Ectopic hepatocellular carcinoma arising in the bile duct

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Abstract
We report a case of ectopic hepatocellular carcinoma arising in the bile duct. A 72-year-old woman was transferred to our hospital with fever, abdominal pain, and jaundice. Contrast-enhanced computed tomography revealed a round mass, measuring 25 mm in diameter, in the bile duct. The mass was causing obstructive jaundice. Endoscopic retrograde cholangiography showed a 27 mm x 21 mm round defect in the superior bile duct. These findings led to a diagnosis of bile duct tumor, and the patient underwent extrahepatic bile duct resection and biliary reconstruction. Gross examination of the tumor showed a fibrous capsule and a stalk arising from the bile duct mucosa. The tumor was diagnosed histopathologically as well-differentiated hepatocellular carcinoma arising in the bile duct.

Key words Ectopic hepatocellular carcinoma · Ectopic liver · Bile duct

Introduction
Ectopic hepatocellular carcinoma (HCC) is defined as HCC arising from hepatic parenchyma that is located in an extrahepatic organ or tissue. The embryologic development of the liver is generally stable; the reported incidence of ectopic liver is only 0.24%–0.47%.¹² Ectopic HCC is even rarer; only about 20 cases have been reported in Japan.¹ The origin of ectopic HCC is not well understood, but it may arise from a functional deficiency of the ectopic liver. We report herein a case of ectopic HCC arising in the bile duct.

Case report
A 72-year-old woman with a history of hypertension and diabetes mellitus, and a surgical history of cholecystectomy and aortic valve replacement for aortic valve stenosis, was admitted to her neighborhood hospital with fever, abdominal pain, and jaundice. Because her symptoms did not improve despite the administration of antibiotics, she was transferred to Yamaguchi University Hospital, on January 17, 2003. Upon admission, her serum bilirubin was elevated, at 6.4 mg/dl. Hepatitis B surface (HBs) antigen, HBs antibody, and hepatitis C virus (HCV) antibody were negative. Contrast-enhanced computed tomography revealed a round mass, (measuring 25 mm in diameter) in the bile duct, but no tumor was detected in the liver. The bile duct tumor had not invaded to the serosa. There was no lymph node swelling in the hepatoduodenal ligament. The extrahepatic bile duct, the tumor, and regional lymph nodes were resected, and the biliary tract was reconstructed by the Roux-en-Y method.

Examination of the resected specimen revealed a round tumor, measuring 25 mm x 22 mm and connected to the surface of the bile duct mucosa by a stalk. The surface of the bile duct mucosa was smooth (Fig. 2a). Histopathologic examination of the tumor revealed cancer cells proliferating sinusoidally. The cells were almost circular, with abundant cytoplasm that was eosinophilic and showed a fine granular appearance (Fig. 2b). These characteristics supported a diagnosis of well-differentiated HCC. There was no evidence that cancer...
cells had invaded the vessels or the biliary ducts. Immunostaining showed many cancer cells to be positive for cytokeratin (CK) 8 and CK18, but negative for CK7 (not shown). These findings indicated that the cancerous tumor cells were derived from hepatocytes, not from bile duct epithelial cells. Thus, we diagnosed the tumor as ectopic HCC arising in the bile duct.

The patient was discharged from our hospital on postoperative day 52, and has been doing well for the past 12 months.

Discussion

Collan et al. classified anomalous liver into four types: accessory liver lobe, small accessory lobe, ectopic liver, and ectopic liver tissue. They distinguished between ectopic liver, which is outside the liver proper without any connection to it, and accessory liver, which is connected to the liver, often by a stalk. Because the tumor in our patient was clearly separate from the liver but had a stalk, this classification system did not apply.

Ectopic liver is often found incidentally during laparotomy or autopsy. The incidence of ectopic liver identified laparoscopically is estimated at 0.47%. Eiserth

Fig. 1. Endoscopic retrograde cholangiography reveals a 27 mm x 21-mm round defect in the superior bile duct

Fig. 2. a A round tumor, 25 mm x 22 mm, is connected to the surface of the bile duct mucosa by a stalk (arrow). The surface of the bile duct mucosa is smooth. b Cancer cells appear almost circular, with abundant cytoplasm, which is eosinophilic, with a fine granular appearance