018) ■–■
https://doi.org/10.1016/j.gtc.2018.04.007
gastro.theclinics.com

response to corticosteroids. In this article, we highlight the imaging features of AIP and the spectrum of other abdominal IgG4 disease.

EPIDEMIOLOGY AND SUBTYPES

It is difficult to estimate the prevalence of AIP, because many cases are undiagnosed. Most reports of AIP come from Japan, where the incidence is estimated to be approximately 1 per 100,000 and AIP accounts for 5% to 6% of patients with chronic pancreatitis.[5] Studies in the United States have reported that 2% to 3% of pancreatic resections show autoimmune pancreatitis at pathologic analysis.[6]

AIP is now classified into types 1 and 2 (**Table 1**). Type 1 AIP is predominantly an IgG4-mediated disease and can affect virtually every organ system in the body (**Table 2**). Nevertheless, IgG4 may not be the only underlying cause of the disease. Type 1 AIP typically affects older, male patients. This type accounts for nearly all cases of AIP in Japan and Korea and 60% to 80% of cases in the United States.[7–9] Type 2 AIP is usually confined to the pancreas and there is an association with inflammatory bowel disease but no association with elevated IgG4 levels. Type 2 subtype is seen more often in Europe than in the United States.[6] The imaging findings of both type 1 and 2 are similar with minor differences, as discussed in this article.

CLINICAL PRESENTATION

The typical presentation of type 1 AIP is an elderly man with painless obstructive jaundice (up to 75% cases).[8] Thus, clinical differentiation from pancreas cancer may be difficult and the diagnosis is often made on imaging. Other clinical features include unexplained chronic abdominal pain, weight loss, and steatorrhea.[10] Exocrine functional abnormalities are seen in up to 80% of patients.[6] Type 2 diabetes mellitus may predate or present synchronously in up to 80% of patients with AIP. Surprisingly, in some patients with AIP, diabetes may improve with steroid therapy.[11] Type 2 AIP presents in younger patients, with roughly equal gender distribution. As with type 1 AIP, the most common presentation of type 2 AIP is painless jaundice. Presentation with acute pancreatitis is more commonly seen in patients with type 2 disease who have concurrent inflammatory bowel disease.[12]

Table 1
Differences between types 1 and 2 autoimmune pancreatitis (AIP)

	Type 1 AIP	Type 2 AIP
Synonyms	Lymphoplasmocytic sclerosing pancreatitis	Idiopathic duct-centric chronic pancreatitis
Mean age, y	Elderly: 64	Middle aged: 43
Male:female ratio	5:1	1:1
Incidence	Japan >> US, EU	EU > US > Japan
Obstructive jaundice, %	50	<5
Acute pancreatitis, %	15	33
Extrapancreatic disease	IgG4-systemic disease	Chronic ulcerative colitis in 30%
Serum IgG4	80% have elevated (>140 mg/dL)	Normal
Response to steroids	Good response (90%) but relapse ≈ 30%–40%	Good (90%) response and relapse <10%

Abbreviation: EU, European Union.
Data from Refs.[6,12,24,32]