We present three cases of IgG4-related sclerosing cholangitis in association with autoimmune pancreatitis. The patients were all men and were aged 69, 71, and 75; their clinical presentations were with obstructive jaundice and considerable weight loss. Computed tomography (CT) scans showed pancreatic head masses in all three patients, mimicking pancreatic malignancy. Magnetic resonance imaging (MRI) scans showed the biliary trees contained multiple areas of common bile duct stenosis and diffuse parietal thickening of the common bile duct wall. Two of the patients also had intrahepatic biliary involvement. Endoscopic ultrasound scans were performed with needle biopsies taken, the results of which were negative for malignancy but also negative for IgG4. Plasma levels of IgG4 were elevated in all of the patients (five times above the upper limit). Oral corticosteroids were initiated.

After 3 weeks of treatment, the clinical status of the patients markedly improved but the cholestasis persisted. New imaging studies were performed, which showed that the pancreatic head mass had almost disappeared in all three patients, but the biliary tract compromise was still present. We decided to perform peroral cholangioscopy, which showed diffuse involvement of the biliary tree with patchy areas of erythema and mucosal friability (▶Fig. 1; ▶Video 1). Biopsies were again negative for malignancy and for IgG4. After 6 weeks of corticosteroids, the patients no longer had evidence of cholestasis and the new MRI scans showed improvement of the biliary tree stenoses, along with disappearance of the pancreatic head masses. IgG4-related sclerosing cholangitis is frequently associated with autoimmune pancreatitis and is currently recognized as a biliary manifestation of IgG4 systemic disease [1]. The differential diagnosis from primary sclerosing cholangitis and cholangiocarcinoma is sometimes diffi-
cult [2]. Peroral cholangioscopy has numerous applications in the diagnosis and treatment of various biliary disorders and can be a helpful diagnostic method to rule out malignancy in patients with IgG4-related sclerosing cholangitis [3].

Competing interests

The authors declare that they have no conflict of interest.

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