Case Report

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A Case Report of Giant Hepatic Hemangioma in A Patient with Budd-Chiari Syndrome

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ABSTRACT

BACKGROUND AND OBJECTIVE: Giant hemangiomas are less likely to be present at birth. This syndrome is caused by obstruction of the portal veins of the liver and requires surgical intervention. Treatment of hepatic hemangiomas varies depending on the symptoms, location and size. In this study, giant hepatic hemangioma in a patient with Budd-Chiari syndrome was reported.

CASE REPORT: The patient is a 57-year-old woman with a giant hemangioma in the liver who has referred to Montaserieh Hospital in Mashhad with symptoms of liver failure, anorexia and early satiety and obstruction of hepatic portal veins. She did not mention any specific drug or food allergies. Except for the heartbeat, all other vital signs were in the normal range. Initial examinations diagnosed giant liver hemangiomas and ascites. After performing the required diagnostic tests, the liver weighing about 10 kg was removed and a liver transplant was performed. After 13 days, the patient was discharged in good general condition.

CONCLUSION: One of the rare indications for liver transplantation is giant hepatic hemangioma, which causes liver dysfunction and Budd-Chiari syndrome. Liver transplantation in these patients improves their life and shows good results.

KEY WORDS: *Hemangioma, Liver, Hepatectomy, Liver Transplant, Budd-Chiari Syndrome, Case Report.*

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Introduction

Liver hemangiomas are present in 3 to 20% of the total population (1, 2) and constitute 73% of all benign liver tumors (3). These masses are the second most common mass in liver tissue after liver metastases (4). The incidence of liver hemangiomas varies in different societies (5) and may occur at any age and gender, but its incidence has been reported more in young and middle-aged women (5, 6). It has an unknown etiology (5), although it is possible that female hormones cause hemangiomas (4).

More than 90% of liver hemangiomas are less than 4 cm in size and are often diagnosed accidentally and do not require surgery (4). However, in cases where the mass is large and symptomatic, segmentectomy, lobectomy and hepatectomy are performed depending on the location and size of the mass. In patients whose liver is completely removed, a liver transplant is required (3). Hemangiomas larger than 5 cm are known as giant hemangiomas and are more common in the posterior part of the right lobe of the liver (7, 8).

The size and location of the tumor in the liver is associated with the incidence of symptoms and complications (2). In general, 85% of hemangiomas are asymptomatic (9). However, in case of bleeding, thrombosis or necrosis becomes symptomatic (4). Bleeding usually occurs after biopsy and trauma and bleeding is a surgical indication in these masses (10). Liver bleeding is a problem that should never be ignored (5, 10) because about 75% of ruptures lead to death from hypovolemic shock (11). In giant hemangiomas, symptoms such as early satiety and fullness in the abdomen, vague pain, thrombosis, anemia, pancytopenia, leukopenia, and coagulation problems may occur (3-5, 12). In addition, obstructive nausea, vomiting, and obstructive jaundice have been reported (9).

Liver hemangiomas are less likely to be present at birth (13). This syndrome is caused by obstruction of the portal veins of the liver and is one of the cases that requires immediate intervention (13, 14). Budd-Chiari syndrome includes a heterogeneous group of different disorders whose occurrence has been reported very rarely in different studies. The main cause of this syndrome in most cases is spontaneous thrombosis of the hepatic veins and usually leads to hypertension in the portal vein (15-18). However, some studies have also reported Budd-Chiari syndrome, which underwent cesarean section during the term pregnancy with severe symptoms of preeclampsia and HELLP syndrome (18). In a meta-analysis examining myeloproliferative neoplasms in coronary artery disease and portal vein thrombosis, myeloproliferative neoplasms were the most common and non-toxic cause of coronary artery disease and portal vein thrombosis (19). New clinical methods with different approaches regarding surgical treatment as well as international guidelines on patient treatment and survival have played an important role. However, the need to conduct clinical studies and report cases in this field and to integrate and combine clinical knowledge and experience to generalize the results of evidence-based clinical studies will be very helpful (20). In this study, a patient with giant hemangioma with Budd-Chiari syndrome was reported..

Case Report

This study was approved by the ethics committee of Kurdistan University of Medical Sciences with the code of IR.MUK.REC.1399.210. The patient is a 57-year-old woman with symptoms of liver failure and giant hemangioma in the right lobe. She referred to Montaserieh transplantation center in Mashhad in July 2019 for liver transplantation. The patient was presented with symptoms of anorexia and early satiety and did not report any specific drug or food allergies. Except for the heart rate (135 beats per minute), all other vital signs were within the normal range and were relatively stable hemodynamically. Giant liver hemangiomas and ascites were diagnosed in initial paraclinical and clinical examinations (Figure 1).

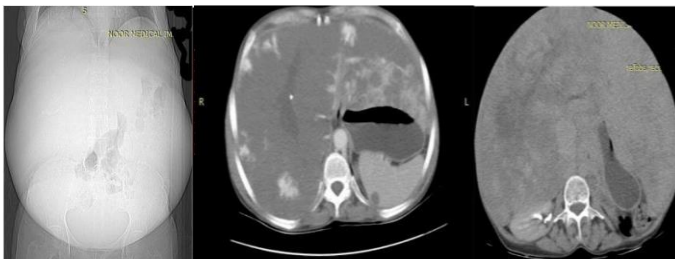


Figure 1. Condition of the liver and adjacent organs in diagnostic tests

In the initial experiments, hemoglobin was 10.1 and hematocrit was 33.3 mg/dL, platelet was 145,000 mm^3, urea was 28 mg/dL and creatinine was 0.6 mg/dL. Total bilirubin was 2.02 mg/dL, direct bilirubin was 0.35 mg/dL, and liver enzymes were normal. The INR was 1.4 and fibrinogen and other laboratory values were normal. The patient's diagnostic and therapeutic measures include CT and MRI findings in the form of

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extensive liver hemangiomas that occupy the entire right and left lobes and contain central calcification foci. The volume of the liver extended to the lower abdomen and other organs appeared normal but were under pressure from the liver and the right branch of the portal vein appeared to be blocked (Figure 1).

On ultrasound, the right lobe of the liver was quite bulky and had heterogeneous parenchymal echo with multiple and scattered foci. A very large hemangioma was seen in the right lobe, which had blocked the right portal vein branch. The density of the left lobe parenchyma was completely heterogeneous, which could indicate infiltrative angiomatosis changes in the left lobe tissue. According to the above findings, the patient is not a good candidate for percutaneous biopsy because the probability of bleeding following biopsy is very high. No significant abnormal findings were observed in other areas of the abdomen and pelvis, and the size of all organs was normal. Therefore, Color Doppler ultrasound of the arteries was performed, which showed compensatory hypertrophy changes in the left lobe. The inferior vena cava was fully compressed but not blocked. The main port vein and left branch were almost normal. The branch of the right port was not recognizable, which could indicate thrombosis or narrowing due to pressure.

After performing preoperative care and necessary examinations, the patient was transferred to the operating room. After general anesthesia, an incision was made under the bilateral ribs (bilateral subcostal). After entering the abdominal cavity, the large liver was observed and then it was fully explored (Figure 2).

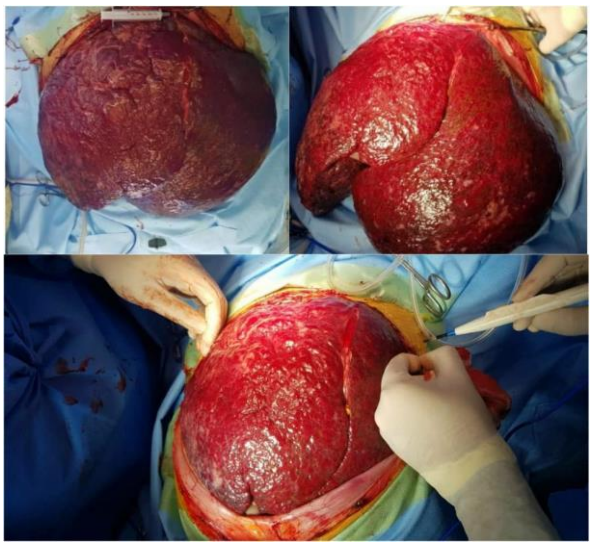


Figure 2. Liver condition after opening the abdominal wall

Then, with standard techniques, the giant liver that weighed approximately 10 kg was removed (Figure 3). After the operation, due to severe bleeding, the presence of diffuse blood coagulation in the patient was confirmed, which was corrected by FFP and then, PT and PTT returned to normal. After the operation, the air was visible under the diaphragm on both sides, and a chest tube was placed to remove it. After follow-ups from different parts of the body, there was no other specific problem and after 13 days, the patient was discharged from the hospital in good general condition.



Figure 3. Weight of the removed liver

Discussion

The findings of this patient showed the very large size of the liver, which weighed 10 kg, indicating Budd-Chiari syndrome. Budd-Chiari syndrome is a complex disease with a wide range of symptoms. In this case, the patient had a relatively stable hemodynamics, despite the relatively high intra-abdominal pressure and the abdominal organs under pressure due to the large and abnormal volume of the liver, which could be due to the patient's long-term adaptation. Hemangioma is the most common benign mass in the liver, which is usually asymptomatic (21). Hemangiomas are more likely to be present in women in their 30s and 50s (9). However, it can occur at any age (22) and is the third most common liver mass in childhood (23).

The macroscopic appearance of hemangiomas may vary due to thrombosis, fibrosis, and calcification, although these masses are usually dark red. Histologically, they are usually composed of a blood-filled space covered by a layer of endothelial cells (24). There are different types of hepatic hemangiomas, but cavernous is the most common (3). Most of these

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masses are present individually in patients but in some cases the presence of different hemangiomas in the liver has been reported (25). Most hemangiomas of the liver are small and asymptomatic, requiring no surgery, and because they are slow to grow (26), follow-up with ultrasound and CT scan every few years is sufficient (6). Of course, there are patients who undergo surgery without symptoms. Studies show that 65% of patients who undergo surgery are symptomatic (24).

Abdominal pain is one of the most common signs and indications for liver hemangioma surgery (6). It is also indicated in cases of advanced abdominal symptoms, spontaneous or traumatic rupture of the mass, Kasabach-Merritt syndrome, rapid enlargement of the mass, and uncertain diagnosis of the surgical mass (3, 4, 6). In some studies, patients underwent surgery for obstructive jaundice and edema of the lower extremities due to hemangiomas (9, 24). Surgery of these masses should be performed carefully due to their proximity to the Portal Vein, Right Hepatic Artery, Right Hepatic Duct, and Right Branch of the Portal Vein to prevent unwanted damage to structures adjacent to the mass and bleeding during surgery (6, 22).

The issue of bleeding prevention is very important (6) and in surgery for hemangiomas larger than 20 cm may also cause hematological complications such as coagulation problems (27). Therefore, considering that methods to reduce bleeding and maintain hemodynamic stability are of great importance and anesthesia examinations and necessary measures to control and replace during surgery are very important, standard and appropriate care should be done before induction of anesthesia and during surgery (28, 29). There is still debate about how to perform surgery in patients with liver hemangiomas (3).

In the case of our patient, the operation was performed under general anesthesia with an incision under the bilateral ribs (bilateral subcostal). After entering the abdominal cavity, complete liver exploration and complete homeostasis were performed. Finally, the liver which was very large and weighed approximately 10 kilogram was removed and finally a liver transplant was performed for the patient. In a study by Zhong et al., a patient with a giant hemangioma in the liver underwent hepatectomy and liver transplantation (30). In Takuya et al.'s study, the patient underwent segmentectomy to treat hepatic hemangiomas (24). In the study of Akbulut et al., a patient with a smaller hemangioma underwent right lobectomy (9). In the study of Okumura et al., a 16 cm hepatic hemangioma was removed laparoscopically (31). In the study of Intaraprasong et al., a patient with bleeding hemangioma in the right and left lobes of the liver underwent hepatectomy and liver transplantation (25). In the study of Mohan et al., the patient underwent embolization with 12 cm hemangioma in the right lobe and there was no particular problem in postoperative evaluations (32).

Regarding the largest reported hemangiomas, Karen et al. reported a hemangioma with a maximum diameter of 21 cm in their study (33). In a study by Zhong et al., The liver weighed 15 kg (30). In the study of Ebina et al., a patient with hemangioma with a maximum diameter of 20 cm in the right lobe was reported (34). Hemangioma caused by Budd-Chiari syndrome is a rare problem that has been reported in a small number of studies (35, 36). In the study of Nuray et al., only 1% of patients with liver masses had Budd-Chiari syndrome (16).

In this study, the very large size of the liver and the incidence of Budd-Chiari syndrome are significant. Liver hemangiomas are usually asymptomatic, but early diagnosis and treatment are essential. In the treatment of liver hemangiomas, attention should be paid to the size, location, and symptoms of the hemangioma. Although the treatment of hemangiomas is mostly non-surgical, in some cases the removal of the hemangioma can solve the patient's problem. A very small percentage of patients with hepatic hemangiomas have symptoms of liver failure and Budd-Chiari syndrome. These people need a liver transplant and the transplant shows good results.

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