

A peculiar case of autoimmune pancreatitis and sclerosing cholangitis

A 65-year-old man presented with recurrent abdominal pain and slight weight loss. Computed tomography (CT) and endoscopic ultrasound (EUS) revealed a hypodense, partially cystic mass located in the pancreatic body and tail that raised the suspicion of pancreatic cancer (Fig. 1). No distant metastases were detected and the patient underwent surgical resection of the mass and splenectomy, which was complicated by a bleeding splenic artery aneurysm. Surprisingly, only massive inflammation of the pancreas was found on histological examination of the resected specimen.

The patient re-presented 1 year later with jaundice for the first time. EUS demonstrated a pseudocyst within the head of

the pancreas. Endoscopic retrograde cholangiopancreatography (ERCP) revealed an isolated stenosis of the distal common bile duct (Fig. 2a). This was successfully treated with regularly exchanged endoprotheses, leading to a complete normalization of laboratory parameters.

The patient returned 2 years later with fever and jaundice. This time ERCP showed a purulent cholangitis with a stricture of the biliary bifurcation as well as several stenotic intrahepatic bile ducts (Fig. 2b). Brush cytology raised a suspicion of cholangiocarcinoma. Serum IgG4 levels, antinuclear antibody (ANA), antineutrophil cytoplasmic antibody (ANCA), anticarbonic anhydrase II and antilactoferrin antibodies were all within the nor-

mal range; however, there was peripheral eosinophilia, which can be associated with autoimmune pancreatitis (AIP) [1]. Histological re-examination of the surgical specimen revealed an impressive lymphoplasmacytic infiltration of the resected pancreas with IgG4-positive cells (Fig. 3a, b). This led finally to the correct diagnosis of AIP type 1 with metachronous autoimmune cholangitis. The patient was treated according to the recently published diagnostic algorithms [2–4] with long-term, slowly tapered prednisone, with resulting improvement in his clinical symptoms, biliary strictures (Fig. 2c), blood eosinophilia, and laboratory parameters.

In summary, distinguishing AIP from pancreatic cancer remains a pitfall for clinicians; in some cases, surgical resection remains the treatment of choice because IgG4 levels can be elevated in 10% of pancreatic cancers [5].

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Fig. 1 Preoperative computed tomography (CT) scan taken 40 seconds after intravenous injection of contrast medium showing a hypodense mass located in the pancreatic body and tail, which is highly suspicious for pancreatic cancer. (Arrow indicates the transition from the pathologic to the normal parenchyma.)

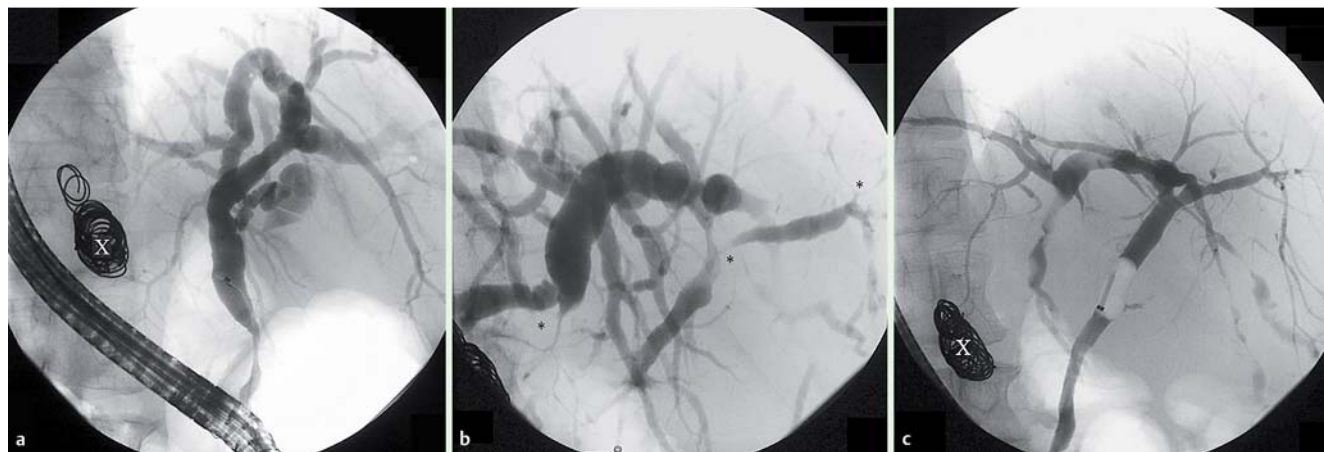


Fig. 2 Images during endoscopic retrograde cholangiopancreatography (ERCP). **a** A stenosis of the distal common bile duct is shown 1 year after surgical resection of the pancreatic mass. **b** A hilar stenosis and several intrahepatic bile duct stenoses (*) are seen 2 years later. **c** Significant improvement of the stenoses is seen following corticosteroid therapy. (X indicates coils within a splenic artery aneurysm.)

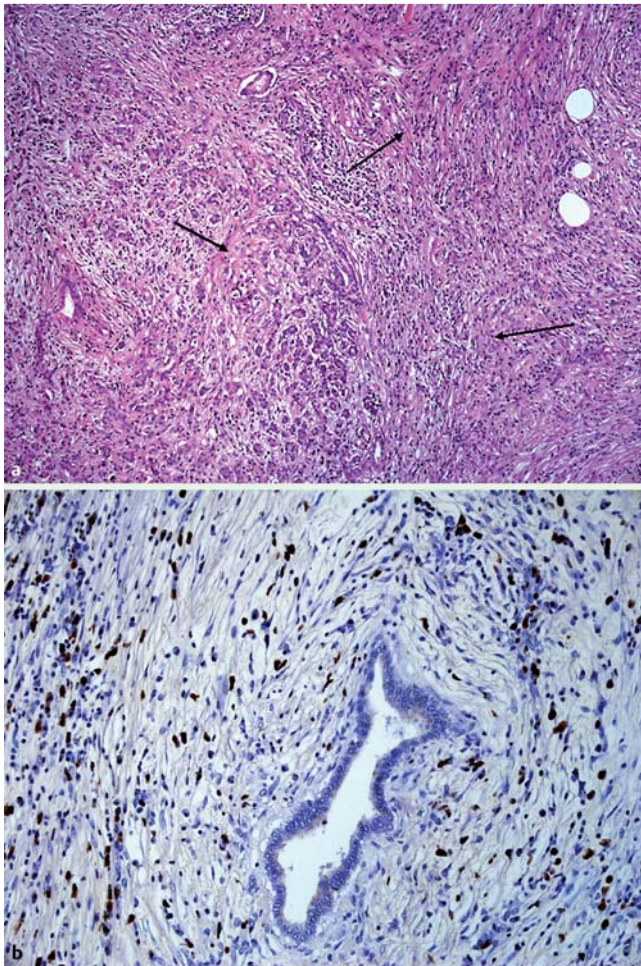


Fig. 3 Histological re-examination of the surgical specimen showing: **a** typical signs of autoimmune pancreatitis including lymphoplasmacytic infiltration with storiform fibrosis (arrows) on hematoxylin and eosin (H&E) staining; **b** cells that are positive for IgG4 (dark brown staining).

References

- 1 Sah RP, Pannala R, Zhang L et al. Eosinophilia and allergic disorders in autoimmune pancreatitis. *Am J Gastroenterol* 2010; 105: 2485–2491
- 2 Otsuki M, Chung JB, Okazaki K et al. Asian diagnostic criteria for autoimmune pancreatitis: consensus of the Japan-Korea Symposium on Autoimmune Pancreatitis. *J Gastroenterol* 2008; 43: 403–408
- 3 Chari ST, Smyrk TC, Levy MJ et al. Diagnosis of autoimmune pancreatitis: the Mayo Clinic experience. *Clin Gastroenterol Hepatol* 2006; 4: 1010–1016
- 4 Maillette de Buy Wenninger L, Rauws EA, Beuers U. What an endoscopist should know about immunoglobulin-G4-associated disease of the pancreas and biliary tree. *Endoscopy* 2012; 44: 66–73
- 5 Ghazale A, Chari ST, Smyrk TC et al. Value of serum IgG4 in the diagnosis of autoimmune pancreatitis and in distinguishing it from pancreatic cancer. *Am J Gastroenterol* 2007; 102: 1646–1653

Bibliography

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