Case Reports

Huge Caudate Lobe of the Liver due to Budd-Chiari Syndrome

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Abstract

Budd-Chiari syndrome is characterized by hepatic venous outflow obstruction. We describe a patient with a huge caudate lobe of the liver due to Budd-Chiari syndrome. A 49-year-old woman was referred to Nippon Medical School Hospital to receive treatment for enlarged gastric varices. She had been followed up for idiopathic portal hypertension with deformity of the liver for 7 years and had undergone surgery for pharyngeal carcinoma 5 years earlier. Upper gastrointestinal endoscopy revealed solitary gastric varices without esophageal varices. Abdominal computed tomography revealed obstructions and scars of the right, middle, and left hepatic veins. The caudate lobe was enlarged, and the portal vein was displaced ventrally, without thrombus. The drainage vein of the caudate lobe, the short hepatic vein on the left side of the inferior vena cava, was dilated. The portal vein and drainage vein of segment 6 were visualized and showed no atrophy. Venography revealed no obstruction of the inferior vena cava. Budd-Chiari syndrome with solitary gastric varices was diagnosed on the basis of these findings. We performed balloon-occluded retrograde transvenous obliteration and partial splenic embolization to treat the gastric varices. The posttreatment course was uneventful, and the patient was discharged 8 days after embolization. The gastric varices shrank.

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Introduction

Budd-Chiari syndrome (BCS) is characterized by hepatic venous outflow obstruction. The obstruction causing BCS can be located at the level of the small or large hepatic veins or in the suprarehepatic portion of the inferior vena cava (IVC)\textsuperscript{1,2}. Liver congestion causes hepatocyte dysfunction, which may progress to hepatocyte necrosis and hepatic fibrosis and cirrhosis. Hypertrophy of the central parts of the liver (mainly the caudate lobe) is related in part to the maintenance of adequate venous drainage through the numerous small caudate lobe veins that reach the IVC directly.

We describe a patient with a huge caudate lobe of the liver due to BCS.
Case Report

A 49-year-old woman was referred to Nippon Medical School Hospital to receive treatment for enlarged gastric varices. She had been followed up for idiopathic portal hypertension with deformity of the liver for 7 years and had undergone surgery for pharyngeal carcinoma 5 years earlier. Initial laboratory tests revealed the following values: serum aspartate aminotransferase, 23 IU/L (normal, <28 IU/L); serum alanine aminotransferase, 13 IU/L (normal, <33 IU/L); serum alkaline phosphatase, 155 IU/L (normal 66 to 220 IU/L); serum lactic dehydrogenase, 284 IU/L (normal, 180 to 460 IU/L); serum gamma glutamic transpeptidase, 36 IU/L (normal, 8 to 39 IU/L); total serum bilirubin, 1.2 mg/dL (normal, 0.2 to 1.2 mg/dL); direct serum bilirubin, 0.3 mg/dL (normal, <0.4 mg/dL); serum hyaluronic acid, 50 ng/dL (normal, <50 ng/dL); serum albumin, 4.1 g/dL (normal, 3.8 to 5.5 g/dL); serum C-reactive protein, 0.1 mg/dL (normal, <0.3 mg/dL); white blood cell count, 3,900/μL (normal, 4,000 to 8,000/μL); red blood cell count, 321 × 10^6/μL (normal, 410 to 550 × 10^6/μL); serum hemoglobin concentration, 10 g/dL (normal, 14 to 18 g/dL); and serum platelet count, 15.0 × 10^9/μL (normal, 20 to 40 × 10^9/μL). The serum concentration of PIVKA-2 was 20 mAU/mL (normal, <40 mAU/mL), and that of alpha-fetoprotein was 12.6 ng/mL (normal, <10 ng/mL). The indocyanine green clearance rate at 15 minutes was 3.8% (normal, <10%). The serum surface antigens for hepatitis B and anti-hepatitis C virus antibodies were negative.

Upper gastrointestinal endoscopy revealed solitary gastric varices without esophageal varices (Lgcf,Cb,Fz,RCs) (Fig. 1).

Abdominal computed tomography revealed obstructions and scars of the right, middle, and left hepatic veins. The caudate lobe was enlarged, and the portal vein was displaced ventrally, without thrombus. The drainage vein of the caudate lobe, the short hepatic vein on the left side of the IVC, was dilated. The portal vein and drainage vein of segment 6 were visualized and showed no signs of atrophy (Fig. 2). Venography revealed no obstruction of the IVC (Fig. 3).

BCS with solitary gastric varices was diagnosed on the basis of the findings described above. Because the patient wanted to undergo prophylactic treatment for the gastric varices, we performed balloon-occluded retrograde transvenous obliteration (B-RTO). A balloon catheter was inserted into the shunt through the femoral vein and the IVC and inflated to occlude blood flow (Fig. 4). A solution of 5% ethanolamine olate with iopamidol was slowly injected through the catheter until it filled the shunt. After 24 hours of balloon occlusion, the catheter was removed**. We also performed partial splenic embolization (PSE) to decrease portal venous pressure to the level before B-RTO. A femoral artery approach was used for superselective catheterization of the splenic artery. The catheter tip was placed as distally as possible in either the hilus of the spleen or an intrasplenic artery. Embolization was achieved by injection of a gelatin sponge cut into 2-mm cubes and suspended in saline solution containing antibiotics**.

The posttreatment course was uneventful, and the patient was discharged 8 days after embolization. The gastric varices shrank.

Discussion

Primary BCS is a rare, multifactorial disease in which several prothrombotic disorders are necessary for the development of thrombosis at this uncommon location. A combination of several
Fig. 2 Abdominal computed tomography revealed obstructions and scars of the right (1), middle (2), and left (3) hepatic veins a). The caudate lobe was enlarged, and the portal vein (4) was displaced ventrally, without thrombus b). The drainage vein of the caudate lobe, the short hepatic vein (5) on the left side of the IVC, was dilated. The portal vein (6) and drainage vein (7) of segment 6 were visualized, with no signs of atrophy c) d).

Fig. 3 Venography revealed no obstruction of the IVC prothrombotic conditions is present in at least 35% of patients, which is a rate several times higher than

Fig. 4 A balloon catheter was inserted into the shunt through the femoral vein and the IVC and inflated to occlude blood flow (B-RTO).

that in the general population**. The most common underlying conditions in patients with BCS are myeloproliferative disorders, such as polycythemia
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vera and essential thrombocytosis. Oral contraceptive use has long been known to be a risk factor for BCS, particularly that associated with pure hepatic vein involvement. Pregnancy also appears to be a triggering factor for hepatic vein thrombosis. In both instances, however, an underlying condition is usually present. Nonetheless, about 20% of cases of BCS are idiopathic, as was the case in our patient.

Obstruction is usually caused by a thrombus, but may result from extrinsic compression (tumors, abscesses, cysts), membranous webs at the level of the hepatic vein or the IVC, or postoperative complications of liver transplantation. Short stenoses are particularly visible in the cephalad segment of the IVC or major hepatic veins. Rarely, all hepatic veins are involved simultaneously. Extrahepatic portal vein thrombosis is superimposed on BCS in about 15% of patients.

Site-specificity might be present within the hepatic venous outflow tract. Myeloproliferative diseases are clearly more common in patients with BCS than in patients with portal vein thrombosis and are far more common than in patients with venous thrombosis at other sites. Factor V Leiden appears to be particularly common in patients with IVC obstruction. Furthermore, oral contraceptives and pregnancy have been specifically associated with hepatic vein involvement. In our patient, the right, middle, and left hepatic veins were occluded, but the short hepatic vein and inferior right hepatic vein were not. Site-specificity might have occurred, leading to enlargement of the caudate lobe.

The diagnosis of BCS is based on the demonstration of an obstructed hepatic venous outflow tract. Langlet et al. has classified BCS into the following types: type 1 (acute injury only, corresponding to the onset of hepatic outflow obstruction), type 2 (chronic lesions only, corresponding to the sequelae of remote hepatic outflow obstruction), and type 3 (acute injury superimposed on chronic lesions). The prevalence was estimated to be 7% for type 1, 45% for type 2, and 48% for type 3. Patients with type 1 BCS had the best outcomes, whereas those with type 3 had the worst. We suspect our patient had type 2 BCS.

The clinical manifestations of BCS have been well characterized. Patients with BCS present with various signs, symptoms, and acuity. The presentation of patients with BCS is governed by the extent and swiftness of hepatic outflow obstruction and the body's ability to decompress the liver via the development of collateral blood flow. Abdominal pain, ascites, liver and spleen enlargement, and portal hypertension are important features, along with prominent dilation of the subcutaneous veins of the trunk in patients with long-standing IVC obstruction. If untreated, symptomatic forms of BCS have a poor prognosis: an estimated 90% of patients die within 3 years.

There are various treatments for BCS, such as anticoagulation therapy, interventional radiology, and liver transplantation. In our patient, enlarged gastric varices were present because of portal hypertension. Various treatments are also available for gastric varices, including endoscopic therapy, interventional radiology, and surgery. B-RTO and PSE were performed to treat the gastric varices. We did not recommend the prophylactic treatment of gastric varices in our previous reports, but our patient requested such treatment. Gastric varices are a type of portal-systemic shunt associated with portal hypertension. Obliteration of the portal-systemic shunt increased the portal venous pressure, leading to portal congestion and increased portal venous pressure. After B-RTO, PSE was performed incrementally to reduce portal venous pressure to the level it was before obliteration B-RTO. A new portal-systemic shunt may be less likely to develop at this lower portal venous pressure.

References


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