—Case Reports—

Mucin-producing Bile Duct Carcinoma Arising from Primary Sclerosing Cholangitis: A Case Report

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Abstract

A 60-year-old woman with primary sclerosing cholangitis (PSC) and high levels of ALP, γ-GTP, and DUPAN-2 was admitted to our institution for examination. The patient did not have ulcerative colitis or pancreatic intraductal papillary mucinous neoplasm. Imaging studies revealed atypical dilation of bile ducts in the left lobe of the liver. Repeated cytologic examinations of the bile showed atypical cells consistent with adenocarcinoma. The patient underwent extended resection of the left lobe of the liver and was found to have intraductal papillary carcinoma with associated mucin-producing bile duct carcinoma. This carcinoma fills dilated bile duct lumens with mucin. This tumor differs morphologically from typical cholangiocarcinoma, which is usually seen in the late stages of PSC. Just one case of mucin-producing bile duct carcinoma arising from PSC has been reported worldwide. The patient has had no signs of recurrence after 27 months. Patients with mucin-producing bile duct carcinoma, as in the case of its pancreatic counterpart, may have a better prognosis and a higher survival rate than patients with typical cholangiocarcinomas.
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Key words: mucin-producing bile duct carcinoma, primary sclerosing cholangitis

Introduction

In primary sclerosing cholangitis (PSC), the bile ducts narrow owing to chronic inflammation. hepatic failure ultimately develops owing to bilious liver cirrhosis. The causes of narrowed bile ducts are thought to be bacterial and viral infections, bile acid, toxins, and abnormalities of immune function, but the details are unclear. In late stages of the illness, PSC may be associated with cholangiocarcinoma. PSC accompanies biliary duct cancer in 7% to 10% of cases.

Mucin-producing bile duct carcinoma is a rare neoplasm in the liver. Mucin causes biliary dilation, obstructive jaundice, and cholangitis.

The bile and pancreatic ducts are different, but have many similarities, such as luminal dilation and the presence of cystic tumors. Mucin-producing bile duct carcinoma and intraductal papillary mucinous neoplasm (IPMN) also may be similar, and there is a possibility that mucin-producing bile duct carcinoma is the counterpart of pancreatic IPMN. Patients with mucin-producing bile duct carcinoma, as in the case of patients with IPMN, may have a better prognosis and a higher survival rate than do patients with typical cholangiocarcinomas. To our knowledge, this is only the second case report to be published in the world.

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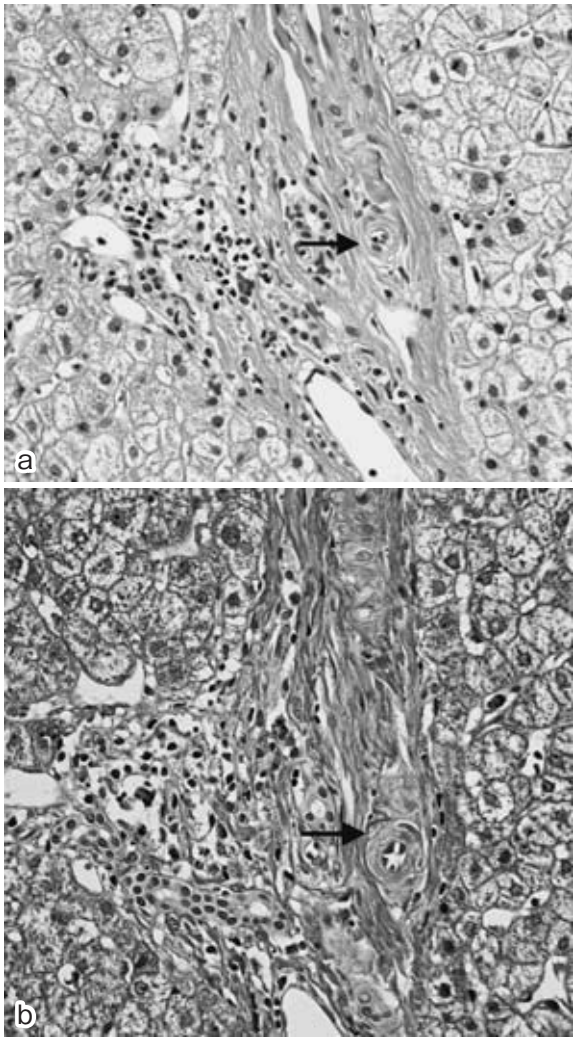

Fig. 1 A liver biopsy indicated that the PSC was Mayo Clinic Group stage I.
(a: hematoxylin and eosin; b: Masson). ←: Bile duct.

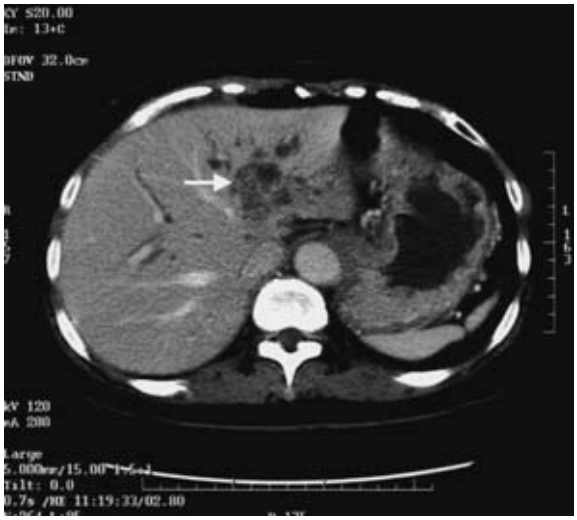

Fig. 2 Computed tomography showed an irregular mass in the liver's medial segment (←). The mass caused severe left bile duct dilation and mild anterior segmental bile duct dilation. Liver dissemination and lymph node metastasis were absent.

Case Report

A 60-year-old woman had received a diagnosis of PSC 1 year before admission. A liver biopsy showed the PSC to be Mayo Clinic Group stage I **(Fig. 1a and 1b)**. Computed tomography scans revealed irregular dilation of the left hepatic duct and an irregular mass in the medial segment of the liver. The mass caused severe dilation of the left bile duct and mild dilation of the anterior segmental bile duct. Liver dissemination and lymph node metastasis were absent **(Fig. 2)**.

Endoscopic retrograde cholangiopancreatography showed bile duct stenosis around the hepatic portal region. The peripheral left bile duct was not contrasted **(Fig. 3a)**. Magnetic resonance cholangiopancreatography showed severe dilation of the left peripheral hepatic duct **(Fig. 3b)**. Repeated cytologic examinations of the bile showed atypical cells consistent with adenocarcinoma. Encasement of the left hepatic artery was revealed by hepatic arteriography, but the right hepatic artery was intact **(Fig. 4a)**. A portohepatogram revealed encasement of the left portal vein **(Fig. 4b)**. A decision was made to perform extended left liver resection and lymph node excision.

The external and cut surfaces of the liver were brown-red and smooth with no evidence of cirrhosis. A thick bile duct in the left lobe and dilated bile duct lumen filled with mucin were seen. There was widening of the periductal regions caused by white gelatinous connective tissue **(Fig. 5)**.

Sections from the left hepatic duct showed intraductal papillary neoplasm associated with invasive, well-differentiated mucinous carcinoma **(Fig. 6a)**. The portal tracts contained proliferating bile ductules accompanied by neutrophils. Periductal concentric fibrosis of the medium bile ducts was present (onionskin-type periductal fibrosis) **(Fig. 6b)**. These findings were consistent with the clinical diagnosis of PSC.

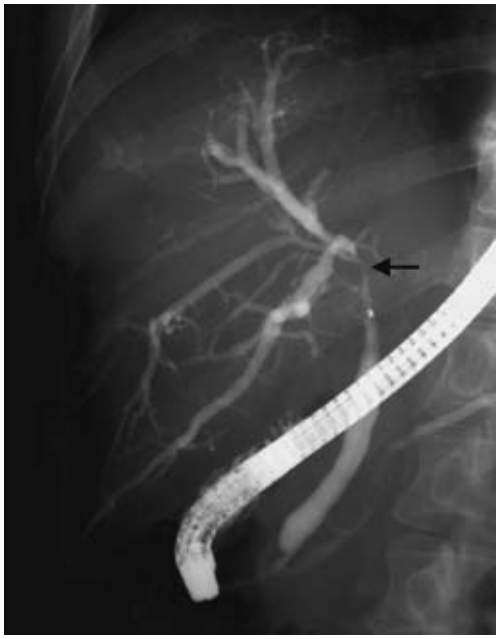

Fig. 3a Endoscopic retrograde cholangiopancreatography showed bile duct stenosis around the hepatic portal region (←). The peripheral left bile duct was not contrasted).

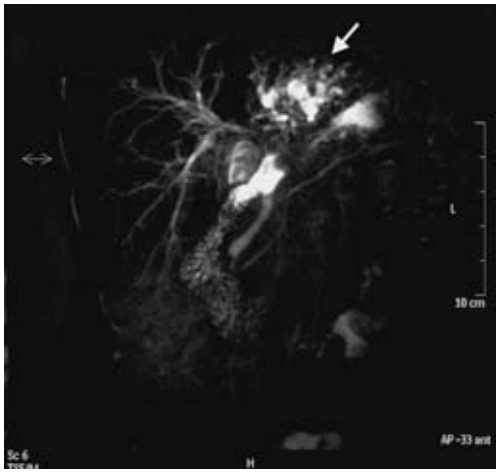

Fig. 3b Magnetic resonance cholangiopancreatography showed severe dilation of the left peripheral hepatic duct (←).

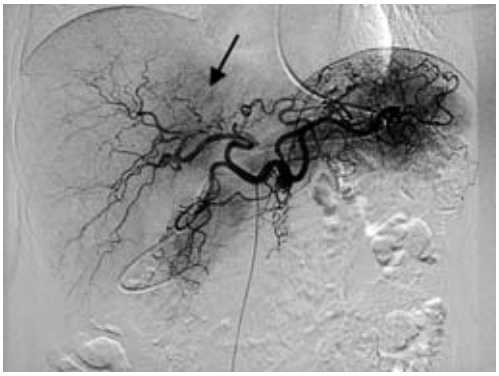

Fig. 4a Hepatic arteriography revealed encasement of the left hepatic artery (←), but the right hepatic artery was intact.

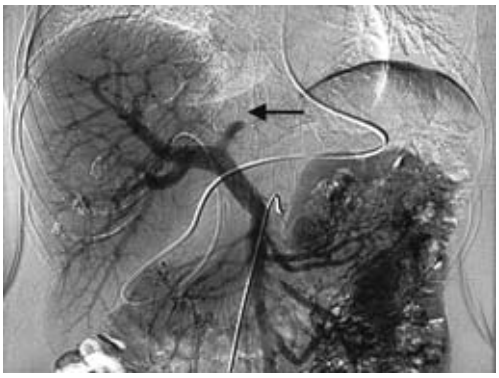

Fig. 4b A portohepatogram revealed encasement of the left portal vein (←).

Fig. 5 The external and cut surfaces of the liver were brown-red and smooth with no evidence of cirrhosis. A thick bile duct in the left lobe and dilated bile duct lumen filled with mucin were seen (←). There was widening of the periductal regions caused by white gelatinous connective tissue.

Although metastasis to lymph nodes of the common hepatic artery was present, the patient has showed no signs of recurrence after 27 months.

Discussion

Some types of bile duct tumor cause bile duct dilation, obstructive jaundice, and cholangitis due to mucin production[3]. When localized, they have been variously referred to as villous adenoma, papillomas, or papillary adenomas, and as mucin-hypersecreting

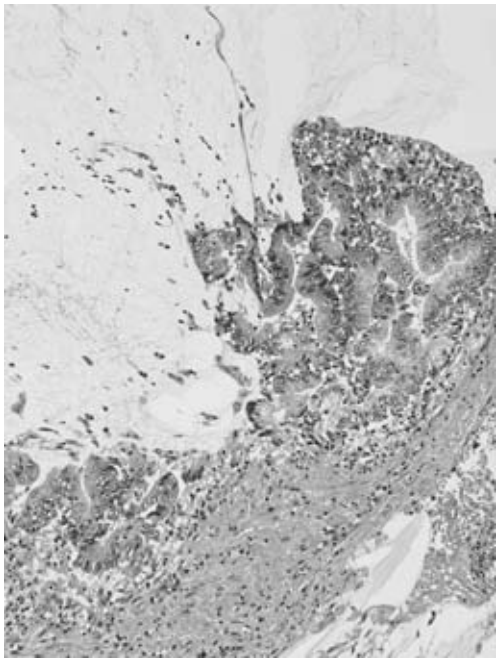

Fig. 6a Sections from the left hepatic duct showed intraductal papillary neoplasm associated with invasive, well-differentiated mucinous carcinoma.

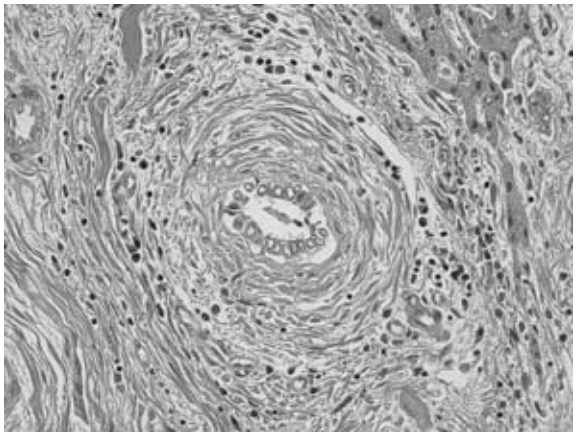

Fig. 6b The portal tracts contained proliferating bile ductules accompanied by neutrophils. Periductal concentric fibrosis of the medium bile ducts was present (onionskin-type periductal fibrosis).

bile duct tumors[3]. Genetically, the bile duct resembles the pancreatic duct. Some mucin-producing bile duct carcinomas of the biliary tract histologically and radiologically resemble pancreatic IPMN[3,4]. Therefore, mucin-producing bile duct carcinoma can be considered the counterpart of pancreatic IPMN[5].

PSC is an idiopathic chronic cholestatic liver disease characterized by inflammatory destruction of the biliary tree. PSC is a slowly progressive disease resulting in biliary fibrosis and obliteration leading to end-stage liver disease. The most feared complication of PSC is cholangiocarcinoma. In a recent report on a PSC cohort, the prevalence of cholangiocarcinoma was approximately 7% over a median follow-up of 11.5 years[6]. Patients with mucin-producing bile duct carcinoma, as in the case of its pancreatic counterpart (IPMN), may have a better prognosis and a higher survival rate than do patients with typical cholangiocarcinomas[7].

Mucin-producing bile duct carcinoma is not known to be associated with PSC. To our knowledge, this is the second reported case of PSC with concurrent mucin-producing bile duct carcinoma.

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