A limited polyarteritis nodosa of the liver and gallbladder accompanied with a solitary cystic dilatation of the intrahepatic bile duct

Sirs,

Although liver involvement in polyarteritis nodosa (PAN) has commonly been found in autopsy studies, clinically evident cases are rare, and may be associated with hepatitis B antigenemia (1). Whereas PAN of the gallbladder has been found at autopsy in 10-40% of cases (2,3), cases manifesting as acute cholecystitis are infrequent (4, 5). Solitary dilatation of the intrahepatic bile duct (IHBD) without choledochal cyst, anomalous pancreaticobiliary ductal union, or congenital cystic change of the kidneys has been rarely reported, and may be described using synonyms such as mucinous cystic neoplasm of the IHBD (6,7).

A 68-year-old female was admitted with complaints of epigastric pain radiating to the back of 2 weeks duration. Previously, she had been quite well except for that she had been a carrier of hepatitis B antigen over 20 years. There were no remarkable signs except for mild epigastric tenderness. Laboratory investigations disclosed the following values: leukocytes 7700/mm^3, hematocrit 37.8%, ESR 38 mm/hr, CRP negative, HBs Ag positive, HBe Ag positive, demonstrated HBV DNA over 2000 pg/ml, ANCA negative, anti-HCV negative, ANA negative, rheumatoid factor negative, C3 70.1 mg/dL (normal, 90-180) and C4 9.0 mg/dL (normal, 10-40). The biochemical tests and uninalysis were within normal limits. A CT scan of the abdomen and ERCP demonstrated a focal dilatation in the IHBD suggesting a biliary stricture or cholangiocarcinoma. Selective hepatic angiography revealed multifocal aneurysms on the branches of the hepatic and cystic arteries, suggestive of PAN. On the 14th day of hospitalisation a laparotomy was carried out to rule out a possible bile duct tumor. Left lobectomy of the liver and cholecystectomy were also done. On examination,the resected liver showed a cystic dilatation of the IHBD and its lumen was filled with clear mucinous fluid. The dilated IHBD was lined by tall columnar mucinous epithelium without stromal invasion or papillary proliferation (Fig. 1). The small and medium-sized arteries in the liver and gallbladder revealed focal and segmental fibrinoid necrosis with infiltration of inflammatory cells. In addition, the hepatic parenchyma was unremarkable. There were histopathologic features consistent with acute cholecystitis, but no calculi were found. A limited PAN of the liver and gallbladder accompanied by a solitary cystic dilatation of the IHBD was diagnosed. Post-operatively, daily prednisolone 30 mg was prescribed and gradually tapered to 5 mg/day within 10 months without any difficulties. The patient has been in remission for almost 2 years.

Our patient had asymptomatic hepatic involvement without laboratory abnormalities, and the gallbladder involvement manifested as acalculous cholecystitis. Most patients with acute cholecystitis can usually be detected preoperatively. However, because there were no remarkable symptoms or signs of acute cholecystitis, nor any abnormalities on the CT scan of the abdomen, acalculous cholecystitis was diagnosed post-operatively.

While clinical involvement of the biliary tree has only been rarely reported, autopsy studies showed the biliary involvement in 10-25% of cases (8, 9). Popovsky *et al.* described a patient with PAN accompanied by Meyenburg complexes (or multiple bile duct adenomas) (10). However, there was no evidence of the biliary involvement by PAN except for the cystic mucinous neoplasm of the IHBD in the current case. In addition,based on high levels of HBV DNA prove and hypocomplementemia, it seems reasonable to assume that the polyarteritis lesions in our case may be associated with hepatitis B antigenemia.

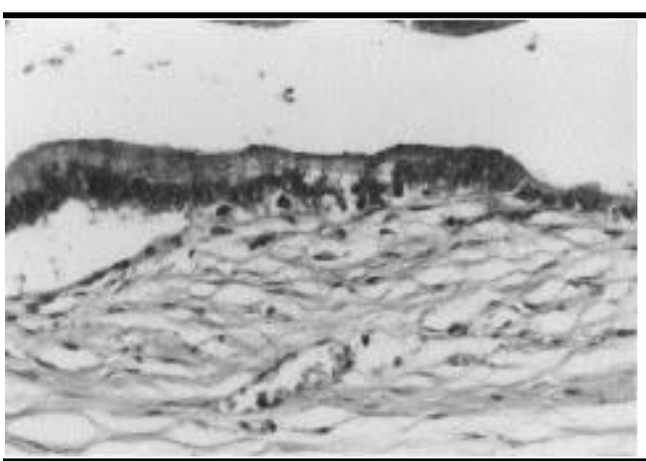

Fig. 1. The dilated bile duct is lined by tall columnar mucinous epithelium. The mucicarmine stain revealed strong positivity of mucin in the cytoplasm. Neither stromal invasion nor papillary proliferation can be seen noted (x200, Mucicarmine stain).

H.K. CHANG[1], *MD* K.T. KIM[2], *MD*
J. CHUNG[3], *MD* H.J. JANG[4], *MD*
D.H. LEE[5], *MD*

[1]Division of Rheumatology, Department of Internal Medicine, [2]Department of Emergency Medicine, Dankook University, Cheonan; [3]Department of Pathology, Gachon Medical School Gil Medical Center, Incheon; [4]Department of General Surgery, [5]Department of Diagnostic Radiology, Asan-Kangnung Hospital, Ulsan University, Kangnung, South Korea.

Address correspondence to: Dr. Hyun Kyu Chang, Division of Rheumatology, Department of Internal Medicine, Dankook University Hospital, 16-5 Anseo-Dong, Cheonan, Chungcheong Nam Do, 330-715, South Korea. E-mail: hanks22@dankook.ac.kr

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