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SUCCESSFUL ORTHOTOPIC LIVER TRANSPLANTATION FOR DIFFUSE HEPATIC HEMANGIOMATOSIS IN AN ADULT PATIENT

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Hepatic hemangioma is the most common benign focal lesion of the liver and is usually small and asymptomatic. However, giant hemangiomas may require surgical intervention, while diffuse hepatic hemangiomatosis with multiple giant hemangiomas is exceedingly rare. We report a rare case of diffuse hepatic hemangiomatosis in a 59-year-old patient complicated by portal hypertension, inferior vena cava syndrome, liver failure, and Kasabach–Merritt syndrome, who successfully underwent orthotopic liver transplantation (LT) from a deceased donor. This case demonstrates the malignant course of a relatively benign disease that required LT. Although giant liver hemangiomas are a rare indication for LT, in selected cases where conservative and surgical treatments are ineffective or contraindicated, transplantation may be a safe and effective therapeutic option.

Keywords: hepatic hemangiomatosis, benign liver tumors, liver transplantation, Kasabach–Merritt syndrome, portal hypertension, inferior vena cava syndrome.

INTRODUCTION

Hepatic hemangioma is the most common benign liver tumor of mesodermal origin. It consists of blood-filled vascular cavities lined by a single layer of flattened endothelial cells and is primarily supplied by the portal venous system [1]. Giant hepatic hemangiomas represents an atypical form characterized by a diameter greater than 5 cm and often require surgical intervention [2].

Diffuse hepatic hemangiomatosis (DHH) is a rare condition in which a substantial portion of the liver parenchyma is replaced by multiple vascular lesions. In adults, DHH may be associated with hereditary hemorrhagic telangiectasia (Osler–Weber–Rendu disease), a systemic fibrovascular dysplasia with an autosomal dominant pattern of inheritance. The most common clinical manifestations include recurrent epistaxis and telangiectasias of the skin and mucous membranes, particularly affecting the face, hands, and oral cavity. Involvement of the gastrointestinal tract, lungs, central nervous system, and liver has also been reported [3].

Isolated DHH without extrahepatic manifestations is extremely rare in adults. An even rarer presentation is the coexistence of DHH with a giant hepatic cavernous hemangioma [2, 3]. This combination may be complicated by the development of Kasabach–Merritt syndrome (KMS).

KMS is a rare but potentially life-threatening complication associated with large vascular tumors, including giant hepatic hemangiomas. It is characterized by a form of consumption coagulopathy resulting from activation of intravascular coagulation, leading to severe thrombocytopenia, hypofibrinogenemia, prolonged prothrombin time, and, in advanced cases, disseminated intravascular coagulation (DIC).

The pathogenesis involves sequestration and destruction of platelets and coagulation factors within the large vascular sinuses of the hemangioma. This process results in rapid clinical deterioration, with a high risk of uncontrolled bleeding, multiple organ failure, and death in the absence of timely intervention.

Given its aggressive and life-threatening course, KMS constitutes a major indication for urgent LT. In cases where conservative and minimally invasive approaches are ineffective, LT as the only definitive treatment in cases where conservative and minimally invasive therapies prove ineffective [4, 5].

The literature to date includes only isolated reports of aggressive disease progression necessitating LT [6].

CLINICAL CASE REPORT

Patient K., a 35-year-old male, first noted abdominal distension in 1999. In 2001, he sought medical evaluation, at which time magnetic resonance imaging (MRI) of the abdomen revealed a cystic mass with irregular contours and septations in the left hepatic lobe, measuring 125×80 mm, with compression of the inferior vena cava and portal vein. Additional lesions measuring 35×25 mm

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and 60×60 mm were identified in segments V and VII, respectively. To clarify the nature of these lesions, diagnostic laparoscopy was performed. Intraoperatively, a large tumor measuring approximately 15×15 cm was observed in the left lobe, soft with prominent vascularity. Multiple smaller lesions, ranging from 1.0 to 1.5 cm and visually consistent with hemangiomas, were distributed across both hepatic lobes.

Based on these findings, DHH was diagnosed. The disease remained asymptomatic for several years until 2016. Beginning in 2017, the patient developed ascites and hepatosplenomegaly.

In October 2023, a 59-year-old patient was admitted to the Internal Medicine Department, Shumakov National Medical Research Center of Transplantology and Artificial Organs, presenting with severe weakness, abdominal distension due to tense ascites, abdominal pain, leg edema, and exertional dyspnea. On physical examination, the patient exhibited marked sarcopenia, multiple tophi on the fingers and toes, and tense ascites, with an abdominal circumference of 156 cm.

Laboratory tests revealed signs of liver failure (coagulopathy, hyperbilirubinemia, and hypoalbuminemia), thrombocytopenia, cholestasis, as well as impaired renal function.

Contrast-enhanced computed tomography (CT) of the abdomen demonstrated pronounced hepatomegaly with diffuse parenchymal alterations. The liver had irregular contours and was markedly enlarged, with a craniocaudal dimension of 457 mm in the right lobe and 218 mm in the left lobe. Multiple hypervascular lesions with lacunar contrast enhancement, along with areas of hypoperfusion, were identified in both lobes.

Signs of portal hypertension were present, including dilation of the portal vein (14 mm) and splenic vein (7.5 mm), esophageal varices. and gastric cardia, splenomegaly, and the presence of a splenorenal shunt. In addition, compression of the inferior vena cava at the hepatic and subhepatic levels was noted (Fig. 1).

The patient underwent diagnostic and therapeutic paracentesis, with a total of 45 liters of light-yellow ascitic fluid evacuated, accompanied by albumin replacement. Medical management included choleretic, hypouricemic, anti-inflammatory, gastroprotective, hypoammonemic, and diuretic therapy. Given the lack of response to conservative treatment and the unfavorable prognosis, orthotopic LT was indicated. The patient was subsequently placed on the waiting list for donor organs at Shumakov National Medical Research Center of Transplantology and Artificial Organs, Moscow.

Surgical Procedure

In November 2023, the patient underwent orthotopic LT. Owing to the markedly enlarged liver, a bisubcostal laparotomy with an upper midline extension (Mercedes incision) was performed instead of the conventional J-shaped approach (Fig. 2). Considering the high risk of hemangioma rupture and massive intraoperative bleeding during hepatectomy, a cell salvage system (Cell Saver) was employed from the outset of the procedure. Despite technical challenges related to liver mobilization, a cava-preserving (piggyback) hepatectomy technique was utilized to minimize hemodynamic instability and reduce the risk of postoperative acute kidney injury.

Caval reconstruction was performed using an end-to-side cava-caval anastomosis incorporating the hepatic vein orifices, according to the Tzakis technique. Portal and arterial revascularization were achieved sequentially through end-to-end anastomoses using the standard

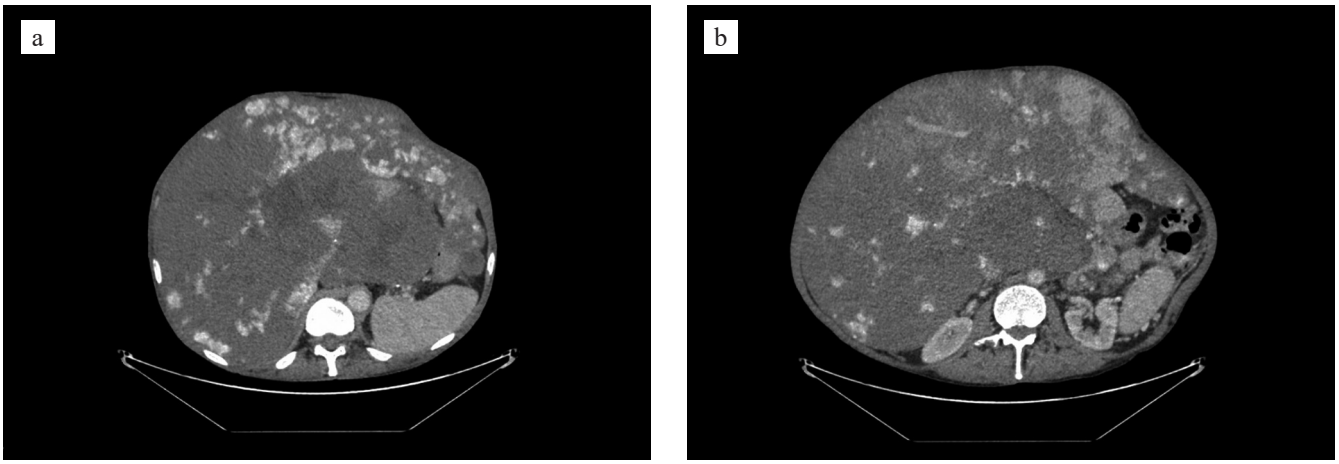


Fig. 1. Contrast-enhanced spiral computed tomography of the abdomen and retroperitoneal space in a patient with diffuse hepatic hemangiomatosis and giant hemangiomas: a, axial section at the liver level during the arterial phase of contrast enhancement: multiple hypodense foci are visible in both lobes of the liver, some of which (>5 cm) have characteristic peripheral nodular enhancement; b, axial section at the liver level during the venous-parenchymatous phase of contrast enhancement: centripetal accumulation of contrast agent in giant hemangiomas is visualized

parachute technique, thereby minimizing the risk of anastomotic kinking or deformation.

Biliary reconstruction was completed via an end-to-end stricture using a 6-0 PDS suture.

The total operative time was 7 hours. Intraoperative blood loss was 1600 mL, with 400 mL reinfused, and transfusion of 2 units of packed red blood cells was required.

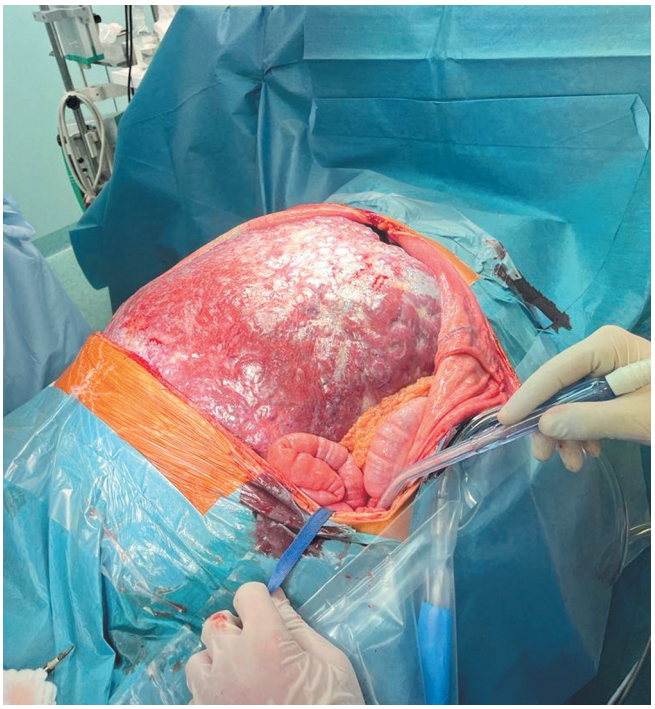


Fig. 2. Subcostal laparotomy supplemented by a midline incision (Mercedes incision)

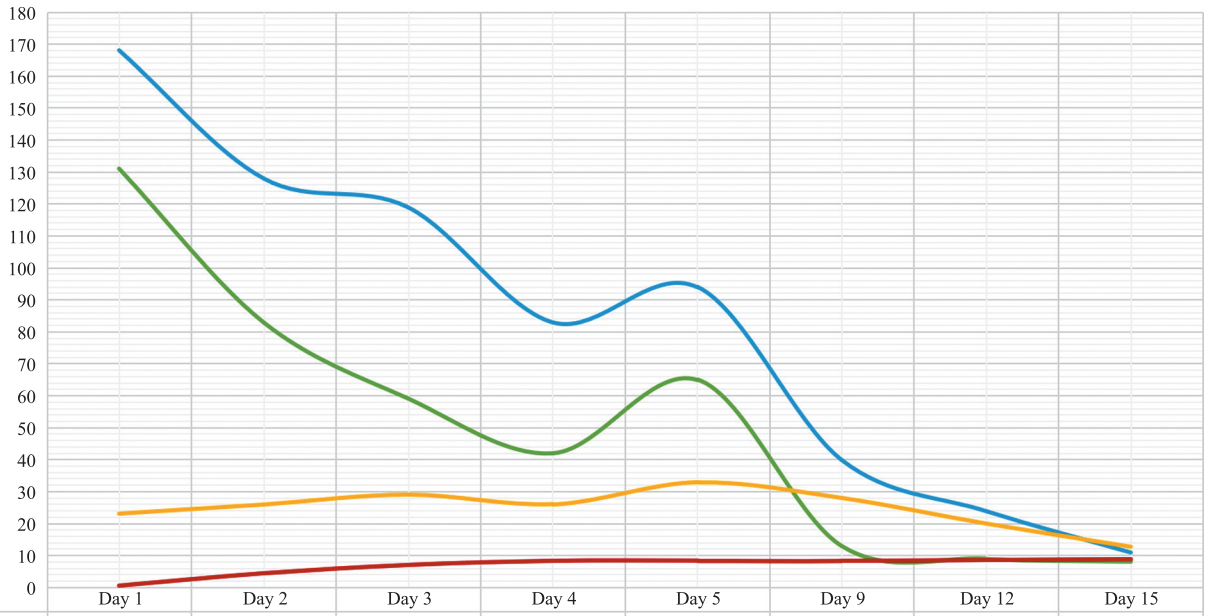
Immunosuppressive Therapy and Postoperative Course

The postoperative period was uneventful, with no complications observed (Fig. 3). The patient was discharged on postoperative day 16 without significant adverse events. The immunosuppressive regimen consisted of extended-release tacrolimus and mycophenolic acid. At one-year follow-up, the patient remains in good clinical condition and continues regular monitoring at Shumakov National Medical Research Center of Transplantology and Artificial Organs. Liver graft function is satisfactory.

DISCUSSION

DHH is a rare disorder characterized by replacement of liver parenchyma by vascular lesions, often associated with hereditary hemorrhagic telangiectasia (Osler–Weber–Rendu), a systemic fibrovascular dysplasia with autosomal dominant inheritance. The most common clinical manifestations include recurrent epistaxis and mucocutaneous telangiectasias involving the face, hands, and mouth. Involvement of the gastrointestinal tract, lungs, central nervous system, and liver has also been reported [3]. Reports of DHH without extrahepatic manifestations in adults are rare.

In the present case, clinical and imaging findings suggest the coexistence of two pathological processes:



	Day 1	Day 2	Day 3	Day 4	Day 5	Day 9	Day 12	Day 15
AST, U/L	131	83	59	42	65	13	9	8.06
ALT, U/L	168	128	119	83	94	40	24	10.93
Bl, µmol/L	23	26	29	26	33	28	20	12.7
Tac, ng/mL	0.54	4.36	7.05	8.3	8.36	8.36	8.51	8.88

Fig. 3. Dynamics of laboratory parameters in the postoperative period. AST, aspartate aminotransferase; ALT, alanine aminotransferase; Bi, total bilirubin; Tac, tacrolimus level

DHH and multiple cavernous hemangiomas (giant focal lesions). This is supported by abdominal MRI performed in 2001, which showed large cystic-appearing lesions in the left lobe (up to 12.5 cm) and right lobe (segment VII, up to 6 cm), exceeding 5 cm in size and causing compression of major vascular structures – features consistent with giant cavernous hemangiomas.

Additionally, diagnostic laparoscopy at that time revealed numerous small hemangiomas (up to 1.5 cm in diameter), indicating concomitant diffuse hemangiomatosis. In subsequent years, contrast-enhanced CT imaging did not clearly delineate the previously identified large lesions, likely due to the diffuse nature of hemangiomatous involvement, which may obscure lesion boundaries and create the impression of a single diffuse process merging with previously identified large foci [7].

The clinical manifestations of combined giant hepatic hemangiomas and diffuse hemangiomatosis are largely similar and primarily result from vascular ectasia and mass effect. Patients commonly present with abdominal pain, while potential complications include intratumoral hemorrhage, rupture with intra-abdominal bleeding, and high-output cardiac failure due to massive arteriovenous shunting. The latter results from abnormal communication between the hepatic arterial and portal venous systems, allowing high-pressure arterial blood to enter the low-pressure portal circulation [8].

In addition, compression of hepatic venous outflow by hemangiomatous masses may lead to Budd–Chiari syndrome. A pronounced intratumoral consumptive coagulopathy can also develop, resulting in Kasabach–Merritt syndrome, which is characterized by the triad of thrombocytopenia, microangiopathic hemolytic anemia, and disseminated intravascular coagulation (DIC) against the backdrop of a giant vascular liver tumor [4–6].

In our case, the patient exhibited features consistent with classical Kasabach–Merritt syndrome (thrombocytopenia, consumption coagulopathy), along with severe ascites due to vascular compression, reflecting a severe disease course [4, 5].

The optimal treatment strategy for DHH has not been clearly defined and depends on the extent of intact liver parenchyma and the severity of clinical manifestations. In asymptomatic patients, even large hemangiomas may be managed conservatively with regular monitoring. However, the development of symptoms or complications necessitates active intervention.

For localized lesions, standard surgical approaches include hepatic resection or enucleation. Minimally invasive techniques, such as transarterial embolization and radiofrequency ablation, may be used to reduce tumor volume and alleviate symptoms. In selected cases, radiation therapy and anti–vascular endothelial growth factor (anti-VEGF) therapy have also been reported.

However, in diffuse hepatic involvement, these treatment modalities are often ineffective. When extensive tumor burden is accompanied by limited functional liver parenchyma, surgical resection becomes either technically unfeasible or associated with prohibitively high risk. In such cases, liver transplantation remains the only definitive treatment option [7, 9].

In the present case, diffuse vascular liver disease with giant hemangiomas led to life-threatening complications, including portal hypertension, inferior vena cava compression syndrome, and Kasabach–Merritt syndrome, ultimately necessitating orthotopic liver transplantation (OLT). This case highlights that, in carefully selected patients with severe benign vascular liver tumors, transplantation can serve as a viable and potentially curative treatment option.

Following OLT, the patient experienced rapid restoration of liver function. Discharge on postoperative day 16 and sustained clinical stability at one-year follow-up further demonstrate the safety and effectiveness of this approach in such complex clinical scenarios.

Overall, this case underscores the successful role of OLT in the management of DHH with giant hemangiomas.

CONCLUSION

DHH presenting with giant hemangiomas is an extremely rare condition that may result in severe, life-threatening complications, including Budd–Chiari syndrome, portal hypertension, and Kasabach–Merritt syndrome. We report a successful case of OLT in a 59-year-old patient with this condition. This case highlights that even histologically benign liver tumors can follow an aggressive clinical course and may ultimately require transplantation as the only life-saving treatment. OLT should be considered a safe and effective treatment option in rare situations where other treatments are not feasible or have failed.

The authors declare no conflict of interest.

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