Autoimmune pancreatitis: when we suspect it?


Learning objectives

Review the radiologic findings in US, CT and MR in patients with autoimmune pancreatitis.

Background

Autoimmune pancreatitis is a rare form of chronic pancreatitis, with multiple synonyms (lymphoplasmacytic sclerosing pancreatitis, sclerosing pancreatitis, sclerosing pancreatic cholangitis); characterized by abundant lymphoplasmacytic infiltrations and fibrosis in the histopathological study.

Autoimmune pancreatitis accounts for 5-11% of all cases of chronic pancreatitis. Age of presentation is variable, and there appears to be a male predilection. The clinical presentation is variable, and includes jaundice, weight loss and abdominal pain. A large portion of patients also have diabetes mellitus.

In most cases, is associated with other autoimmune disorders: sclerosing cholangitis and primary biliary cirrhosis, Sjögren syndrome, ulcerative colitis, thyroiditis and retroperitoneal fibrosis.

There are three patterns of autoimmune pancreatitis: diffuse, focal and multifocal. The focal pattern is the most important to recognize because it can simulate pancreatic carcinoma.

Findings and procedure details

We describe the radiological findings of 3 patients diagnosed of autoimmune pancreatitis in our hospital. There were two men and one woman, age range between 5 and 58 years.

US:

In patients with autoimmune pancreatitis the affected area of the pancreas
appears hypoechoic (Fig 1).

CT:

Enlargement of the pancreas with loss of lobular architecture, and hypoattenuating parenchyma. Sometimes enlargement is focal, and differentiation from pancreatic carcinoma can be difficult. At dynamic contrast-enhanced CT, progressive enhancement of the focal mass due to autoimmune pancreatitis can help in differentiation from pancreatic adenocarcinoma.

Peripancreatic rim of low attenuation, forming the typical image of “sausage pancreas”, is a characteristic finding of autoimmune pancreatitis (Fig 2).

MR:

MR images shows diffuse enlargement of pancreatic gland, with hypointense at T1-weighted MRI and hyperintense at T2-weighted MRI (Fig 3 and fig 4).

At post-contrast images, affected pancreas shows delayed enhancement on dynamic imaging. A peripheral rim of hypoenhancement is also seen.

Other important feature of autoimmune pancreatitis is focal, segmental or diffuse narrowing of the pancreatic duct in the MR cholangiopancreatography (Fig 5).

Autoimmune pancreatitis has a partial or complete response to treatment with corticosteroids. After steroids therapy, pancreatic function and morphologic characteristics usually returns to normal within 4-6 weeks (Image D in figure 4).
**Fig. 1**: US appearance of autoimmune pancreatitis in 58 years old male. The image shows a diffuse hypoechoic area in the body and tail.
Fig. 2: Patient of figure 1. Contrast-enhanced CT (Fig 2a) reveals diffuse enlargement of body and tail of pancreas. Image 2b shows the tipical form of “sausage pancreas” It consists in a peripheral rim of hypoattenuation of the pancreatic gland (white arrow).
Fig. 3: Patient of figure 1 and 2. Fat-suppressed T2-weighted MR image (Fig 3a) shows diffuse enlargement of the pancreas. Postcontrast arterial phase fat-suppressed T1-weighted MR image (Fig 3b) shows heterogeneous enhancement. Note the hypointense peripheral ring (white arrow), a characteristic finding of autoimmune pancreatitis. MR cholangiopancreatography (Fig 3c) shows stenosis of the intrapancreatic segment of the common bile duct (white arrow), with dilatation of the upstream bile ducts.
Fig. 4: Autoimmune pancreatitis in a 5 years old woman with abdominal pain, jaundice and hepatomegaly. Fat-suppressed T1-weighted MR images (Fig 4 a) shows increased size of the pancreatic gland. Short tau inversion recovery (STIR) MR image (Fig 4b) shows increased pancreas and the absence of intra-abdominal fluid. MR cholangiopancreatography (Fig 4c) shows diffuse dilatation of the bile duct. After 20 days of treatment with steroids (Fig 4d), symptoms and imaging tests improved significantly (reducing the size of the pancreatic gland).
Fig. 5: 52 years-old man with autoimmune pancreatitis. MR cholangiopancreatography shows the tipical stenosis of the main pancreatic duct, a frequent finding in autoimmune pancreatitis.
Conclusion

Autoimmune pancreatitis is an infrequent type of chronic pancreatitis. Recognition of this entity and its characteristics is important because it allows for the differential diagnosis with other entities and let’s start treatment at an early stage.

References