Autoimmune pancreatitis

Overview

Autoimmune pancreatitis (AIP) is a chronic inflammation that is thought to be caused by the body's immune system attacking the pancreas and that responds to steroid therapy. Two subtypes of AIP are now recognized, type 1 and type 2.

Type 1 AIP is the pancreatic manifestation of a disease called IgG4-related disease (IgG4-RD). This disease often affects multiple organs including the pancreas, bile ducts in the liver, salivary glands, kidneys and lymph nodes.

Type 2 AIP seems to affect only the pancreas, although about one-third of people with type 2 AIP have associated inflammatory bowel disease.

Type 1 AIP can be mistakenly diagnosed as pancreatic cancer. The two conditions have overlapping signs and symptoms, but very different treatments, so it is very important to distinguish one from the other.

Symptoms

Autoimmune pancreatitis (AIP) is difficult to diagnose. Often, it doesn't cause any symptoms. Symptoms and signs of type 1 AIP are similar to those of pancreatic cancer.

Pancreatic cancer signs and symptoms can include:

- Dark urine
- Pale stools or stools that float in the toilet
- Yellow skin and eyes (jaundice)
- Pain in your upper abdomen or middle part of your back
- Nausea and vomiting
- Weakness or extreme tiredness
- Loss of appetite or feelings of fullness
- Weight loss for no known reason

The most common sign of type 1 AIP, present in about 80% of people, is painless jaundice, caused by blocked bile ducts. Type 2 AIP can present with recurrent episodes of acute pancreatitis. Pain in the upper abdomen, a common symptom of pancreatic cancer, is frequently absent in autoimmune pancreatitis.

Differences between type 1 and type 2 AIP are:

- In type 1 AIP, the disease may affect other organs in addition to the pancreas. Type 2 AIP affects only the pancreas, although the disease is associated with another autoimmune condition, inflammatory bowel disease.
- Type 1 AIP predominantly affects men in the sixth to seventh decade of life.
- Type 2 AIP affects both men and women equally and has a younger age of onset compared with type 1 AIP.
- Type 1 AIP is more likely to relapse after treatment is discontinued.

When to see a doctor

Autoimmune pancreatitis often doesn't cause any symptoms. See your doctor, however, if you experience unexplained weight loss, abdominal pain, jaundice, or other signs and symptoms that bother you.

Causes

Doctors don't know what causes autoimmune pancreatitis, but as in other autoimmune diseases, it is thought to be caused by the body's immune system attacking healthy body tissue.

Risk factors

The two types of AIP occur with different frequency in different parts of the world. In the United States, about 80 percent of people with autoimmune pancreatitis have type 1.

People with type 1 autoimmune pancreatitis often:

- Are over age 60
- Are male

People with type 2 autoimmune pancreatitis:

- Are often over age 40 (one or two decades younger than those with type 1)
- Are as likely to be female as male
- Are more likely to have inflammatory bowel disease, such as ulcerative colitis

Complications

Autoimmune pancreatitis can cause a variety of complications.

- **Pancreatic exocrine insufficiency.** AIP may affect the ability of your pancreas to make enough enzymes. Signs and symptoms may include diarrhea, weight loss, metabolic bone disease, and vitamin or mineral deficiency.
- **Diabetes.** Because the pancreas is the organ that produces insulin, damage to it may cause diabetes and you may need treatment with oral medication or insulin.
- **Pancreatic and bile duct stricture.**
- **Pancreatic calcifications or stones.**

Treatments for autoimmune pancreatitis, such as long-term steroid use, also can cause complications. However, even with these complications, people who are treated for autoimmune pancreatitis have a normal life expectancy.

There is no established association between AIP and pancreatic cancer.

By Mayo Clinic Staff