Focal autoimmune pancreatitis

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A 70-year-old man was referred for evaluation of mild epigastric discomfort with tiredness. He had no particular medical history and admitted drinking two glasses of wine a day. Biology showed a small increase in CRP and pancreatic enzymes (lipases and amylases).

Ultrasound examination was negative for gallstone or biliary dilatation. Abdominal contrast-enhanced CT showed a sharply delineated enhancement defect at arterial time of the caudal region of the pancreas (Fig. A1) and a relative swelling of the region with some peri-pancreatic fat blurring at portal time (Fig. A2). MRI showed edema of the caudal region of pancreas, appearing hyper-intense on T2-weighted imaging and diffusion (Fig. B1), hypo-intense on T1-weighted imaging with disappearance of interlobular septas (Fig. B2), and a mild enhancement after intravenous injection of gadolinium (Fig. B3). Diagnosis of auto-immune chronic pancreatitis was suggested.

Increase in IgG4 and negative tumor markers (CEA and CA 19.9) consolidated the hypothesis of an auto-immune origin. A FNA by echo-endoscopy confirmed the radiological suspicion.

Comment

Auto-immune chronic pancreatitis is a rare variant of chronic pancreatitis (representing less than 5% of cases) that can be misdiagnosed as a pancreas neoplasms. Imaging, especially MRI, and laboratory findings are helpful to establish the diagnosis and avoid an inappropriate surgical resection.

This pathology is characterized by a minor symptomatology, absence of stone, calcification, pseudo-cyst, adenopathy, vascular lesion and is also by a ductal abnormality: most often, long irregular stenosis of the principal pancreatic duct with mild or absence of pancreatic ductal dilatation is seen. There are two variants of the disease: diffuse or pseudo-tumoral (1).

On contrast-enhanced CT, it appears as a focal or diffuse swelling of pancreas with loss of normal lobulated shape, enhancement defect at arterial time and enhancement at late time. MRI demonstrates an edema appearing hyper-intense on T2-weighted imaging with enhancement after intravenous injection of gadolinium. MRI is the best technique to appreciate ductal abnormalities.

Auto-immune chronic pancreatitis is inconstantly associated with others auto-immune diseases like diabetes, cholangitis, inflammatory bowel diseases or Sjögren disease. Treatment consists in corticotherapy leading to regression of lesions and symptoms in a few days or weeks, but some cases of recurrence have been described.

Reference