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Case Report

Bilateral Lower Limb Edema Caused by Compression of the Retrohepatic Inferior Vena Cava by a Giant Hepatic Hemangioma

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Hemangiomas are the most common benign primary tumors of the liver and their prevalence ranges from 0.4% to 20%. Approximately 85% of hemangiomas are clinically asymptomatic and are incidentally detected in imaging studies performed for other causes. In a very small minority of patients, nausea, vomiting, abdominal pain, distension, palpable mass, obstructive jaundice, bleeding, and signs and symptoms of Budd-Chiari syndrome may develop due to compression of bile duct, hepatic vein, portal vein, and adjacent organs. Occasionally, external compression of inferior vena cava may lead to edema and/or indirect symptoms such as deep vein thrombosis of the lower limbs. In this report, we present a case of giant hepatic hemangioma that completely filled the right lobe of the liver. The patient presented with bilateral lower limb edema and pain. A computed tomography scan detected a 9 × 11 × 12 cm mass indicative of a hemangioma in the right lobe of the liver that compressed the inferior vena cava. The patient refused treatment initially but returned 6 months later presenting with the same symptoms. At that time, the mass had increased in size and a hepatectomy was performed, preserving the middle hepatic vein. By postoperative month 13, the swelling in the lower extremities had decreased significantly and the inferior vena cava appeared normal.

Key words: Liver – Giant hemangioma – Venous obstruction – Lower limb edema

Hemangiomas are the most common benign primary tumors of liver and their prevalence in different autopsy series ranges from 0.4% to 20%.[1–4] Liver hemangiomas are generally smaller than 3 cm and a considerable majority of patients are asymptomatic. Thus, they are incidentally de-

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tected in imaging studies performed for other causes.[1] In a very small minority of patients, signs and symptoms including nausea, vomiting, abdominal pain, palpable mass, obstructive jaundice, bleeding, Budd-Chiari syndrome, and edema and deep vein thrombosis in the lower limbs develop due to compression of the bile duct, hepatic vein, portal vein, inferior vena cava, or gastrointestinal system organs.[4–8] The most important factors contributing to these symptoms are the size, growth rate, and localization of the hemangioma in the liver. To our knowledge, only one case of liver hemangioma has been published so far that has reported edema and deep vein thrombosis of the lower limbs resulting from compression of the retrohepatic inferior vena cava.[4] We present in this report a case of giant hepatic hemangioma that completely filled the right lobe of the liver and caused serious bilateral edema in the lower limbs by compressing on the inferior vena cava.

Case Report

A 43-year-old female presented to a private health facility for bilateral lower extremity swelling and pain, which had begun 3 years prior and had become more prominent in the last 6 months preceding presentation. Blood, urine, and cardiac tests including echocardiography at that facility revealed no cardiac or renal abnormalities. Deep vein thrombosis in the bilateral lower extremities was also ruled out using Doppler ultrasonography (US). However, abdominal ultrasonography detected a mass that completely filled the right lobe of the liver and was suggestive of a hemangioma. For this reason, the patient was referred to our institution for liver transplantation and further workup. Previous examinations and tests of the patient were reassessed in our institution and a computed tomography (CT) scan was scheduled to delineate the relationship of the mass with the adjacent structures in the liver. A $9 \times 11 \times 12$ cm mass in the right lobe of the liver, hypodense on noncontrast images and demonstrating nodular enhancement following intravenous contrast administration, was present and caused marked compression in the retrohepatic inferior vena cava, indicative of a lobulated cavernous hemangioma. The patient did not consent to any surgical or invasive intervention and was discharged upon her request.

Six months later, she presented again for increased bilateral swelling and pain in the lower limbs. Physical examination showed diffuse pitting edema in both limbs. The circumference of the right and left thigh, 15 cm proximal to the knee, measured 71.0 and 70.5 cm, respectively, while the circumference of the right and left leg, 10 cm distal to knee, measured 49.0 and 49.2 cm, respectively. On contrast multislice CT, the mass was noted to have grown to $9 \times 12 \times 17$ cm (Fig. 1A and 1B). An operation was scheduled immediately because of the increased mass size, compression of the inferior vena cava, pain, and diffuse edema in the lower extremities.

After laparotomy, the mass filling the right lobe of liver was noted to compress the inferior vena cava (Fig. 2). The hepatoduodenal ligament was turned using the Pringle maneuver. Next, the right hepatic artery and the right portal vein were suspended and the demarcation line for right hepatectomy was determined. A standard right hepatectomy was carried out in a way to preserve the middle hepatic vein. A Cavitron Ultrasonic Surgical Aspirator (CUSA; ValleyLab, Boulder, Colorado, USA) system was used for the parenchymal transection. On postoperative day 6, the measurements of the right and left thighs, 15 cm proximal to knee, were 60.0 and 60.5 cm, respectively. Measurements of the thigh, 10 cm distal to knee, were 41.0 and 41.5 cm for the right and left legs, respectively. Multislice CT scans on postoperative day 7 showed a normal-sized inferior vena cava and a normally drained and perfused liver parenchyma (Fig. 1C and 1D). At the 13-month follow-up, edema of the lower limbs completely resolved.

Discussion

Hemangiomas are benign hamartomatous tumors developing as a result of rapid proliferation of vascular endothelial cells.[9] They are classified histopathologically as capillary and cavernous hemangiomas. Although they are most commonly located in the skin of the head or neck, they may theoretically be found in nearly all organs having vascular endothelial cells. Of the cases in nonskin tissues, the most common gastrointestinal hemangiomas are in the liver. Hepatic hemangiomas are usually red-blue areas, which are subcapsularly located and easily distinguished from normal liver tissue.[1–3,9,10] They are solitary in 90% of cases and tend to be located in the right lobe of the liver.[6] Hemangiomas are usually seen between the ages of 30–50 years, with a female-to-male ratio of 5–6:1.[2,6,8,9,11] Thus, it has been suggested that the disease is related to exogenous steroid hormones or

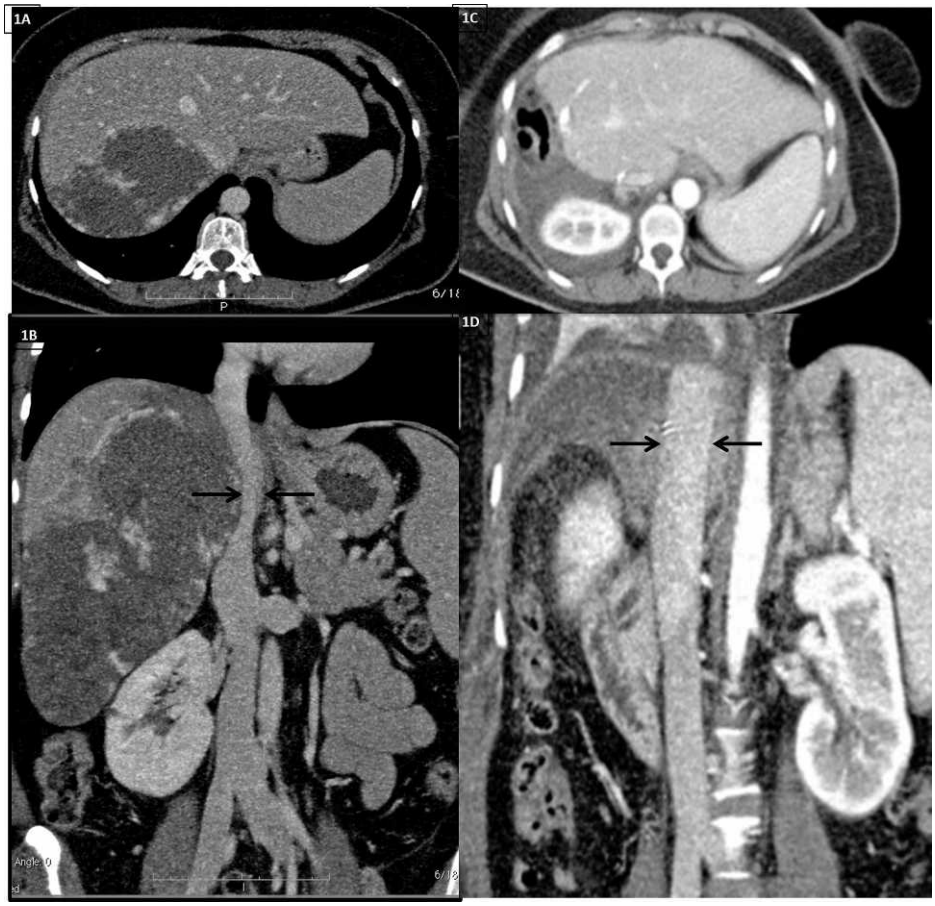

Fig. 1 A peripheral nodular enhancing mass in segments VI and VII of the liver, causing marked compression of the inferior vena cava and showing peripheral nodular contrast uptake, was demonstrated in axial (a) and coronal (b) contrast computed tomography (CT) images taken during the preoperative period. Axial (c) and coronal (d) contrast CT images taken on day six postoperatively show that the compression completely disappeared. In addition, fluid collection secondary to the operation is visible in the hepatectomy space.

gynecologic hormones, albeit studies have failed to show a definite connection.[1,3,6,9]

Previous studies have shown that liver hemangiomas are usually smaller than 3 cm and 85% of lesions are asymptomatic. Therefore, the majority of lesions are detected incidentally by radiologic tests done for screening purposes. Despite there being a lack of consensus with regards to the classification of giant hemangiomas, Adam *et al* described hemangiomas larger than 4 cm as "giant."[3–5,8,11] Studies have suggested that there is a strong correlation between hemangioma diameter and symptoms. Hemangiomas larger than 10 cm are nearly always symptomatic.[1,2]

The most common complaint in symptomatic patients is right upper quadrant pain due to stretching of the Glisson's capsule.[9] However, other signs of gastric outlet obstruction also may be seen as a result of stomach and duodenum compression.[6,8] Portal vein compression may lead to portal vein thrombosis or portal hypertension. Bile duct compression may result in obstructive jaundice or biliary colic.[1] The hepatic vein and inferior vena cava may be compressed, resulting in congestion of various hepatic segments, renal failure, edema in the lower limbs, and clinical signs and symptoms of Budd-Chiari syndrome.[1,4,8] Moreover, hematologic complications, such as Kasabach-Merritt syndrome characterized by thrombocytopenia, micro-angiopathic hemolytic anemia and consumptive coagulopathy, have been reported.[2,7] Although there are many studies about compressive effects of liver hemangiomas, there is only one case report mentioning edema or deep vein thrombosis of the lower limbs due to inferior vena cava compression.[4] However, in this case we detected a problem in

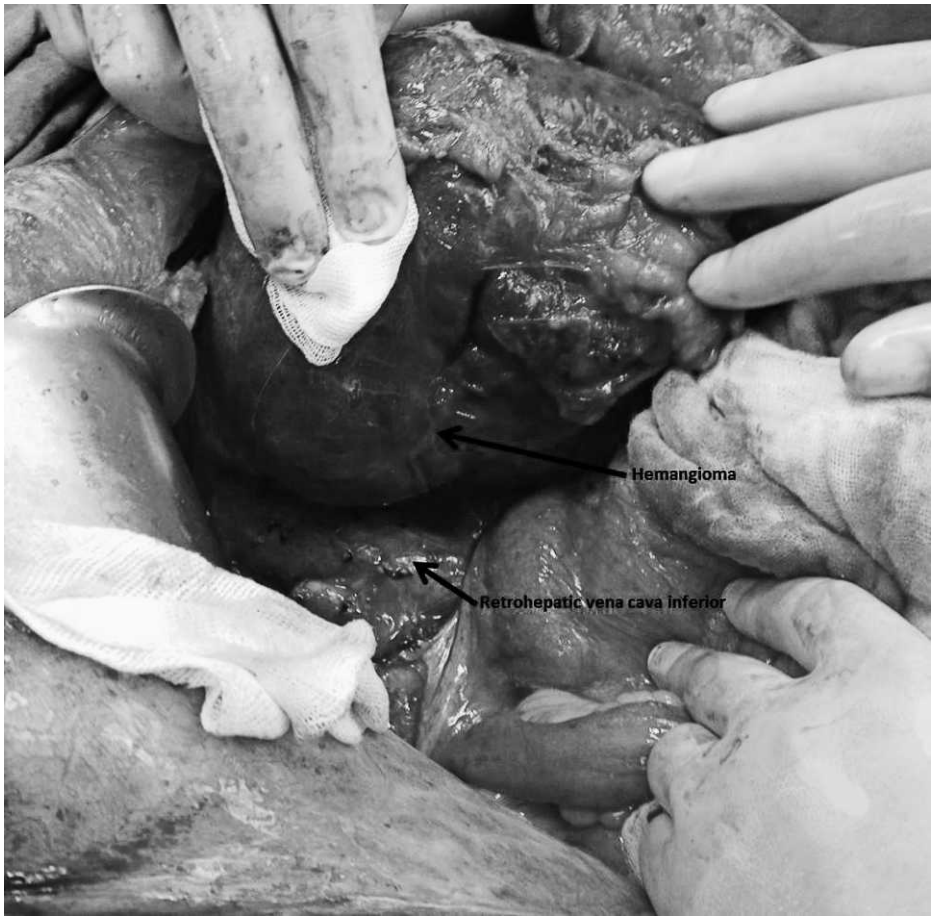

Fig. 2 Dissection of the hemangioma compressing the retrohepatic inferior vena cava from the inferior vena cava by blunt and sharp dissections.

venous return without the development of deep vein thrombosis.

Diagnosis of hepatic hemangioma can be made easily with diagnostic imaging modalities such as US, CT, or magnetic resonance imaging (MRI), either alone or in combination.[10] US is often used in the diagnosis of hemangiomas but because of different imaging patterns, it is difficult to differentiate them from other liver tumors. The clearly demarcated images typical of hemangioma detected by US are usually uniform, sharply demarcated, and hyperechoic. Dynamic CT with contrast may be used in the diagnosis of hemangioma but there is the possibility of confusion with metastatic disease or hepatoma. Small cavernous hemangiomas often demonstrate diffuse contrast enhancement whereas larger hemangiomas characteristically demonstrate peripheral nodular enhancement.[12]

Tc-99m-labelled red blood cell single-photon emission computed tomography (SPECT) has a high level of specificity in the diagnosis of hemangioma.[13] It has been reported that it can be the first choice in the differential diagnosis of hemangioma from other liver lesions. In T1-weighted MRI, hemangiomas are seen in the form of regular, lobular, homogenous, sometimes septated, and hypointense lesions. The hyperintense images created in T2-weighted images are important in the differentiation of hemangioma from other malignancies.[14] Although angiography is the gold standard in the diagnosis of hemangioma, its greatest disadvantage is that it is an invasive procedure.

There is not any algorithm upon which a consensus exists regarding treatment of liver hemangiomas. Close follow-up with no intervention is the best way of management of asymptomatic cases.[1,8,11] Spontaneous or traumatic rupture, intratumoral hemorrhage, consumption coagulopathy, and a rapid growth rate are absolute surgical indications.[15] On the other hand, persistent abdom-

inal pain, obstructive jaundice, portal hypertension, compressive symptoms, tumor size greater than 5 cm, superficial localization (due to increased trauma risk), and suspected malignancy are relative surgical indications.[6,8,10] The most commonly employed surgical treatment modalities are ligation of the hepatic artery, enucleation, resection, and rarely liver transplantation.[2,6,8,15] In addition, transarterial embolization, liver radiotherapy, systemic corticosteroid therapy, and interferon therapy have been reported with positive results.[2,15] To date, there is no consensus on the surgical technique that is best for hepatic hemangioma removal.

Ultimately, we decided to operate because the patient had a hemangioma that was large in size and compressed the inferior vena cava. Furthermore, a standard right hepatectomy was performed since the mass nearly filled the right lobe completely and the demarcation line was close to the hemangioma during the Pringle maneuver. While there are proponents of both the anatomic or nonanatomic resection techniques, many others support the enucleation technique. In our opinion, the most suitable approach to treat hemangiomas with a diameter greater than 10 cm or with multiple lesions is standard or extended hepatectomy accompanied by the Pringle maneuver or total hepatic vascular exclusion.[8,11] However, residual hepatic volume should be taken into account in patients with a planned extended hepatectomy operation.

Acknowledgments

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