Caroli Disease

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Caroli disease is a rare autosomal recessive disorder characterized by multiple segmental communicating dilatations of the intrahepatic bile ducts affecting all or part of the liver that is often associated with renal disorders. It occurs in 2 forms: the rare isolated variety (type I) characterized by recurrent episodes of cholangitis and absence of periportal fibrosis and the more common variety (type II) associated with congenital hepatic fibrosis and consequently occurs in patients with cirrhosis and portal hypertension with esophageal varices.[1]

A 27-year-old, otherwise healthy man was admitted to our hospital because of septic fever of 7 days' duration associated with mild pain in the right upper abdominal quadrant and anorexia. Physical examination findings were normal except for tenderness of the right hypochondrium above an enlarged liver. Laboratory test results (reference ranges shown parenthetically) were suggestive of cholangitis: serum alkaline phosphatase, 192 U/L (35-110 U/L); γ-glutamyl transpeptidase, 102 U/L (10-75 U/L); C-reactive protein, 21 mg/dL (<0.50 mg/dL); and white blood cell count, 15.3×10^9/L ($3.5\text{-}5.0 \times 10^9$/L). Magnetic resonance imaging of the abdomen showed hepatomegaly and multiple communicating, irregular, tubular (white arrows on left-hand figure), and cystic (black arrows on left-hand figure) dilatations of the intrahepatic bile ducts mainly involving the right lobe. Splenomegaly and other findings suggestive of portal hypertension were absent, but bilateral renal tubular ectasia was evident. Magnetic resonance cholangiopancreatography also showed communications between saccular dilatations of the intrahepatic biliary tract and normal bile ducts (white arrows on right-hand figure). The gallbladder (black star on right-hand figure) was dilated but without thickened walls, and the extrahepatic bile duct was normal (white arrowhead on right-hand figure). These findings were indicative of type I Caroli disease.[2] The patient recovered after a course of ceftriaxone, but during the following 4 months, he experienced at least 5 other episodes of cholangitis. His clinical status stabilized, and he is currently awaiting liver transplantation.

1. Caroli J. Diseases of the intrahepatic biliary tree. *Clin Gastroenterol*. 1973; 2:147-161.
2. Levy AD, Rohrmann CA Jr, Murakata LA, Lonergan GJ. Caroli's disease: radiologic spectrum with pathologic correlation. *AJR Am J Roentgenol*. 2002; 179:1053-1057.