VIDEO CASE REPORT

Autoimmune pancreatitis masquerading as pancreatic cancer

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CASE PRESENTATION

A 77-year-old man presented with a history of painless jaundice, loss of weight, and anorexia for 3 weeks. He had a significant history of atrial fibrillation on warfarin. His physical examination revealed icteric sclera. Murphy's sign was negative. Initial laboratory markers showed increased bilirubin (169 umol/L) with elevated Ca 19-9 (144 u/mL).

Abdominal ultrasound revealed an ill-defined heterogeneous mass in the distal end of the common bile duct causing intrahepatic and extrahepatic biliary ductal dilatation. MRCP showed moderate dilatation of the biliary system down to the distal common bile duct with an abrupt change in the caliber, which is consistent with a stricture (Fig. 1). There was also a stricture in the main pancreatic duct in the region of the pancreatic neck and body (Fig. 2) without any upstream dilatation. Further evaluation revealed a high serum immunoglobulin subclass 4 (IgG4) level above 3.40 g/L (normal value: 0.04-1.57 g/L).

EVALUATION AND PROGRESS

EUS was done to exclude pancreatic malignancy. It showed diffuse enlargement of the pancreas with heterogeneous, predominantly hypoechoic pancreatic parenchyma, which assumed a sausage-shaped appearance (Fig. 3). The intrahepatic bile duct was dilated. There was a hypoechoic rim around the pancreatic parenchyma, which is consistent with a pseudo capsule.[1] The main pancreatic duct was tortuous and irregular but still within normal caliber. A mass-like lesion was seen in the distal pancreas.

When observed closely, the main pancreatic duct was visible within the mass. This is called the duct penetrating duct sign (Fig. 4) and is suggestive of autoimmune pancreatitis (AIP).[2] The head of the pancreas was relatively unremarkable. Fine-needle biopsy was preferred over FNA[3,4] because of its favorable detection rates for

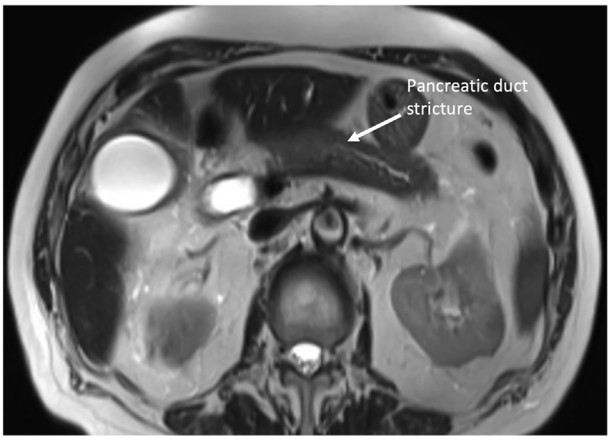

Figure 2. Pancreatic duct stricture at the level of the body.

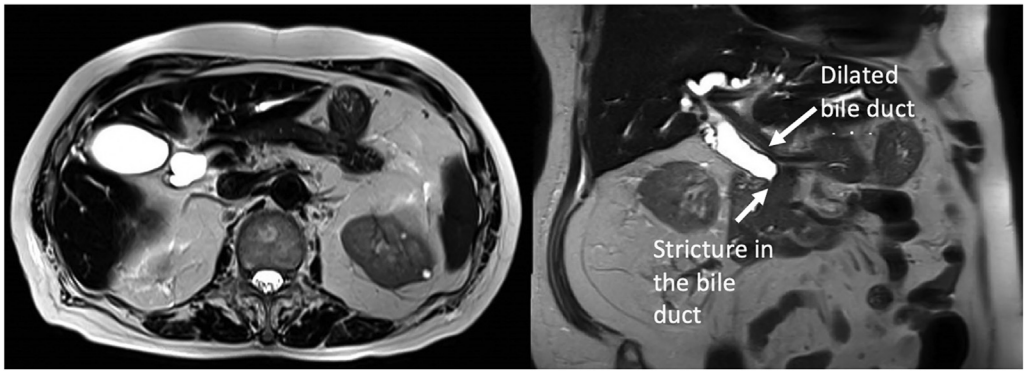

Figure 1. Distal bile duct stricture with upstream dilatation.

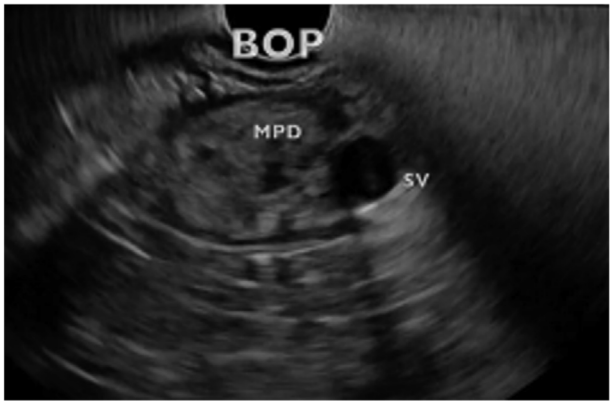

Figure 3. Pseudo capsule of the pancreatic parenchyma.

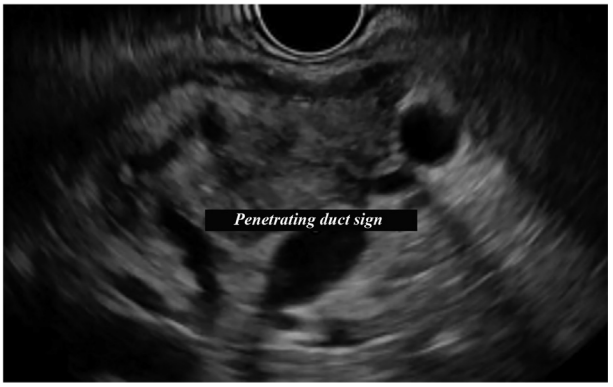

Figure 4. Penetrating duct sign.

AIP. This was done using a 20-gauge Procore histology needle (Cook-Medical, Winston-Salem, NC, USA). Histology showed IgG4-positive plasma cells (Fig. 5). Together with the raised IgG4 levels, the presence of IgG4-positive plasma cells confirmed the diagnosis of AIP.

The patient was started on oral prednisolone 40 mg once a day for a month with subsequent tapering. Liver function improved (bilirubin 30 umol/L), and the interval MRCP scan showed resolution of the pancreatic swelling. ERCP was not indicated because the patient did not have cholangitis and the diagnosis was in keeping with AIP.

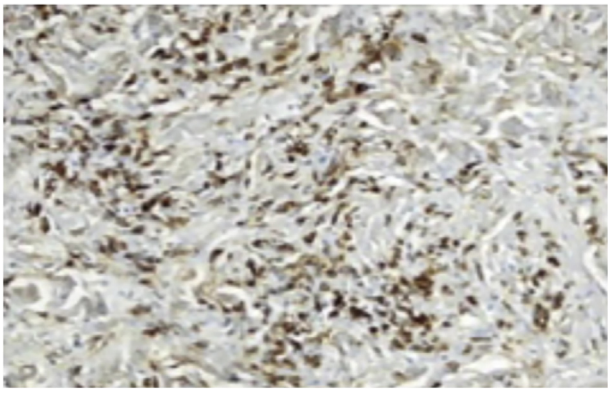

Figure 5. Immunoglobulin subclass 4–rich plasma cells.

DISCUSSION

This patient's clinical presentation, serology, and radiological and histological features fulfill the diagnostic criteria for type I AIP based on the international consensus diagnostic criteria for AIP (2011).[5] Following the international consensus for the treatment of AIP (2017),[6] corticosteroid therapy (prednisolone 40 mg/day) was started because the patient had both pancreatic and bile duct involvement. Prednisolone was tapered by 5 mg every week until a daily dosage of 20 mg was reached, followed by tapering 5 mg every 2 weeks. Repeat imaging 4 weeks after the diagnosis showed the swelling in the pancreas had resolved.

Biochemically, the patient's jaundice was resolved. As per the international consensus for the treatment of AIP, this patient had predictors of relapse that included high IgG4 levels (>4 times the upper limit of normal) and diffuse enlargement of the pancreas. Hence, he was kept on a low, maintenance dose of prednisolone 5 mg/day to prevent relapse.

We have presented a case of painless obstructive jaundice with imaging initially detecting a pancreatic mass; however, with careful examination using EUS and fine-needle biopsy, it was found to have features of AIP, and the diagnosis was confirmed with histology. Recognizing the EUS features for AIP raises the index of suspicion in patients who present with obstructive jaundice and pancreatic mass. The case summary and evaluation have been explained in the video (Video 1, available online at www.giejournal.org).

DISCLOSURE

All authors disclosed no financial relationships.

Abbreviations: AIP, autoimmune transaminase; IgG4, Immunoglobulin subclass 4.

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