

## **AUTOIMMUNE PANCREATITIS**

### **Inflammatory Pancreatic Disease**

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Until 1995, when autoimmune pancreatitis was first described, pancreatitis was considered a disease induced exclusively by alcoholism.

### **The Pancreas and its Function**

The pancreas is an endocrine gland situated in the abdominal cavity that helps regulate blood glucose levels and assists with the digestion of fatty lipids. Inflammation of the pancreas, which is known as pancreatitis, has traditionally been associated with alcohol abuse. However, in 1995 researchers first described a form of chronic pancreatitis associated with autoimmune manifestations.

### **Autoimmune Origins**

Today it's known that about 5-6 percent of all cases of chronic pancreatitis are autoimmune in nature. And according to recent reports, the incidence of autoimmune pancreatitis appears to be rising, particularly in Japan, Europe, the United States, and Korea.

Autoimmune pancreatitis is generally considered a benign disorder because patients typically respond favorably to treatment with corticosteroids. Autoimmune pancreatitis is also known by other names including lymphoplasmacytic sclerosing pancreatitis with cholangitis, idiopathic duct destructive pancreatitis, primary inflammatory pancreatitis, non-alcoholic duct destructive chronic pancreatitis, pseudotumorous pancreatitis, tumefactive pancreatitis, and destructive pancreatitis depending on the specific tissue changes found on biopsy or the predominant and accompanying symptoms.

### **Who Is Affected?**

Autoimmune pancreatitis occurs in twice as many men as women. The initial presentation usually occurs between ages 50-60, but patients can also develop autoimmune pancreatitis as early as age 30 as well as late in life.

Autoimmune pancreatitis can occur as alone or in association with other autoimmune disorders including sclerosing cholangitis, primary biliary cirrhosis, inflammatory bowel disease, rheumatoid arthritis, hypothyroidism, sarcoidosis, and Sjogren's syndrome. In addition, autoimmune pancreatitis has been seen in association with retroperitoneal fibrosis and lung nodules.

Because certain markers of autoimmune pancreatitis, such as IgG4 positive plasma cells can be detected in other tissues besides the pancreas in affected patients, some researchers believe that autoimmune pancreatitis may be a systemic autoimmune disease affecting multiple organs besides the pancreas, including the gallbladder, bile ducts, salivary glands, lungs, biliary tree, and the kidney's renal tubules.

### **Signs and Symptoms**

Signs of autoimmune pancreatitis include elevated levels of gamma globulins and marked elevations of alkaline phosphatase with only slight elevations of transaminase enzymes, elevated serum IgG4 levels, and autoantibodies directed against carbonic anhydrase and lactoferrin. The blood sugar may also be elevated and the stools may have increased fat content. The pancreas is often enlarged and is surrounded by a halo of lymphocytes and plasma cells. Granulomas may also be present around the ducts of the pancreas, and a mass may obstruct the ducts.

Autoimmune pancreatitis can cause a wide variety of symptoms that tend to occur as a relapsing-remitting type of disease, with periods of symptoms alternating with periods of remission. Common symptoms include jaundice, weight loss, and mild abdominal pain. Severe abdominal pain or other symptoms of acute pancreatitis are unusual.

### **Diagnosis**

Blood tests for carbonic anhydrase and lactoferrin antibodies are positive in autoimmune pancreatitis. Imaging tests show an enlarged pancreas with white blood cell infiltration and fibrosis. Cross-sectional imaging shows diffuse gland enlargement and a long attenuated segment of the pancreatic duct. A favorable response to corticosteroids also differentiates autoimmune pancreatitis from alcohol-induced pancreatitis. Because a mass obstructing the biliary ducts is often seen in autoimmune pancreatitis, autoimmune pancreatitis must be differentiated from pancreatic cancer with fine needle aspiration biopsy and tissue studies of the pancreas.

### **Resources:**

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