A Case of Intrahepatic Cholangiocarcinoma Associated with Primary Sclerosing Cholangitis

A 31-year-old man was referred to our hospital for the treatment of obstructive jaundice. Dynamic CT scan revealed an ill-defined mass in the right liver, 4.0 × 3.5 cm in size, involving the right branch of the portal vein (Fig. 1A). Percutaneous transhepatic cholangiogram showed widespread biliary stricture in the left liver (Fig. 1B), which suggested primary sclerosing cholangitis (PSC) or IgG4-related cholangitis. However, serum IgG4 level was within normal limit and needle biopsy of the tumor in the right liver revealed adenocarcinoma. Thus, under the diagnosis of mass forming-type intrahepatic cholangiocarcinoma with periductal infiltration, the patient underwent extended right hemihepatectomy combined with resection of the extrahepatic bile duct and portal vein.

Pathologically, the tumor consisted of poorly differentiated adenocarcinoma (Fig. 2A; a color version of this figure is available as supplementary data at http://www.jjco.oxfordjournals.org) with marked perineural, portal and venous invasion, and not only bile ducts around the tumor but also scattered intrahepatic bile ducts in the left liver were accompanied with inflammatory cell infiltration and periductal onion skin type fibrosis (Fig. 2B; a color version of this figure is available as supplementary data at http://www.jjco.oxfordjournals.org). The final diagnosis would be intrahepatic cholangiocarcinoma associated with PSC.

Eisho Kanemitsu and Minoru Esaki
Hapatobiliary and Pancreatic Surgery Division
National Cancer Center Hospital
Tokyo, Japan
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