

# IgG4 related autoimmune pancreatitis and sclerosing cholangitis

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Dear Editor,

IgG4-related disease is an increasingly recognized disorder that is characterized by an IgG4-positive plasma cell infiltration of tissue. The affected tissues also have a variable degree of fibrosis (1,2).

A 55-year-old female was admitted to the emergency unit with increasing nausea, vomiting, and abdominal pain since two days prior to presentation. Physical examination showed abdominal tenderness on the right upper quadrant and guarding in the epigastric region. She had decreased blood pressure of 95/50 mmHg, normal blood oxygen level (95%; normal range, 90%-100%), and increased heart rate (114 beats/minute; normal range, 80-100 beats/minute). No features were found in her medical history. Laboratory investigations revealed an elevated white blood count (14.8 K/uL; normal range, 4-10.8 K/uL) and low hemoglobin level (11.9 g/dL; normal range, 14-17 g/dL). Elevated liver enzymes were observed as follows: gamma-glutamyl transferase (GGT; 156 U/L; normal range, 10-71 U/L), alanine transaminase (55 IU/L; normal range, 4-40 IU/L), and aspartate transaminase (90 IU/L; normal range, 4-35 IU/L). Total bilirubin (3.7 mg/dL; normal range, <1.3 mg/dL) and C-reactive protein levels (1.04 U/mL; normal range, 0-0.6 mg/dL) were elevated. Notably, other laboratory results were within the normal limits.

Abdominal ultrasonography was obtained, and it showed dilatation of the intrahepatic and extrahepatic biliary ducts. Abdominal magnetic resonance imaging (MRI) with intravenous contrast medium and MR cholangiopancreatography (MRCP) were obtained. MRI showed diffuse swelling of the pancreatic corpus and tail, subtle edematous rim of pancreatic parenchyma, consistent with autoimmune pancreatitis (Figure 1). MRCP showed biliary

duct beading, including the areas of strictures and focal aneurysmal dilatations of the common bile duct (CBD), consistent with sclerosing cholangitis (SC; Figure 2). An endoscopic retrograde CP (ERCP) was performed, and it showed intrahepatic biliary strictures and CBD strictures, similar with MRCP. Brushing and biopsies were taken, and sphincterotomy was performed. The biopsies were negative for malignancy. Immunostaining of IgG4 for biopsies showed positive cells with indeterminate clinical significance. She had elevated blood IgG4 level of 322 mg/dl (normal range, 8-130 mg/dL). Based on the clinical, histopathological, and imaging findings, a diagnosis of autoimmune pancreatitis (AIP) and IgG4-related SC was done. The patient was initiated on 40 mg/day of prednisone that was continued for one month. The patient was also initiated on 1 gr/day of paracetamol and intravenous (IV) fluid hydration. Within the first week of the treatment initiation, the patient showed clinical improvement, and her laboratory results were normalized. Control MRCP was obtained on the 27<sup>th</sup> day of the treatment, and it showed improvement in the biliary duct beading and swelling of the pancreatic corpus. Her corticosteroid treatment was slowly tapered gradually and stopped. No recurrence was seen at the 6-month follow-up.

Autoimmune pancreatitis is a rare form of chronic pancreatitis. It is characterized by parenchymal lymphocyte infiltration and fibrosis, and it may be associated with various additional diseases and conditions such as SC and high serum IgG4 level. In cases with IgG4-related AIP and SC, intensive tissue infiltration of IgG4-positive plasma cells and increased fibrosis are common. The pancreas, bile ducts, and gallbladders are frequently affected, as in our case (1). IgG4-related SC should be distinguished from cholangiocellular carcinoma and primary SC (PSC) (2). Segmental strictures and aneurysmal dilatation of the lower CBDs were found to be more common in IgG4-related SC

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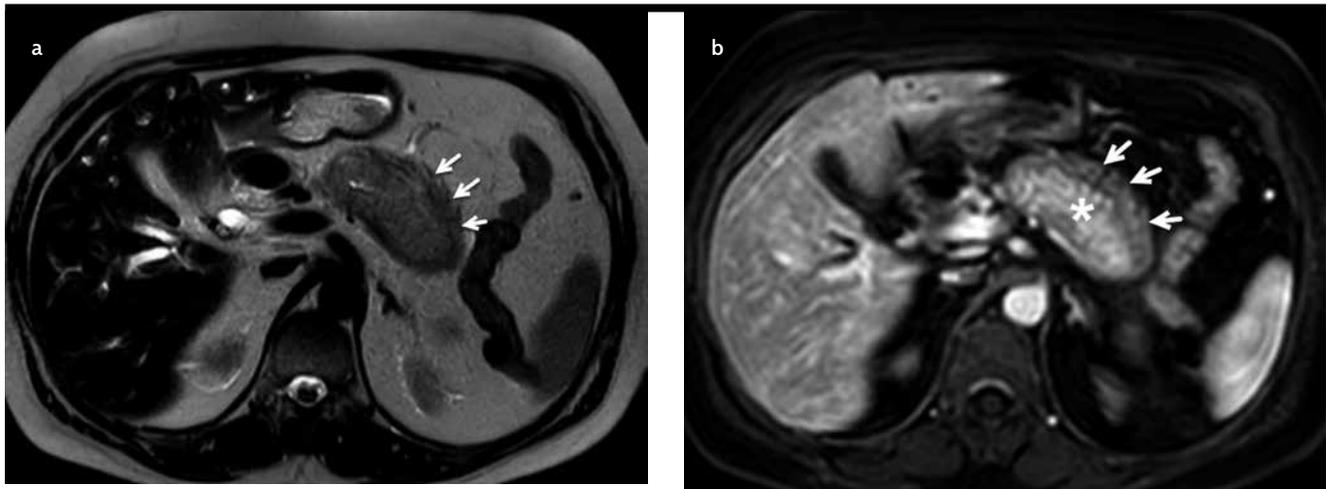


Figure 1. a, b. MRI showed diffuse swelling of the pancreatic corpus and tail

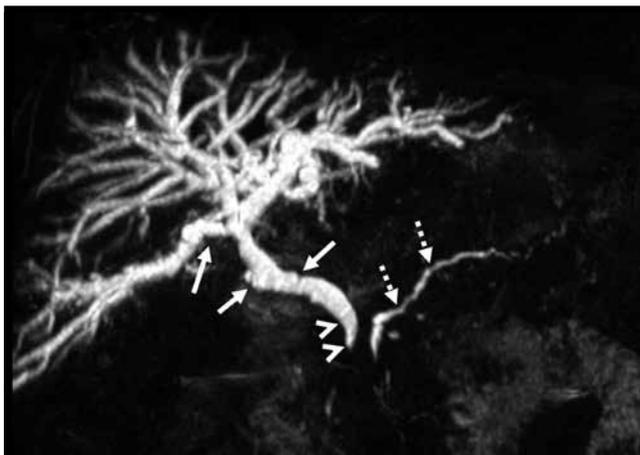


Figure 2. MRCP showed biliary duct beading

than PSC (3). Similarly, in our case, we observed segmental strictures and aneurysmal dilatation of the lower CBD.

The decisive MRI findings of IgG4-related SC and AIP are smooth distal CBD stricture with proximal intrahepatic biliary dilatation, strictures and aneurysmal dilatation of the lower CBD, and diffuse pancreatic enlargement with a hypoenhancing halo, as in our case (4). In conclusion, our case clearly shows that the MRI findings of IgG4-related SC and AIP and the awareness of MRI findings will lead to prompt diagnosis and early treatment.

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