Immunoglobulin G4-Related Sclerosing Cholangitis without Pancreatic Involvement

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A 62-year-old man with a past history of retroperitoneal fibrosis was admitted with obstructive jaundice. CT demonstrated dilated intrahepatic bile ducts and a non-swollen pancreas. Endoscopic retrograde cholangiography (ERC) disclosed an irregular stricture of the hepatic hilar bile ducts (fig. 1). Magnetic resonance cholangiopancrea-

tography (MRCP) demonstrated that the biliary stricture did not spread to the root of the left medial branch (fig. 2). Right hepatic lobectomy, caudate lobectomy and resection of the extrahepatic bile duct with D2 lymph node dissection were performed. A resected specimen showed an irregular but smooth surface of hepatic hilar mucosa and

Fig. 1. ERC disclosed an irregular stricture of the upper part of the common hepatic duct, the root of the right and left hepatic duct and intrahepatic biliary branch of the right lobe, and dilation of the intrahepatic biliary branches.

Fig. 2. MRCP demonstrated that the biliary stricture did not spread to the root of the left medial branch.

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Fig. 3. Gross dissection of the common bile duct and hepatic duct showing an irregular but smooth surface of hepatic hilar mucosa and severe stricture of hepatic hilar, right anterior and right posterior branches.

severe stricture of hepatic hilar, and right anterior and posterior branches (fig. 3). Immunohistological study revealed thickening by diffuse fibrosis and inflammatory cell infiltration mainly composed of IgG4-positive cells.

As a disease entity of autoimmune pancreatitis, IgG4-related disease was established mainly on a report from Japan, and became to be well known as autoimmune pancreatitis accompanied by a sclerosing cholangitis (IgG4-SC) like a primary sclerosing cholangitis or bile duct cancer [1–3]. Zen et al. [4] reported that IgG4-SC occurs in association without autoimmune pancreatitis. Clinically, the differentiation of IgG4-SC from primary sclerosing cholangitis and hilar cholangiocarcinoma becomes to be the problem. Serological examination and immunohistologic evaluation in addition to direct cholangiography are important for diagnosis of IgG4-SC.

References

- 1 Mendes FD, Jorgensen R, Kearch J, Katzmann JA, Smyrk T, Donlinger J, et al: Elevated serum IgG4 concentration in patients with primary sclerosing cholangitis. *Am J Gastroenterol* 2006;101:2070–2075.
- 2 Nakazawa T, Ohara H, Sano H, Aoki S, Kobayashi S, Okamoto T, et al: Cholangiography can discriminate sclerosing cholangitis with autoimmune pancreatitis from primary sclerosing cholangitis. *Gastrointest Endosc* 2004;60:937–945.
- 3 Nakazawa T, Ohara H, Sano H, Ando T, Aoki S, Kobayashi S, et al: Clinical differences between primary sclerosing cholangitis and sclerosing cholangitis with autoimmune pancreatitis. *Pancreas* 2005;30:20–25.
- 4 Zen Y, Harada K, Sasaki M, Sato Y, Tsuneyama K, Haratake J, et al: IgG4-related sclerosing cholangitis with and without hepatic inflammatory pseudotumor, and sclerosing pancreatitis-associated sclerosing cholangitis: do they belong to a spectrum of sclerosing pancreatitis? *Am J Surg Pathol* 2004;28:1193–1203.