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LETTER TO THE EDITOR

Immunoglobulin G4-related Autoimmune Pancreatitis in a Patient Misdiagnosed with Abdominal Pain and Obstructive Jaundice

A 60-year-old male patient who was a heavy drinker presented with yellow discoloration of his skin for 1 week, dull epigastric pain, and a 3-kg weight loss over a period of 1 month. Physical examination showed a yellowish skin, icteric sclera, and mild epigastric tenderness. Laboratory results showed abnormal liver function tests (aspartate aminotransferase 164 U/L, alanine aminotransferase 292 U/L), hyperbilirubinemia (total bilirubin 3.95 mg/dL, direct bilirubin 2.53 mg/dL), and increased Carbohydrate Antigen 19-9 (CA 19-9) (44.34 U/mL). Figure 1A shows an abdominal magnetic resonance imaging scan. After the diagnosis of a pancreatic tumor with obstructive jaundice, the patient underwent a cholecystojejunostomy bypass operation and a biopsy sample was taken of the pancreatic head tumor. Under microscopic examination, the specimen showed chronic inflammation with marked fibrosis, but no cancer cells.

The patient was seen 1 year later with acute renal failure. Retrograde pyelography showed bilateral hydronephrosis. He therefore underwent a percutaneous nephrostomy (Figure 1B). He was treated with steroids (prednisolone 25 mg/day by mouth) as a result of marked retroperitoneal fibrosis. Figure 1C shows cholangiopancreatography on a magnetic resonance imaging scan. The histopathological findings showed a positive immunohistochemical stain of immunoglobulin G4 (IgG4) in the pancreatic cells (Figure 1D). In addition, his serum IgG4 level was increased (626 mg/dL, normal reference range 3–200 mg/dL) and the patients was therefore diagnosed with IgG4-related autoimmune pancreatitis (AIP).

AIP is a rare disease first reported by Yashida et al[1] in 1995. A patient with AIP may have symptoms and signs of abdominal pain, weight loss, and obstructive jaundice. The findings on imaging usually show enlargement of the pancreas and a tumor-like mass.[2] It is clinically important to distinguish between pancreatic cancer and AIP. Our patient underwent surgery after the diagnosis of a pancreatic tumor with superior mesenteric vein encasement. He was also found to have retroperitoneal fibrosis, hydronephrosis, and biliary tract stenosis. He was treated long term with steroids at an average dose of 25 mg/day of prednisolone by mouth and his clinical status improved dramatically.

To avoid the misdiagnosis of AIP as pancreatic cancer, the Japanese consensus[1] suggests three major pathognomonic points: (1) enlargement of the pancreas and narrowing of the main pancreatic duct; (2) high serum IgG or IgG4; and (3) histological findings of lymphoplasmacytic infiltration and fibrosis in the pancreas.[1]

Clinicians were misled in the diagnosis of pancreatic cancer or chronic pancreatitis in this alcoholic patient with an enlarged pancreatic head. The patient should have been diagnosed with IgG4-related AIP because of the early encasement of the superior mesenteric vein and later marked retroperitoneal fibrosis, increased serum IgG4, lymphoplasmacytic infiltration, the positive immunohistochemical stain of IgG4 of the pancreas (Figure 1D), and his good response to treatment with steroids.[3]

In summary, this case report serves as a reminder to clinicians to keep the possibility of IgG4-related AIP in mind during the differential diagnostic work-up of a patient with an enlarged pancreatic head and jaundice.

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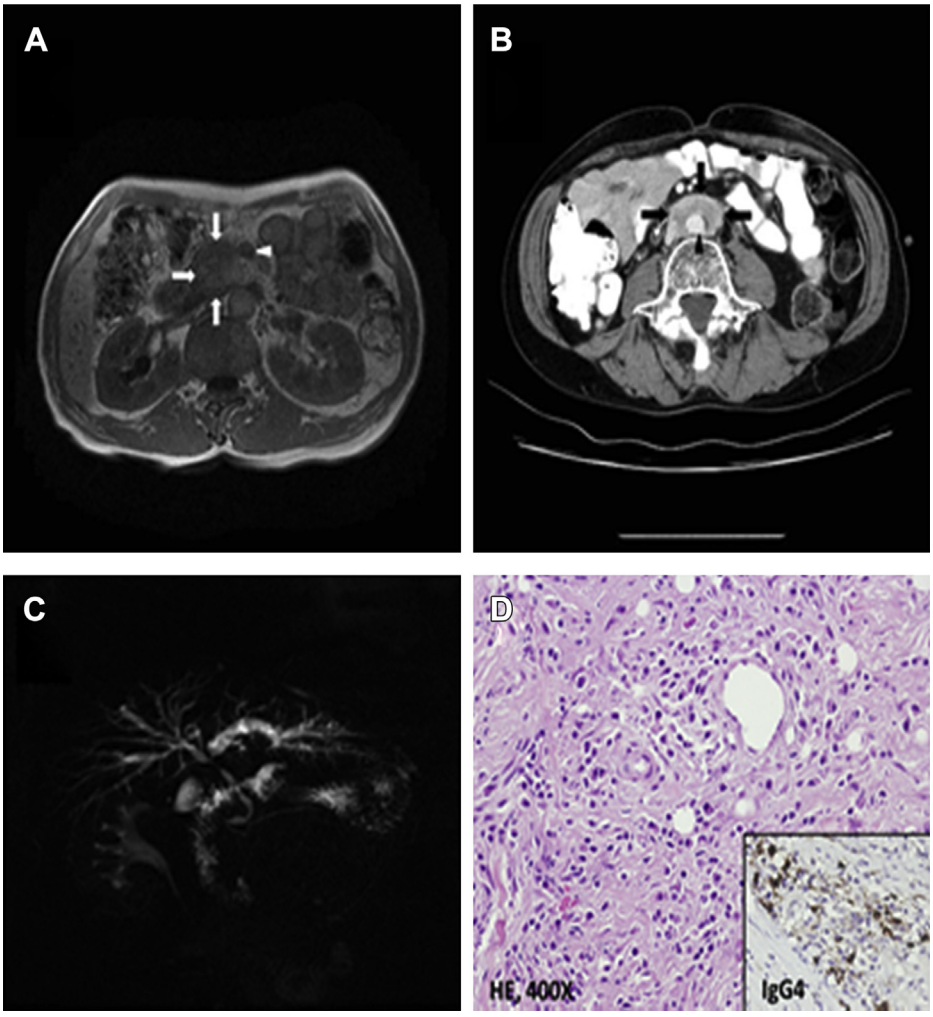

Figure 1 (A) Computed tomography T1-weighted image showing a 3.7 cm pancreatic head tumor (three arrows) with mild dilatation of the main pancreatic duct, partial superior mesenteric vein encasement (arrowhead), and mild right hydronephrosis. (B) Computed tomography scan 1 year after surgery showing retroperitoneal fibrosis (3 black arrows) and encasement of the abdominal aorta (arrowhead), common iliac arteries, and bilateral distal ureters. (C) Magnetic resonance image of cholangiopancreatography showing a narrow common hepatic duct and dilated bilateral intra-hepatic ducts after the patient had discontinued treatment with steroids after he was found not to have a definite pancreatic tumor. (D) Microscopic image (400×) of the hematoxylin and eosin stain showing increased serum immunoglobulin G4, lymphoplasmacytic infiltration, and a positive immunohistochemical stain of immunoglobulin G4 of the pancreas in the right lower corner.