Report on Experiments and Clinical Cases

Extreme Left Hepatic Lobar Atrophy in a Case with Hilar Cholangiocarcinoma

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Abstract

We describe an unusual case of extreme hepatic left lobar atrophy with hilar cholangiocarcinoma. A 67-year-old woman was referred to Nippon Medical School with obstructive jaundice. On admission, computed tomography revealed dilated intrahepatic bile ducts and a defect in the area drained by the left side of the middle hepatic vein. A Spiegel lobe was demonstrated, but the left lobe could not be detected to the left side of the gallbladder. Percutaneous transhepatic cholangiography was performed and demonstrated obstruction of the intrahepatic bile duct at the hepatic hilum. A drainage catheter was left in place. Angiography revealed that the left hepatic artery was present, but there was narrowing of the left portal vein. A diagnosis of agenesis of the left hepatic lobe with hilar cholangiocarcinoma was made. At surgery, the left lobe appeared extremely atrophic without atrophy of the Spiegel lobe. The right anterior branches of the hepatic artery and portal vein had been invaded by carcinoma, so a left trisegmentectomy was performed. Final pathology was advanced hilar cholangiocarcinoma with invasion of the hepatic parenchyma, portal vein, and nervous system. The left lobe was atrophic without hepatolithiasis. The left portal vein was narrow distal to the Spiegel branch. The serum total bilirubin concentration was elevated postoperatively, and the patient was treated for hepatic failure. The patient died of pneumonia without recurrence 7 months after surgery.

This rare case of extreme hepatic left lobar atrophy with hilar cholangiocarcinoma was successfully treated by left trisegmentectomy. Preoperative portal embolization was unnecessary because the left lobe was already atrophic. (J Nippon Med Sch 2002; 69: 278–281)

Key words: cholangiocarcinoma, hepatic lobar atrophy, trisegmentectomy

Introduction

Atrophy of a hepatic lobe or segment is a well-documented, but rare, condition that can be caused by a variety of hepatic diseases. This report describes a case of extreme left lobar atrophy with hilar cholangiocarcinoma.

Case Report

A 67-year-old woman was referred to Nippon Medical School with obstructive jaundice. Medical history was significant for left breast carcinoma resected 6 years previously. On admission, computed tomography (CT) revealed dilated intrahepatic bile

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ducts and a defect in the area drained by the left side of the middle hepatic vein. A Spiegel lobe was demonstrated, but the left lobe to the left side of the gallbladder was not detected (Fig. 1). Ultrasound examination revealed dilated intrahepatic bile ducts in the right lobe, but no ducts were seen in the left lobe. The initial laboratory tests revealed: glutamic-oxaloacetic transaminase, 74 IU/L (normal, <28 IU/L); glutamic-pyruvate transaminase, 200 IU/L (normal, <33 IU/L); alkaline phosphatase, 524 IU/L (normal 66 to 220 IU/L); lactate dehydrogenase, 556 IU/L (normal, 180 to 460 IU/L); gamma-glutamyl transpeptidase, 306 IU/L (normal, 8 to 39 IU/L); total bilirubin, 9.2 mg/dL (normal, <1.0 mg/dL); direct bilirubin, 8.2 mg/dL (normal, <0.4 mg/dL); C-reactive protein, 0.23 mg/dL (normal, <0.3 mg/dL); and white blood cell count, 4,500/µL (normal, 4,000 to 8,000/µL). The serum concentration of CA 19-9 was 178 U/mL (normal <37 U/mL). DUPAN 2 was 230 U/mL (normal <150 U/mL), and CEA was 1.4 ng/mL (normal <2.5 ng/mL). Viral markers for hepatitis B were all negative. Anti-hepatitis C virus (anti-c-100) was also negative.

Percutaneous transhepatic cholangiography was performed and demonstrated an obstruction of the intrahepatic bile duct at the hepatic hilum (Fig. 2). A drainage catheter was left in place. Angiography revealed that the left hepatic artery was present, but that the left portal vein was narrowed (Fig. 3). A diagnosis of agenesis of the left hepatic lobe with hilar cholangiocarcinoma was made.

After 20 days of drainage, an operation was performed. The left lobe was present, but was extremely atrophic without atrophy of the Spiegel lobe. The right anterior branches of the hepatic artery and portal vein had been invaded by carcinoma, so a left trisegmentectomy was performed. The right posterior branch of the intrahepatic bile duct was resected 15 mm distant to the bifurcation of the anterior and posterior branches. Frozen section histologic examination revealed carcinoma at the line of resection, so the resection was extended by 7 mm along the posterior branch of the intrahepatic bile duct. Repeat frozen section examination of the new
line of resection showed no carcinoma. Final pathology revealed advanced hilar cholangiocarcinoma with invasion of the hepatic parenchyma, portal vein, and nervous system. The left lobe was extremely atrophic without hepatolithiasis. The left portal vein was narrow distal to the Spiegel branch.

The postoperative serum total bilirubin concentration was elevated, and the patient was treated for hepatic failure. She died 7 months after operation of pneumonia without recurrence.

Discussion

Lobar atrophy of the liver is rare, although recent routine use of ultrasound and CT in the assessment of hepatobiliary disease has made this diagnosis more common. It turn, increased awareness has led to its recognition on angiograms and at laparoscopy more often than previously.

Hann et al.\textsuperscript{1} described 13 cases of hepatic lobar atrophy that were evaluated for vascular patency and bile duct obstruction. Hepatic lobar atrophy usually occurs in the setting of combined biliary and portal vein obstruction. A correlation exists between lobar atrophy and ipsilateral portal vein obstruction. Ishida et al.\textsuperscript{1} reported six cases of lobar atrophy and investigated the relationship between lobar atrophy and portal flow disturbance. Atrophy of the right lobe was always associated with marked enlargement of the left lobe, but obstruction of flow to the left lobe did not universally result in hypertrophy of the right lobe.

Even though a retrohepatic gallbladder and severely distorted hepatic morphology due to compensatory hypertrophy of the left and caudate lobes may raise the suspicion of agenesis of the right lobe of the liver, the absence of the right hepatic vein, right portal vein and its branches, and dilated right intrahepatic ducts are prerequisites for diagnosing agenesis of the right hepatic lobe on CT. In severe lobar atrophy, at least one of these structures is recognizable\textsuperscript{1}.

Recently, animal models have suggested that a decrease in portal blood flow plays a role in the formation and progression of hepatolithiasis. To determine whether this causal relationship is clinically relevant, Terada et al.\textsuperscript{3} examined the histology of the intrahepatic portal venous and hepatic arterial systems. They found that vascular changes were more severe in patients with hepatolithiasis than in patients who had biliary sludge and microliths, or extrahepatic biliary obstruction. The vascular changes may represent progression of the inflammation associated with cholangitis, which in turn may cause parenchymal atrophy, and if sufficiently severe, lobar atrophy if the hepatolithiasis is progressive.

The following features are characteristic of hepatic lobar atrophy: 1) Advanced septal fibrosis with or without nodular parenchymal changes; 2) Biliary piecemeal necrosis with formation of vascular structures; 3) Ductal proliferation, frequently extending into the septa and involving the parenchyma; 4) Capillarization of the sinusoids with type IV collagen deposition in Disse’s space; 5) Factor VIII-associated antigen expression by the sinusoidal endothelium; and 6) A seemingly paradoxical increase in proliferative activity of hepatocytes, based on evidence from PCNA staining\textsuperscript{4}.

The frequency of hepatic lobar atrophy was 5.3% among 1,208 patients who underwent laparoscopy\textsuperscript{5}.

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**Fig. 3** Angiography reveals that the left hepatic artery (LHA) exists. (a) Narrowing of the left portal vein is observed. (b)
In genesis, bilateral hepatic lobes are the same volume. The right lobe grows and enlarges more than the left lobe at birth. Hepatic lobar atrophy can occur as a congenital anomaly and subsequent to a variety of liver diseases, including chronic hepatitis, idiopathic portal hypertension, primary biliary cirrhosis, drug-induced liver injury, hepatic scarring, autoimmune hepatitis, and metastases from other visceral organs. The most common causes are idiopathic portal hypertension, scarred liver, and primary biliary cirrhosis. Very rare causes of hepatic lobar atrophy include alveolar echinococcosis\(^1\) and primary sclerosing cholangitis\(^1\).

There have been several reported cases of hepatolithiasis and cholangiocarcinoma with hepatic lobar atrophy\(^3,\)\(^4\) Kusano et al.\(^5\) studied 11 patients with Asian cholangiohepatitis to define the relative contributions of portal vein, bile duct, and parenchymal involvement by correlating findings on cholangiography, ultrasound, CT, and angiography. They concluded that the degree of portal vein obstruction correlates most closely with the degree of liver atrophy, although complete central portal obstruction does not necessarily prove that cancer is present.

Hepatolithiasis was not present in this case. The left portal vein was narrow distal to the Spiegel branch. In hilar cholangiocarcinoma, invasion by carcinoma may occasionally narrow the portal vein. Slight hepatic lobar atrophy could result from such narrowing, but extreme atrophy is more likely to represent a congenital anomaly. Extreme hepatic left lobar atrophy with hilar cholangiocarcinoma, as occurred in this case, is rare.

Postoperative liver failure following extensive liver resection for hilar cholangiocarcinoma with jaundice is a major cause of morbidity and mortality. To minimize postoperative liver dysfunction, preoperative portal vein embolization is often used to induce atrophy of the lobe to be resected and hypertrophy of the contralateral lobe in the patients with hilar cholangiocarcinoma\(^6\). In this case, portal embolization was not necessary, because the left hepatic lobe was atrophic already.

In conclusion, we report a rare case of extreme hepatic left lobar atrophy with hilar cholangiocarcinoma. Left trisegmentectomy was performed safely without preoperative portal embolization.

**References**


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