CASE REPORT

A patient with adult extrahepatic portal obstruction, of which distinction from intrahepatic cholangiocarcinoma was difficult

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Abstract : A 51-year-old Japanese male with chief complaints of slightly high fever and epigastralgia was hospitalized at our facility. The inflammatory response was enhanced, and liver dysfunction was observed. Abdominal ultrasonography demonstrated a hyperechoic lesion occupying the left portal vein, and abdominal plain CT indicated a low density of the lesion with a clear boundary, measuring about 3 cm× 2 cm, between the porta hepatis and segment IV of the liver. Contrast CT showed no enhancement in the arterial and portal phases, but a reduction in the density inside the tumor in the equilibration phase was noted. MRI showed hypointensity by T1-weighted imaging and hyperintensity by T2-weighted imaging. Angiography demonstrated an obstruction of the left portal vein and superior mesenteric vein, and endoscopic retrograde cholangiography revealed a constriction in the left intrahepatic bile duct. Since the possibility of intrahepatic cholangiocarcinoma could not be excluded, extended left hepatectomy combined with caudate lobectomy was performed. The tumor, measuring 31 mm× 21 mm×20 mm, was pathohistologically diagnosed as an extrahepatic portal obstruction. Extrahepatic portal obstruction is an important disease that is sometimes difficult to rule out oncologic origin.
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INTRODUCTION

Extrahepatic portal obstruction (EHO) is a disease including an obstruction of the porta hepatis, and shows portal hypertension. EHO occurs most often in infants, and the cause in most patients is reported to be pylethrombosis (1). Adult EHO is often caused by hepatic cirrhosis, idiopathic portal hypertension, tumor embolism, conditions derived from a celiotomy, and pyogenic pylephlebitis followed by abdominal inflammation, such as cholecystitis and appendicitis (2). However, adult EHO, for which apparent causal diseases cannot be clinically identified and which shows abnormalities in liver function but is not accompanied by portal hypertension symptoms, is rare, and its diagnosis as well as distinguishing it from a malignant hepatic tumor is not always easy.

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CASE REPORT

The patient, a 51-year-old Japanese male, consulted a local physician because he had had fever and epigastralgia. Abdominal ultrasonography demonstrated a hyperechoic lesion occupying the left portal vein,

and the patient was admitted to our hospital for close examination and treatment one month later.

The patient had a mild fever but neither anemia nor jaundice, and the abdomen was flat and soft without tenderness. The results of a biochemical examination at the time of admission were as follows: WBC 11,860/mm^3 and CRP 10.8 mg/dl l (normal range, <0.3 mg/dl), showing increased inflammatory reactions, and aspartate aminotransferase 72 U/l (normal range, 8-38 U/l), alanine aminotransferase 126 U/l (normal range, 8-40 U/l), lactate dehydrogenase 576 U/l (normal range, 220-450 U/l), and gamma-glutamyl transpeptidase 91 U/l (normal range, 0-60 U/l), indicative of abnormalities in liver function. In the coagulation system, the prothrombin time was 11.3 sec (normal range, 11-15 sec) and the active thromboplastin time 29.5 sec (normal range, 23-36 sec), showing no abnormalities. The number of thrombocytes was 21.9×10^4/ mm^3, total bilirubin 0.6 mg/dl (normal range, 0.2-1.2 mg/dl), alkaline phosphatase 340 U/l (normal range, 250-380 U/l), and hemoglobin 15.3 g/dl, all of which were within the normal range. Viral markers, such as the hepatitis C virus antibody, hepatitis B surface antigen, and hepatitis B core antibody, were negative, and tumor markers, such as alpha-fetoprotein, carbohydrate antigen 19-9, and carcinoembryonic antigen, were also negative. Abdominal ultrasonography demonstrated a hyperechoic lesion occupying the left portal vein (Fig. 1), and abdominal plain CT showed that the lesion had a low density with a clear boundary, measuring about 3 cm × 2 cm, between the porta hepatis and segment IV of the liver. Contrast CT showed no enhancement in the arterial and portal phases, but reduction of the density inside the tumor in the equilibration phase was noted (Fig. 2). Abdominal MRI demonstrated a node with a diameter of about 2 cm around liver segment IV showing hypointensity by T1-weighted imaging and hyperintensity by T2-weighted imaging (Fig. 3). Splenomegaly was not detected by the imaging methods, nor were abnormalities in the esophagus and stomach, including varices observed by endoscopy of the upper digestive tract. No abnormalities, such as tumor enhancement, were observed by angiography of the common hepatic artery, but the left portal branch and superior mesenteric vein were not enhanced by portography via the superior mesenteric vein (Fig. 4). Endoscopic retrograde cholangiography (ERC) using a balloon catheter revealed a local constriction at the starting point of the left intrahepatic bile duct (Fig. 5). At that point, considering that the elevation of the CRP, fever and alteration of liver function as well as epigastralgia might be caused by a cholangitis, intravenous injection of antibiotics and parenteral nutrition in a fasting state were administered. Thereafter, the presenting symptoms such as fever and epigastralgia remitted, and inflammatory responses normalized. Liver function tests indicated gradual im-

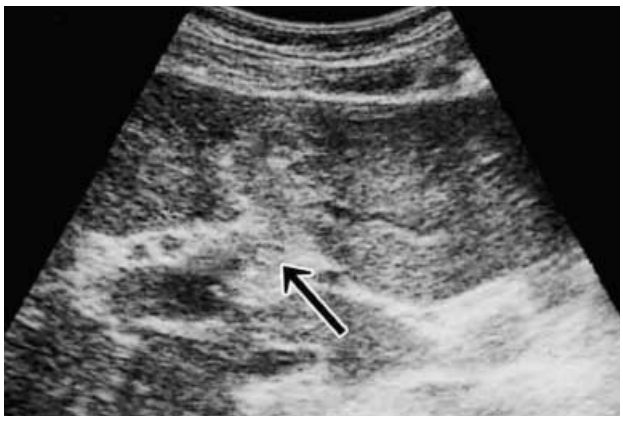

Fig. 1. Abdominal ultrasonography
A hyperechoic lesion occupying the left portal vein was observed (arrow).

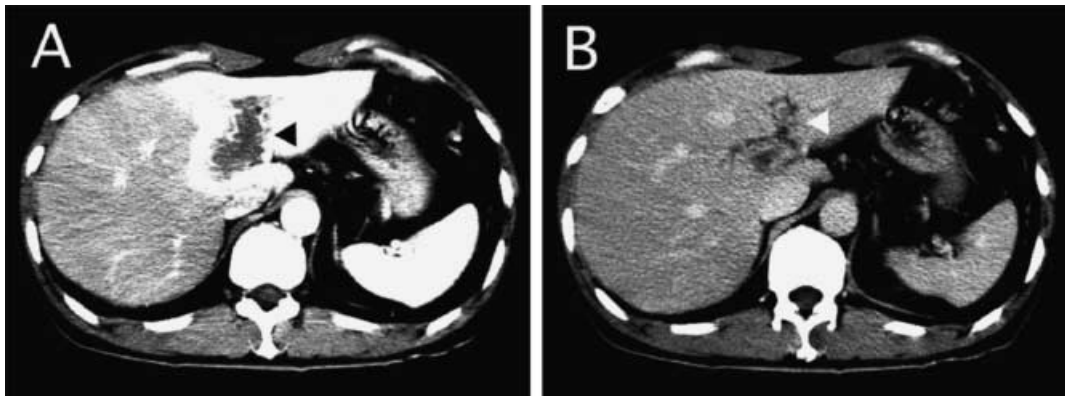

Fig. 2. Dynamic computed tomography of the liver
A low density lesion (about 3 cm × 2 cm in size) (arrow head) was observed between the porta hepatis and segment IV of the liver. No enhancement was detected (A), but a low density was observed in the equilibration phase at the center inside the tumor (arrow head) (B).

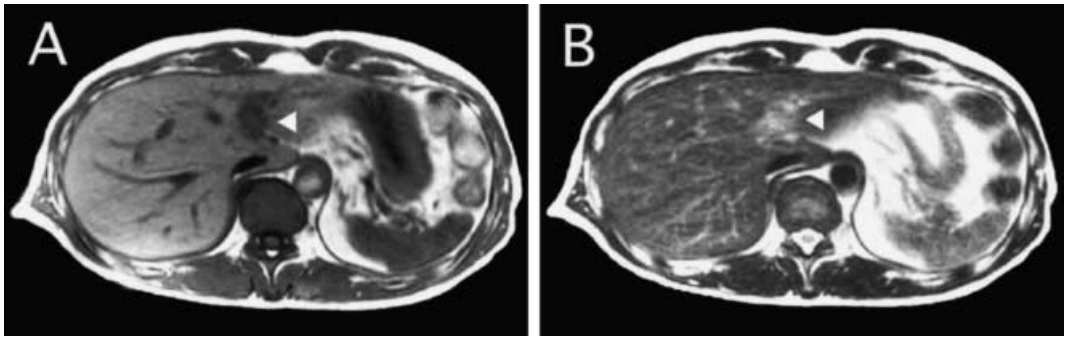

Fig. 3. Magnetic resonance imaging of the liver
A node showing hypointensity (arrow head) by T1-weighted imaging (A) and hyperintensity (arrow head) by T 2-weighted imaging (B) was observed around the liver segment IV.

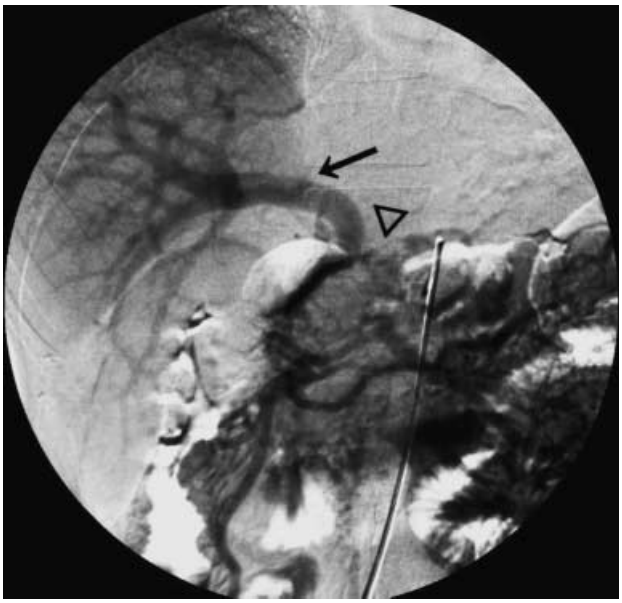

Fig. 4. Portography via the superior mesenteric vein
The left portal branch (arrow) and superior mesenteric vein (arrow head) were not enhanced.

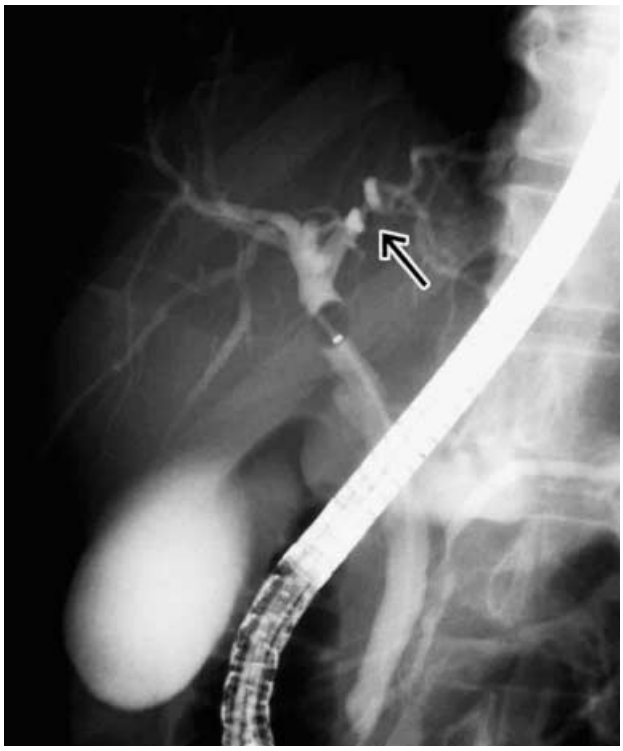

Fig. 5. Endoscopic retrograde cholangiography
A focal stenosis was noted at the origin of the left intrahepatic bile duct (arrow).

provement, but not normalization.

Since the possibility of intrahepatic cholangiocarcinoma could not be excluded based on these findings, extended left hepatectomy combined with caudate lobectomy was performed 35 days after admission. The size of the tumor was 31 mm × 21 mm × 20 mm. What appeared to be a tumor was, in fact, an increased portal area, and proliferation of connective tissues was observed around the starting point of the obstructed left portal vein (Fig. 6). Although portal vein pressure was not measured, findings derived from portal hypertension such as the development of collateral circulation and splenomegaly were not observed. Pathohistologically, no malignancy was detected. Proliferation of bile duct and periductal fibrosis around the portal area was noted with chronic inflammation, and old thrombi that had obstructed the left portal vein were scattered, showing chronic cholangitis and portal thrombosis, respectively (Fig. 7). Neither infiltration by inflammatory cells nor the proliferation of fibroblasts nor capillary vascularization was observed.

Based on these findings, the patient was diagnosed as having EHO. The postoperative course was satisfactory. All results of biochemical blood examinations were normal. As of about 5 years after surgery, no sign of recurrence was observed.

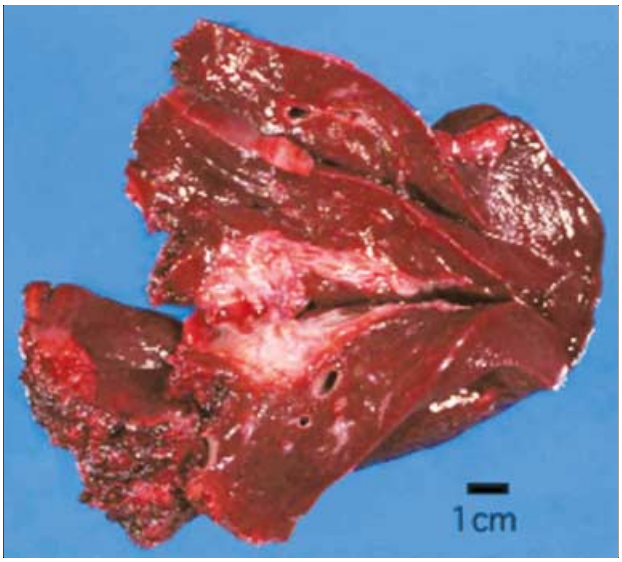

Fig. 6. The cut surface of the tumor in the excised liver specimen The tumor, measuring 31 mm × 21 mm × 20 mm, was a milk-white solid tumor with a clear boundary.

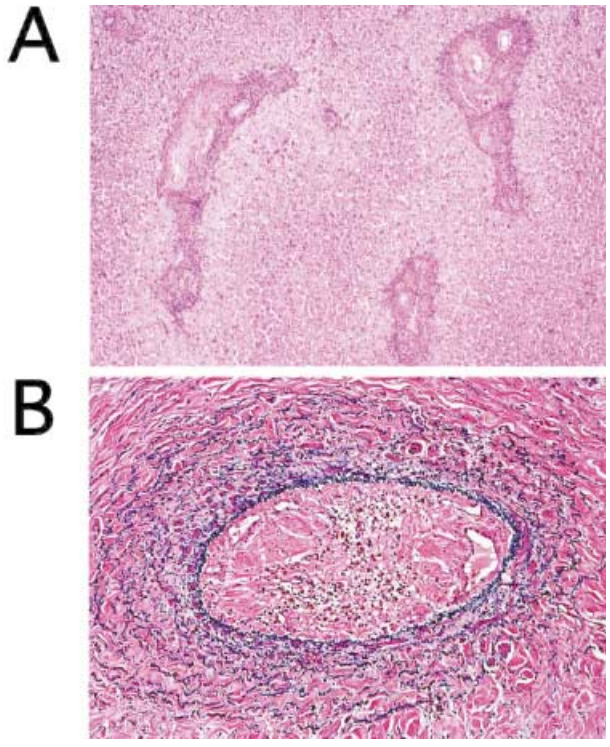

Fig. 7. Microscopic view of the hepatic mass
(A) Most of the portal veins within the mass were occluded by fibrous connective tissue without recanalization, and were surrounded by concentric collagenous connective tissue. The wall of the occluded portal vein did not show any inflammatory or destructive changes (HE stain, × 40). (B) The portal vein is occluded by fibrous connective tissue. The architecture of the venous wall is preserved. The vein is surrounded by dense collagenous connective tissue (Victoria blue & HE stain, × 200).

DISCUSSION

Acquired EHO is caused by tumor embolism, pyogenic pylephlebitis, and pylethrombosis. Pyogenic pylephlebitis and pylethrombosis are frequently caused in infants as the result of an infection in the umbilical vein. In adults, it has been reported that EHO is caused by inflammatory diseases in the abdominal and pelvic cavity, such as cholecystitis, choledochitis, pancreatitis, and appendicitis (1,2). However, EHO patients who had no clearly evident causal infection are often observed. In the patient in this study, epigastralgia, enhanced inflammatory responses and abnormalities of liver function test were observed despite the normal value of the bilirubin in the first examination. The symptoms observed at the time of admission led to consider as a cholangitis. However, these initial findings cannot be the cause of EHO, because the thrombus obstructing the portal vein had not been formed recently. The occlusions of the left portal vein and superior mesenteric vein might be derived from a pyogenic pylephlebitis followed by chronic cholangitis. In the acute EHO stage, a gas image is observed in the portal vein (3), and in the chronic stage, portal hypertension and hepatopetal collateral vessel formation caused by an obstruction of the major portal vein are often observed, particularly, cavernous transformation of the portal vein (CTPV) around the bile duct in the porta hepatis.[1] However, neither a gas image of the portal vein nor CTPV was detected in our patient.

The diagnosis of EHO is clinically made on the basis of history of upper gastrointestinal bleeding, near-normal liver functions, and evidence of portal hypertension and obstruction in the extrahepatic portal vein (1, 4). Thus, histological confirmation should be oriented to the definition of portal or periportal fibrosis as was observed in our patient. In brief, the architectural pattern of the liver is reserved in EHO. There is concentric condensation of reticulin fibers around portal tracts. Such periportal fibrosis could arise from nonspecific, or specific inflammation, extension of the extrahepatic thrombophlebitic process into the intrahepatic radicals of the portal vein, or chemical irritation as a result of hepatocellular breakdown products or bile imbibition (1). The most common site of portal obstruction is at the portal trunk, and often the entire length of the portal vein is occluded with extension into the splenic vein and sometimes into the upper portion of the superior mesenteric vein (1). Obstruction of the porta hepatis and portal trunk results in portal hypertension and is a common cause of upper gastrointestinal bleeding and CTPV. In our patient, however, the detected obstruction site was at the superior mesenteric vein and left portal branch, which might be a reason not to lead to portal hypertension. The segmental obstruction involving the splenic or mesenteric vein is infrequent

(5). Ogawa *et al.* reported that EHO with intrahepatic obstruction of the portal branch tended to predominate (6).

The incidence of EHO is high in India and Southeast Asian countries (7,8). Studies of portal hypertension in Japanese patients with EHO using a questionnaire indicated that esophageal varices and splenomegaly, respectively, were observed in 89% and 70% of the patients (9), and that liver dysfunction was mild (10). Furthermore, abnormalities in coagulability were often observed, and a prolongation of prothrombin time and a decrease in thrombocytes have been reported (7,8,11). In our patient, esophageal varices, splenomegaly, and abnormalities in coagulability were not detected, but sustained liver dysfunction was observed. The CT and MRI findings showed a grossly resemblance to those of intrahepatic cholangiocarcinoma. The obstruction of the left portal vein, observed by abdominal angiography, and a constriction in the left intrahepatic bile duct, observed by ERC, also gave the misleading possibility of tumor embolism or invasion. Although the standard surgical treatment of EHO was portosystemic shunts, the patient in this study has been left untreated after liver resection, because no symptomatic portal biliopathy and hypersplenism including variceal bleeding were observed thereafter. In EHO, the secondary exclusion of the intrahepatic bile duct caused by the proliferation of connective tissues around the obstructed portal vein sometimes shows a constriction in the bile duct, as was observed in our patient. Bayraktar *et al.* reported that a partial obstruction extending along the length of the common bile duct was found in EHO, which was referred to as pseudo-cholangiocarcinoma (12). In some patients, an esophageal varix is formed and ruptured in the bile lumen, in which conditions similar to primary sclerosing cholangitis are observed by cholangiography (13). It is necessary to consider adult EHO, as observed in our patient, as a disease to be distinguished from intrahepatic tumor lesions.

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