Session 4. Pancreatic Cancer-Mimicking Autoimmune Pancreatitis

Autoimmune pancreatitis overview: Diagnosis, classification, and manifestations

Myung-hwan Kim

Division of Gastroenterology, Asan Medical Center,
University of Ulsan College of Medicine, Korea

Curriculum Vitae

- Professor, Division of Gastroenterology
  Asan Medical Center
  University of Ulsan College of Medicine
- Director, Center for Pancreatobiliary Disease, Asan Medical Center
- Past President, Korean Pancreatobiliary Association
- Past President, Korean Society of Gastrointestinal Endoscopy

Autoimmune pancreatitis overview: Diagnosis, classification, and manifestations

During the last decade, there has been growing recognition of a peculiar type of chronic pancreatitis known as autoimmune pancreatitis (AIP). AIP consists of two distinct clinical & histopathological forms of pancreatitis; type 1 and type 2. Unlike type 1 AIP, which is recognized as part of IgG4-related systemic disease, type 2 AIP may be an isolated pancreatic disorder. Type 2 AIP appears relatively common in the USA and Europe but rare in East Asia. Type 2 AUP responds well to steroid therapy as did type 1 AIP (table 1).

Table 1. Differences between clinicopathologic profiles of type 1 and type 2 AIP

	Type 1 AIP	Type 2 AIP
Synonym	Lymphoplasmacytic sclerosing pancreatitis (LPSP)	Idiopathic duct-centric chronic pancreatitis (IDCP)
Epidemiology	Asia > United States, Europe	Europe, United States > Asia
Clinical presentation	Obstructive jaundice (painless)	Obstructive jaundice/acute pancreatitis
Age at diagnosis	Old	Young
Serum IgG4 level	Elevated	Normal
Histological hallmarks	Periductal lymphoplasmacytic infiltrate, storiform fibrosis, obliterative phlebitis	Granulocytic epithelial lesion (GEL)
Tissue IgG4 stain	Many IgG4 (+) cells	nNone or very few IgG4 (+) cells
Other organ involvement	Bile duct, salivary gland, kidney, retropertineum	Not seen
Ulcerative colitis	rare	20-30%
Steroid responsiveness	Excellent	Excellent
Recurrence	Common	Rare

Type 1 AIP is the pancreatic manifestation of a systemic disease characterized by a steroid-responsive fibroinflammatory process involving multiple organs. The term IgG4-related disease (IgG4-RD) has been used to describe this multisystem disorder because it is characterized by a peculiar elevation in serum levels of the IgG4 subclass of immunoglobulin G (IgG) and tissue infiltration with IgG4-positive plasma cells. Patients with type 1 AIP often have other organs involvement (OOI) as part of IgG4-RD. Many organs involved in IgG4-RD share common histologic features of abundant infiltration with lymphocytes and plasma cells accompanied by storiform fibrosis. The infiltrate has a unique predilection to involve

veins, leading to their obstruction (obliterative phlebitis).

Type 1 AIP is a disease of elderly people. Its most common acute presentation is obstructive jaundice. The demographics and presentation usually raise suspicion of pancreatic/biliary cancer. Characteristic imaging includes a diffusely enlarged gland with featureless borders (sausage-shaped) and capsule-like low-density rim of the pancreas. The pancreatic duct is characteristically not dilated at this stage and shows a diffusely narrowed duct with irregular wall. Cases of AIP with segmental/focal pancreatic enlargement or mass-forming AIP are difficult to differentiate from pancreatic cancer.

The diagnosis of AIP in this setting requires a high index of suspicion and familiarity with the clinical manifestations of the disease. The diagnosis is suspected in patients with obstructive jaundice who have elevated serum IgG4 levels, OOI (e.g., proximal bile duct stricture, retroperitoneal fibrosis, renal mass, salivary/lacrimal gland enlargement/mass) or if core biopsies of the pancreas show typical histology. The response in patients with AIP to even short duration (2 weeks) of steroid therapy (prednisolone 30-40mg/day) is dramatic and diagnostic.

AIP is much less common than pancreatobiliary malignancies. Negative work-up for cancer including EUS-FNA is a prerequisite for a steroid trial. If the steroid responsiveness is equivocal or negative, a definitive diagnosis should be pursued by surgical exploration or resection.

Our understanding of AIP and IgG4-RD continues to evolve.