Autoimmune pancreatitis (AIP) is a distinct form of chronic pancreatitis. Its incidence ranges between 4% and 6% of chronic pancreatitis. AIP affects a wide age range of subjects (40-60 years), typically males (60%) without a history of alcohol abuse, biliary stone disease or duodenal wall inflammation. Histologically, AIP is characterized by a dense lymphoplasmacytic infiltrate of mainly CD4+ T lymphocytes and immunoglobulin G4 plasma cells, located around the pancreatic ducts with mass-forming regions of fibrosis.

Other organs, such as the biliary tract may also be involved. In some cases other organs such as salivary glands, stomach, duodenum, kidney and retroperitoneum may be involved. For this reason some authors postulated that AIP is one aspect of a systemic IgG4-mediated inflammatory process with a predominantly pancreatic involvement. Furthermore, the autoimmune mechanism is supported by the remission of signs and symptoms after a short-term steroid treatment.

The dense lymphoplasmacytic infiltrate is responsible for the increase in size of the organ and the loss of its lobular structure; depending on the extent of the parenchyma affected, AIP has been classified morphologically into focal or diffuse forms. The differential diagnosis between focal AIP and pancreatic adenocarcinoma represents a medical need, since AIP responds to steroid therapy, and surgery should be avoided.

Magnetic resonance imaging (MRI) findings of AIP are characterized by an enlargement of the gland, either focal or diffuse, with hypointense signal on T1-weighted images within the lesion, because the areas of fibrosis in AIP. In the early stage of AIP the lesion shows mild hyperintensity on T2-weighted

images due to the periductal inflammation. The lesion appears hypovascular during the arterial phase, with progressive enhancement and delayed retention of contrast in the venous and late phases.

MR cholangiopancreatography (MRCP) is able to assess the pancreatic duct system and the demonstration of its involvement has an increasing importance in the diagnosis of AIP. These features are important in the differential diagnosis with the pancreatic adenocarcinoma in which the main pancreatic duct is characterized by single short stenosis, with marked dilation of the upstream ductal system. The bile ducts can also be involved. In cases in which AIP involves the head of the pancreas, dilatation of the common bile ducts and the intrahepatic ducts can be seen. In other cases multiple stenoses of the common bile duct and intrahepatic bile ducts due to cholangitis can be documented.

MRI is also helpful in monitoring the disease following corticosteroid therapy. A prompt response to the corticosteroid treatment is seen after two to three weeks of therapy with a reduction of the pancreatic enlargement and a gradual restitution of the normal signal intensity of the gland.