



## Case Report

# Giant Infantile Hepatic Hemangioma: Case Report and Surgical Technique

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### Abstract

Infantile hepatic hemangioma is the most common liver tumor in children. The most common symptoms are mass in her stomach, anemia and heart failure. According to the findings of the patient, the treatment may vary from the clinical follow-up to liver transplantation. In our study, the details of the surgical technique were presented.

An 11-day-old newborn case with giant hepatic hemangioma causing postnatal respiratory arrest was presented in this study.

Large and symptomatic patients with infantile hepatic hemangiomas who face us with different clinical behaviors are operated. It will be useful to share the surgical technique for these rarely seen cases for surgeons.

**Keywords:** Child; hepatic hemangioma; surgery.

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Infantile hepatic hemangioma (IHH) is the most common benign liver tumor of infants younger than six months.<sup>[1]</sup> Eighty-five percent of the cases are children younger than six months.<sup>[2]</sup> IHH is the third most common liver tumor in children, with a prevalence of 1% and an incidence of 0.4–1.9/1.000.000/year.<sup>[2]</sup> IHH is characterized by its rapid dissemination in the first year of life. IHH grows slower in the following years. It can start after birth and grow rapidly for 9–12 months. Its incidence is twice higher in girls than in boys. This neoplasia is accompanied by skin hemangiomas at a rate of 40–50 percent.<sup>[3]</sup> Apart from the liver, it may also be seen, for instance, in tissues of ovaries, heart, central nervous system, and bone.<sup>[4]</sup> Clinical manifestations cannot be seen in asymptomatic cases, but they may also manifest severe symptoms. In the physical examination, besides the

mass finding in the abdomen, anemia, congestive heart failure and jaundice are the most common findings.<sup>[5]</sup>

In this study, a patient whose IHH caused postpartum respiratory arrest in a newborn who was successfully treated by surgery based on the results of the tests performed after resuscitation of the case is presented.

### Case Report

A 36-week-old 2950 g, 11-day-old baby boy was intubated following cardiopulmonary resuscitation performed upon respiratory arrest at birth. During his physical examination, he was intubated and tachycardic (150 bpm) with a poor general health condition. In the abdominal examination, the mass filling the right abdominal quadrant and descending into the pelvis was detected. In its ultrasonog-

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raphy (USG), in hepatic segments of 4a, 4b, 2,3; structurally hyperechoic patchy areas with punctate echoes and occasionally linear hypoechoic vascular structures with sharp borders and large nodular formations were observed. In addition, it was reported that the segment 4b localization showed an approximately 172x17 mm hyperechoic lesion with lobulated and smooth contours. Computed tomography (CT) demonstrated an extremely enlarged liver with its longest diameter being 182 mm. In the contrast-enhanced CT examination, multiple numbers of disseminated lesions which filled up left lobe segments 2, and 3 which are characterized by patchy areas of nodular formations with sharp borders were seen. However, in the arterial phase, peripheries of these lesions were intensely contrast-enhanced, and in portal and delayed venous phase contrast uptake gradually became more intense towards central regions of these lesions.

Levels of serum enzymes (AST 155 U/L, ALT 45.31, GGT 230.6 U/L,) and tumor markers (AFP >300 IU/ml, CA 125: 112.3 U/ml, CEA 1.47 ng/ml, CA 19-9: 6.43 U/ml.) were as recorded.

The patient was operated with diagnoses of hepatoblastoma, infantile hepatic hemangioma, atypical giant hemangioma, and undersigned. Informed consent form was obtained from the family of the patient.

### Statistical Analysis

After proper application of antibacterial solution and draping of the surgical site, the abdomen was entered with an inverted T incision. Hepatic tumoral mass with approximate dimensions of 15x10x20 cm was observed. The mass at first glance was seen to originate from the left lateral part of the liver and extended into the pelvis. The left lobe of the liver was liberated from its attachments and delivered out of the abdomen. The hilar region was made visible. Following cholecystectomy, the left hepatic artery (left HA) was found and dissected. It was observed that the left HA diameter was very enlarged. The right hepatic artery (right HA) and segment 4 artery were exposed. Segment 4 artery was found to originate from the left HA. The main portal vein was exposed. The left portal branch was dissected. The left portal branch was dissected from the umbilical fissure and the arterial branches of segments 4, 2, and 3 were revealed. The main arterial branches leading to segments 2 and 3 were ligated to protect segment 4. A nearly 50% decrease in the size of the avascularized mass was observed. Then, liver parenchyma was dissected with the help of CUSA (Cavitron ultrasonic surgical aspirator) and crushing clamp applied at a safe distance between the mass border and the parenchyma. Bile canaliculi and vascular structures were closed with a titanium clip. The left hepatic vein was dissected away from the area where it entered inferior vena

cava over the vascular clamp and closed in two layers with 5/0 non-absorbable suture. The left lateral bile duct was exposed and ligated. The mass completely separated from the liver was taken out of the abdomen. After bleeding control, an absorbent drain was placed in the hilar region and the abdominal layers were closed.

After the operation, the patient was intubated and taken to the neonatal intensive care unit. For five days, his vital signs remained stable, so he was weaned from the mechanical ventilator. Any pathological finding was not detected in the control USG, so the absorbent drain in the abdomen was removed. The dressing of the surgical wound and follow-up of the patient was continued for ten days. Since any complication did not develop, the patient was discharged and followed up in the outpatient clinic.

The histopathological examination of the removed mass revealed a 10.5x7 cm mass lesion containing stroma-rich and diffusely sinusoidal vascular tissue compatible with infantile hepatic hemangioma which can be clearly differentiated from liver tissue.

### Discussion

Infantile liver hemangiomas are the most common benign liver tumors before the age of two. It is more common in girls. The most common finding is hepatomegaly. Apart from this, jaundice, cutaneous hemangiomas, coagulopathy and heart failure are seen.<sup>[6]</sup> Our case was an 11-day-old baby boy, and because of his size, he immediately showed symptoms after birth and hepatomegaly was noticed at the first examination. Such cases may be part of Kasabach-Merritt syndrome,<sup>[7]</sup> but our case did not have skin hemangioma and thrombocytopenia.

USG, CT and magnetic resonance are being used for radiological diagnosis of IHH. Biopsy, on the other hand, is not usually performed since it carries a risk of bleeding.<sup>[8]</sup> In our case, the diagnosis was first made with USG and then supported with contrast CT. Peripheral contrast enhancement of the tumor in the arterial phase in the CT of patients with IHH and the progression of the contrast enhancement towards the center at a later stage are characteristic features for the diagnosis of IHH.<sup>[1]</sup> In our case, a tumor that had up-taken contrast material intensely in the arterial phase, and in the portal and delayed venous phase, it progressively demonstrated greater hyperintensity towards the central region (Fig. 1).

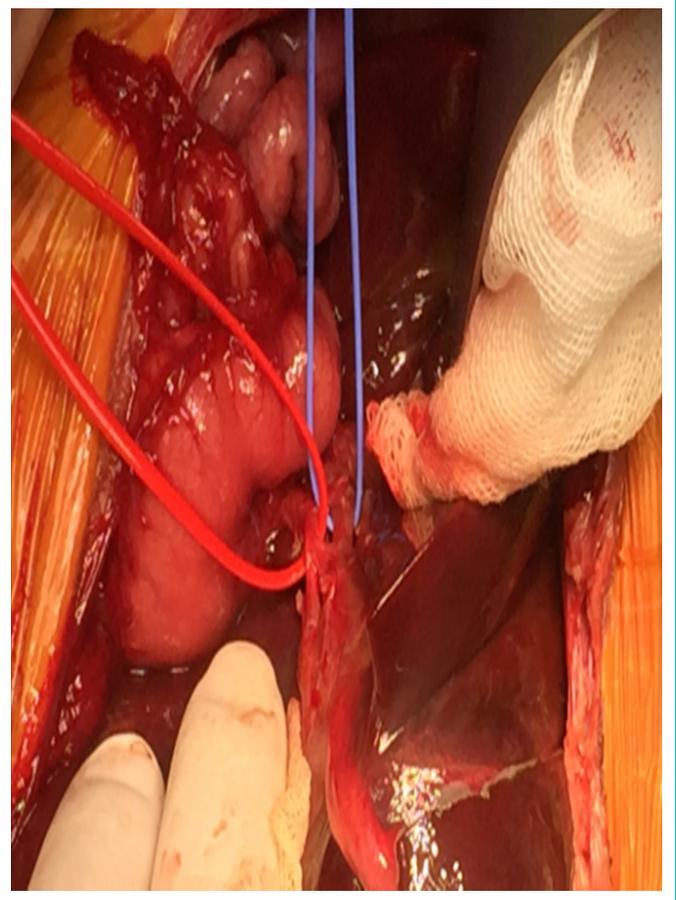
Tumor sizes vary in IHH, and it has been reported in the literature that tumors reaching up to 20 cm from millimetric sizes.<sup>[9]</sup> In our case, it was nearly 20 cm in size during the operation. In this respect, it is one of the large tumors reported in the literature.



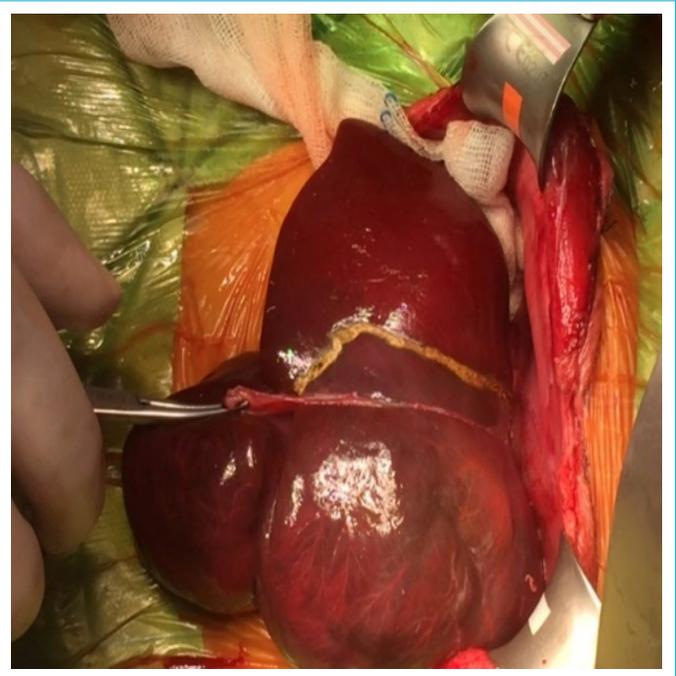
**Figure 1.** Contrast-enhanced abdominal tomography.

Steroid therapy is the priority in symptomatic cases with infantile liver hemangiomas. Corticosteroids, interferon-alpha2a or propranolol can be used to shrink the vascular bed of the lesion. However, due to studies reporting the side effects of these drugs and variable success rates, the risk/benefit ratio could not be determined well<sup>[10]</sup> Surgical treatment may be preferred primarily if there is a large, single hepatic lobe lesion causing life-threatening symptoms.<sup>[11]</sup> It is recommended in patients with a low probability of spontaneous regression or in cases where somehow, the possibility of malignancy could not be excluded radiologically/clinically.<sup>[10]</sup> In our patient, depending on the size of the mass at birth, the patient stopped breathing and was intubated. Given that the mass is stealing a large amount of blood from the systemic circulation, due to its large size, it caused us to think primarily of surgical treatment.

During the surgery of IHH, any tumor residue should not be left behind as much as possible, and liver circulation should not be impaired.<sup>[12]</sup> In this respect, after the liver is liberated, the segments where the tumor is located must be determined. Recognition of the hepatic artery that fed the segment containing the tumor will prevent unnecessary dissection.<sup>[3]</sup> In our case, the tumor seemed to cover segments 2-3 and 4 at first glance. However, after dissection of the right and left hepatic arteries, it was understood that the tumor invaded only segments 2 and 3, and segment 4 can be saved. When the arterial branches of the segments 2 and 3 separated from the left hepatic artery were ligated, the tumor mass shrank very much (Fig. 2). After the tu-



**Figure 2.** Suspension, and then Identification of vessels supplying the tumor.



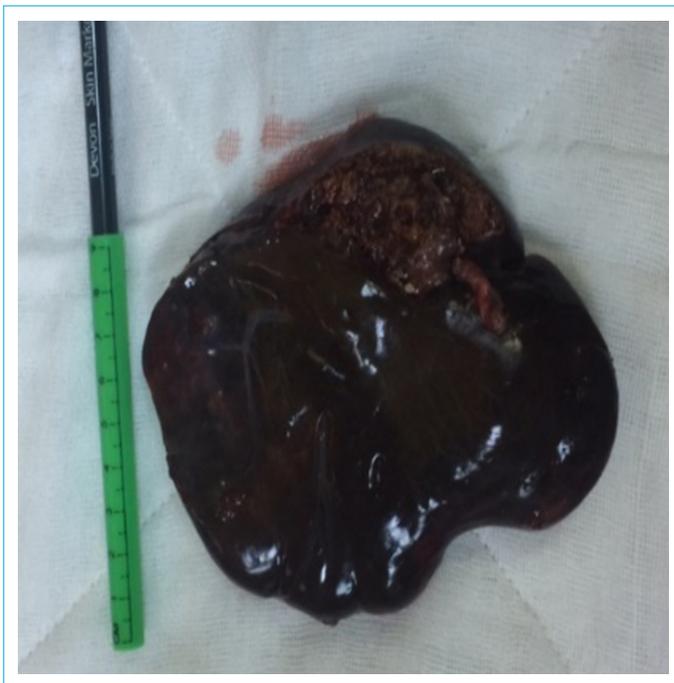
**Figure 3.** Determination of the dissection line between the tumor and the border of the parenchyma.

mor vessels were ligated, we proceed with the stage of parenchymal dissection (Fig. 3).

In the literature, for the dissection of liver parenchyma, techniques, such as finger dissection, cutter dissection, Kelly technique (clamping technique), hydro-jet and radiofrequency dissection and sealing, have been reported. CUSA (Cavitron Ultrasonic Surgical Aspirator, Tyco healthcare, Mansfield, MA) is a device that uses ultrasonic energy to dissect parenchymal tissue. It dissects by protecting the liver parenchyma and reveals bile canaliculi and vascular structures without damaging them and allows them to be clipped.<sup>[13]</sup> The need for postoperative blood transfusion is lower, and the risk of infection and bile fistula is lower in liver parenchymal dissection combined with CUSA and clamping technique.<sup>[12-14]</sup> In our case, we made a parenchymal dissection using these two methods. Our patient did not need a blood transfusion and did not develop an intra-abdominal infection after surgery.

Pathologically, the macroscopic image changes depending on the size of the tumor. Pressure-related pseudocapsule may be present, especially in large tumors, necrotic changes and bleeding may occur due to infarction in the tumor center.<sup>[16]</sup>

In histological examination, single-layered bulging endothelial cells and anastomotic vascular structures are observed. Intervascular spaces usually consist of small-diameter capillaries and sinusoids. In some areas, cavernous type vascular structures are also seen.<sup>[12,14]</sup> In our case, a pseudocapsule was formed due to severe pressure, and the



**Figure 4.** Appearance of the extracted tumor.

tissue with sinusoid vascular features rich in stroma could be distinguished. Therefore, there was no need for an immunohistochemical study (Fig. 4).

As a result, different treatment options for IHH should be discussed due to their different sizes and clinical behaviors. Surgical treatment of the giant infantile hepatic hemangiomas can be decided upon vein in the neonatal period. It is important to present the technical details regarding the surgery of these rare cases in the literature.

#### Disclosures

**Informed Consent:** Written informed consent was obtained from the parents of the patient for the publication of the case report and the accompanying images.

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**Conflict of Interest:** None declared.

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