—Case Reports—

Extrahepatic Portal Venous Obstruction due to a Giant Hepatic Hemangioma Associated with Kasabach-Merritt Syndrome

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Abstract

We describe a patient with extrahepatic portal venous obstruction due to a giant hepatic hemangioma associated with Kasabach-Merritt syndrome. A 67-year-old woman presented with upper abdominal distension and appetite loss. The medical history was not relevant to the current disorder. Initial laboratory tests revealed the following: serum platelet count, $9.9 \times 10^4/\mu L$; serum fibrinogen degradation products, 12 μg/mL; prothrombin time, 1.26; and serum fibrinogen, 111 mg/dL. Computed tomography demonstrated homogenous low-density areas, 15 cm in diameter, in the left lobe of the liver. Common hepatic arteriography revealed a hypervascular tumor with pooling of contrast medium in the delayed phase. The portal venous phase of supramesenteric arteriography revealed obstruction and cavernous transformation of the portal vein. We diagnosed extrahepatic portal venous obstruction due to a giant hepatic hemangioma associated with Kasabach-Merritt syndrome. Laparotomy was performed, and the liver was found to be markedly enlarged. After mobilization of the left lobe, left hepatectomy was performed with intermittent clamping. After resection, Doppler ultrasonography revealed recovery of the portal venous flow. The cavernous transformation shrank. Pathologic examination of the surgical specimen confirmed the presence of a giant benign hepatic cavernous hemangioma. The patient was discharged 16 days after operation. Laboratory data and complications improved after 2 months.
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Key words: hemangioma, Kasabach-Merritt syndrome, giant, portal venous obstruction

Introduction

Hepatic hemangiomas are congenital vascular malformations and the most common benign tumors arising in the liver, with an estimated prevalence of 0.4% to 7.3% in the general population[1,2]. Most hepatic hemangiomas are asymptomatic, but some (especially large lesions) cause various complications. We describe a patient with extrahepatic portal venous obstruction due to a giant hepatic hemangioma associated with Kasabach-Merritt syndrome.

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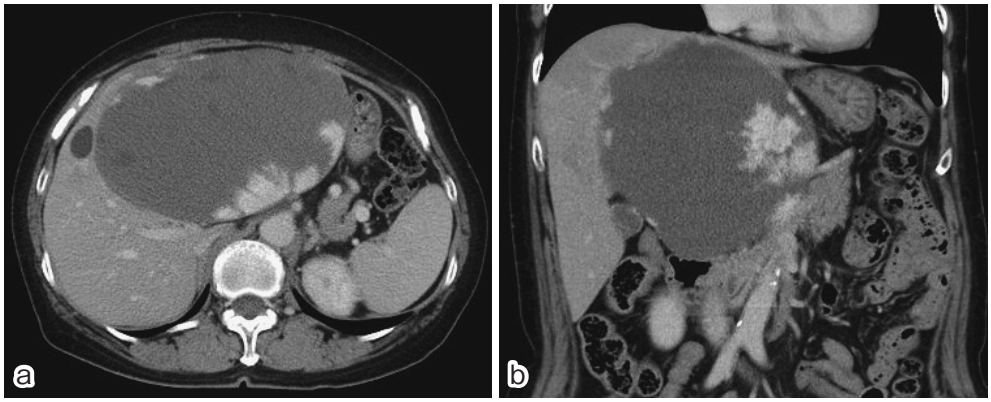

Fig. 1 A computed tomographic (CT) scan, showing homogenous low-density areas, 15 cm in diameter, in the left lobe.

Case Report

A 67-year-old woman presented with upper abdominal distension and appetite loss. The medical history was not relevant to the current disorder. Initial laboratory tests revealed the following: serum aspartate aminotransferase, 37 IU/L (normal, <28 IU/L); serum alanine aminotransferase, 40 IU/L (normal, <33 IU/L); serum alkaline phosphatase, 415 IU/L (normal 66 to 220 IU/L); serum lactic dehydrogenase, 230 IU/L (normal, 180 to 460 IU/L); serum gamma glutamic transpeptidase, 151 IU/L (normal, 8 to 39 IU/L); serum C-reactive protein, 0.10 mg/dL (normal, <0.3 mg/dL); white blood cell count, 4,500 /μL (normal, 4,000 to 8,000 /μL); red blood cell count, 407 × 10^4/μL (normal, 410 to 550 × 10^4/μL); serum hemoglobin concentration, 12.7 g/dL (normal, 14 to 18 g/dL); serum platelet count, 9.9 × 10^4/μL (normal, 20 to 40 × 10^4/μL); serum fibrinogen degradation products, 12 μg/mL (normal, <10 μg/mL); prothrombin time (international normalized ratio), 1.26 (normal, 0.70 to 1.13); and serum fibrinogen, 111 mg/dL (normal, 170 to 410 mg/dL). The serum concentration of carcinoembryonic antigen was 1.0 ng/mL (normal <2.5 ng/mL), and that of CA 19-9 was 1.0 U/mL (normal <37). Computed tomography (CT) demonstrated homogenous low-density areas, 15 cm in diameter, in the left lobe of the liver (**Fig. 1**). Drip infusion cholangiographic CT showed that the intrahepatic bile duct was compressed by the tumor (**Fig. 2**). Common hepatic arteriography revealed a hypervascular tumor with pooling of contrast medium in the delayed phase. The portal venous phase of supramesenteric arteriography revealed obstruction and cavernous transformation of the portal vein (**Fig. 3**). We diagnosed extrahepatic portal venous obstruction due to a giant hepatic hemangioma associated with Kasabach-Merritt syndrome.

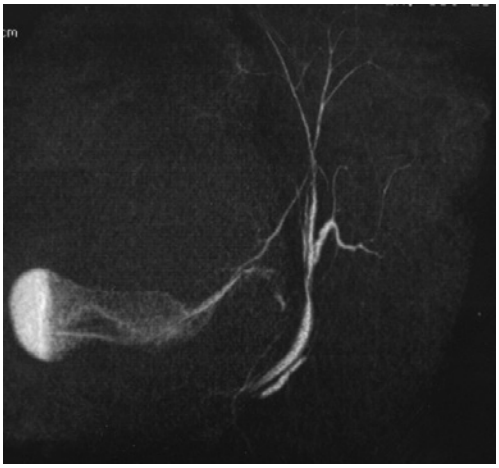

Fig. 2 A drip infusion cholangiographic CT scan, revealing compression of the intrahepatic bile duct by the tumor.

Laparotomy was performed, and the liver was found to be markedly enlarged. Dissection of the hilar structures at the mid-hilum was not performed to avoid injuring the cavernous transformation of the portal vein. Intraoperative ultrasound was done to determine the exact cutting line. After mobilization of the left lobe, left hepatectomy was performed with intermittent clamping (Pringle maneuver)[3]. After resection, Doppler

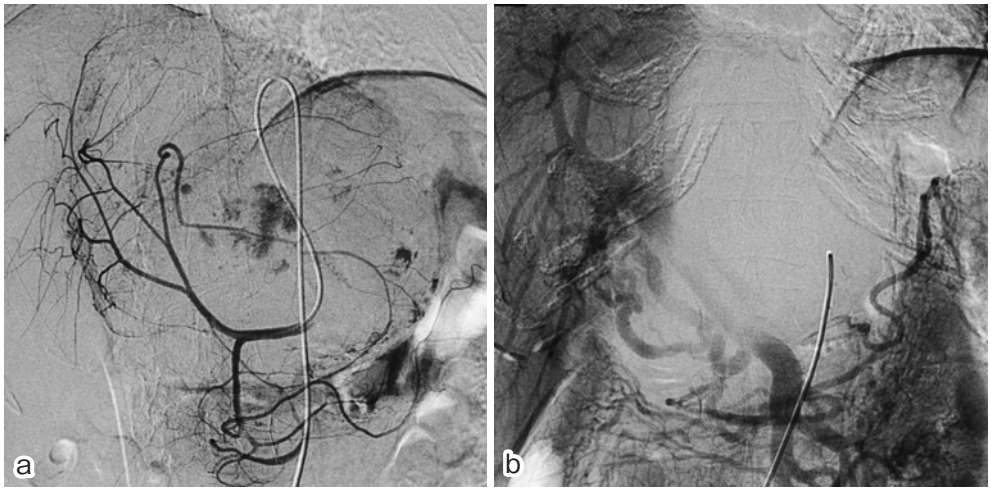

Fig. 3 Common hepatic arteriography revealed a hypervascular tumor with pooling of contrast medium in the delayed phase (a). The portal venous phase of supramesenteric arteriography revealed obstruction and cavernous transformation of the portal vein (b).

ultrasonography revealed recovery of the portal venous flow. The cavernous transformation shrank. Biliary leakage tests using an injection of saline and air were performed, and several leakage points were repaired with fine sutures. Hemostasis of the cut surface of the liver was achieved by ligation and the application of a fibrin glue spray (Bolheal; Chemo-Sero Therapeutic Research Institute, Kumamoto, Japan). The greater omentum was fixed to the peritoneum to prevent delayed gastric emptying[4,5]. An external drainage catheter (19 Fr. BLAKE Silicon Drain; Ethicon, Somerville, NJ, USA) was positioned in the space created by surgery. The weight of the resected specimen was 801 g.

Pathological examination of the surgical specimen confirmed the presence of a giant benign hepatic cavernous hemangioma. After operation, minor biliary leakage and right pleural effusion occurred. The biliary leakage decreased, and the patient recovered after drainage of the pleural effusion. The patient was discharged 16 days after operation. Laboratory data and complications improved after 2 months.

Discussion

Hemangiomas are the most common primary liver tumor. The size of most hepatic hemangiomas remains stable[6,7]. A giant hemangioma, defined as a hemangioma exceeding 4 cm in diameter, can cause symptoms and require treatment[8]. Complications of hemangiomas include congestion, bleeding, thrombosis, infarction, Kasabach-Merritt syndrome, spontaneous rupture, obstructive jaundice, and gastric outlet obstruction[8–13].

Many cases of Kasabach-Merritt syndrome have been described, mostly in infants with cutaneous hemangiomas. Rarely, hepatic hemangiomas associated with Kasabach-Merritt syndrome and diffuse intravascular coagulopathy have been described. The primary pathophysiologic event of Kasabach-Merritt syndrome is platelet trapping by clotting and fibrinolysis within a vascular lesion[14]. The lesion may be acute and massive or chronic and low grade. More rarely, portal venous obstruction can be caused by a giant hepatic hemangioma[15]. In our patient, cavernous transformation of the portal vein was caused by extrahepatic portal venous obstruction by a giant hemangioma, 15 cm in diameter, associated with Kasabach-Merritt syndrome.

Various treatments are available for portal hypertension[16–20]. Because the portal vein was occluded by the tumor in our patient, portal hypertension improved after hepatic resection. Thrombocytopenia can be caused by hypersplenism due to portal hypertension, as well as by clotting and fibrinolysis within vascular lesions such as hemangiomas.

Because a high risk of bleeding is a primary cause

of death in Kasabach-Merritt syndrome, aggressive management is required. Symptomatic giant hemangiomas require some form of treatment, such as interferon, radiation, arterial embolization, surgical resection, and liver transplantation. However, long-term outcomes are disappointing for treatments other than liver transplantation or resection. Recent interest has focused on surgical therapy, which has produced excellent results[10,12,21–27]. In our patient, the laboratory data improved after operation. Surgical resection was considered an effective treatment.

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