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 normal 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].

Endoscopy_UCTN_Code_CCL_1AZ_2AG

Competing interests: None

H.-M. Curth1, U. Drebber2, D. Stippel3, C. Bangard4, U. Töx1, H.-M. Steffen1
1 Department of Gastroenterology and Hepatology, University of Cologne, Germany
2 Institute of Pathology, University of Cologne, Germany
3 Department of General, Visceral and Cancer Surgery, University of Cologne, Germany
4 Department of Radiology, University of Cologne, Germany
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
4 Maillette de Buy Wenninger L, Beuers U. What an endoscopist should know about immunoglobulin-G4-associated disease of the pancreas and biliary tree. Endoscopy 2012; 44: 66–73

Bibliography
DOI http://dx.doi.org/10.1055/s-0032-1309350
Endoscopy 2012; 44: E226–E227
© Georg Thieme Verlag KG
Stuttgart · New York
ISSN 0013-726X

Corresponding author
H.-M. Curth, MD
Department of Gastroenterology and Hepatology
University of Cologne
Kerpener Str. 62
50924 Cologne
Germany
Fax: +49-221-4786758
harald-morten.curth@uk-koeln.de