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Giant Congenital Liver Hemangioma: Kasabach Merritt Syndrome

Carlos Cortés-Caballero, MD,[1] Margarita Rosa Mogollón-Zehr,[2] Silvia Nathalia Vera-Campos[2]

1 Surgeon, graduated from the University of Antioquia; Pathologist, graduated from Western Pennsylvania Hospital in Pittsburg, PA USA; Centro clinico Bucaramanga in Bucaramanga, Colombia
2 Eighth semester medical student at the Industrial University of Santander in Bucaramanga, Colombia

Correspondence: Carlos Cortés-Caballero: cccaballero@gmail.com

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Abstract
Hemangiomas are the most frequent proliferations in children, and liver hemangiomas are the most common among those in the viscera. Early diagnosis to focus treatment is important. We present the case of a neonate with compromised breathing secondary to the effect of a giant mass that echographically appeared to be a hepatic hemangioma. Surgical resection and subsequent pathology confirmed the diagnosis.

Keywords
Hepatic hemangioma, Kasabach Merrit syndrome, giant hemangioma.

INTRODUCTION

Hemangiomas are the most frequent liver neoplasms in children. About 70% present at birth, and they represent 10% of pediatric tumors. They can compromise any organ, although they are more common in the skin, liver and brain. (1) Their incidence is higher in women than in men at a ratio of 3:1. (2)

Hemangiomas are characterized by a proliferation of endothelial cells without atypia and angiogenesis which leads to rapid growth of this lesion. They are usually small and asymptomatic, but there have been cases of giant hemangiomas (greater than 5 cm) which can result in complications such as heart failure, hypothyroidism, abdominal compartment syndrome and Kasabach syndrome -Merritt which has a high mortality rate in neonates. (3)

Diagnosis is generally incidental through ultrasound findings, although it can be confirmed histologically using glucose-1 transporter markers (GLUT1), factor VIII, CD31 and CD38 which help distinguish it from other vascular pathologies. (2, 4)

CLINICAL CASE

The patient was born prematurely at 33.3 weeks of gestation. The mother was a 29-year-old woman with a history of controlled gestational diabetes and chorioamnionitis. We observed a female neonate weighing 2565 g who was 45 cm long and had bradycardia and respiratory compromise secondary to restriction by a mass. She was admitted into the intensive care unit (ICU) where she underwent orotracheal intubation. During the physical examination, her abdomen was distended and a mass of hard consistency was palpable on the flank and in the left hypochondrium but without pain or signs of peritoneal irritation. The abdominal perimeter was 35 cm which was large for her chronological age. She had petechiae and her skin and eyes were jaundiced.

Abdominal ultrasound showed a mass of heterogeneous echogenicity dependent on the left hepatic lobed. It was difficult to quantify due to its size, but it extended to the pelvis and displaced neighboring structures. The mass had internal vascularization, and there were cystic areas without calcifications. Diagnostic possibilities included

hepatic hemangioma and mesenchymal hamartoma with a less likely possibility of a hepatoblastoma. A chest x-ray suggested cardiomegaly.

On the third day, paraclinical tests showed total bilirubin of 8.9 mg/dL, indirect bilirubin of 7.9 mg/dL, ALAT of 21 U/L, ASAT of 213 U/L, prolonged prothrombin time, platelets of 56,000/mm^3, hemoglobin (Hb) of 10.8 g/dL, and hematocrit (Hct) of 34.4%. Her evolution showed progressive decreases of these values. Her alpha-fetoprotein (AFP) was 1,000 IU/mL. The diagnostic impression from computed tomography (CT) was that the patient had a congenital hepatic hemangioma, so it was decided to perform a left lobectomy.

On day 14 of the preoperative period, total bilirubin was 0.7 mg/dL and direct bilirubin was 0.56 mg/dL. Liver function tests and prothrombin time were within normal ranges. Ten days after surgery, the patient developed massive pulmonary hemorrhaging that was difficult to manage. The diagnosis was disseminated intravascular coagulation (DIC). Rescue treatment was started with adrenaline, tranexamic acid, and cryoprecipitate and platelet transfusion. Results were satisfactory. In addition, hyperbilirubinemia was found at the expense of direct bilirubin, with total bilirubin values of 17 mg/dL and direct bilirubin of 12 mg/dL. Subsequently, the patient was referred to a pediatric cardiology clinic for suspected persistent ductus arteriosus. It was decided that she did not require surgery, and she was discharged at 45 days under optimal conditions.

The resection piece was sent to pathology. Histopathological findings included a firm, ovoid mass weighing 268 g, measuring 10.5 x 6.5 x 5 cm. It had a smooth, shiny external surface, and the implantation base was bloody and measured 4 x 3 cm. It was composed of red tissue surrounded by a pale band of variable thickness that reached 0.3 cm thick at its maximum point (Figure 1).

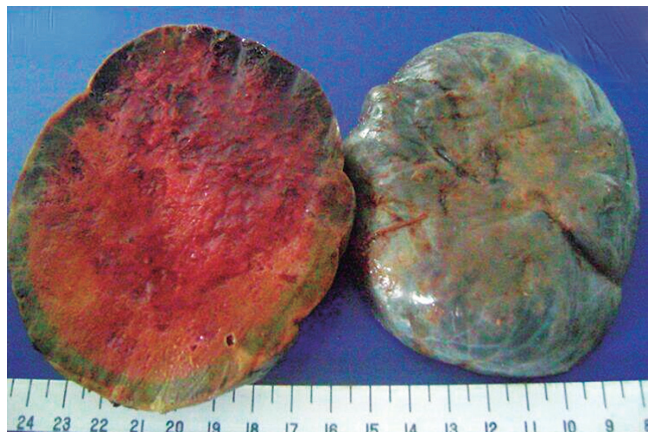

Figure 1. Hepatic hemangioma composed of internal red tissue with smooth outer surface

Microscopically, vascular spaces of varying sizes predominate. There were capillaries and cystic spaces covered by a monomorphic endothelium. The spaces contained red blood cells and histiocytes with pigment. A fibrous capsule of variable thickness could be seen in the peripheral area. There were bile ducts in some fields and hepatocytes and capillaries in others. A diagnosis of capillary and cavernous mixed hepatic hemangioma was made (Figure 2).

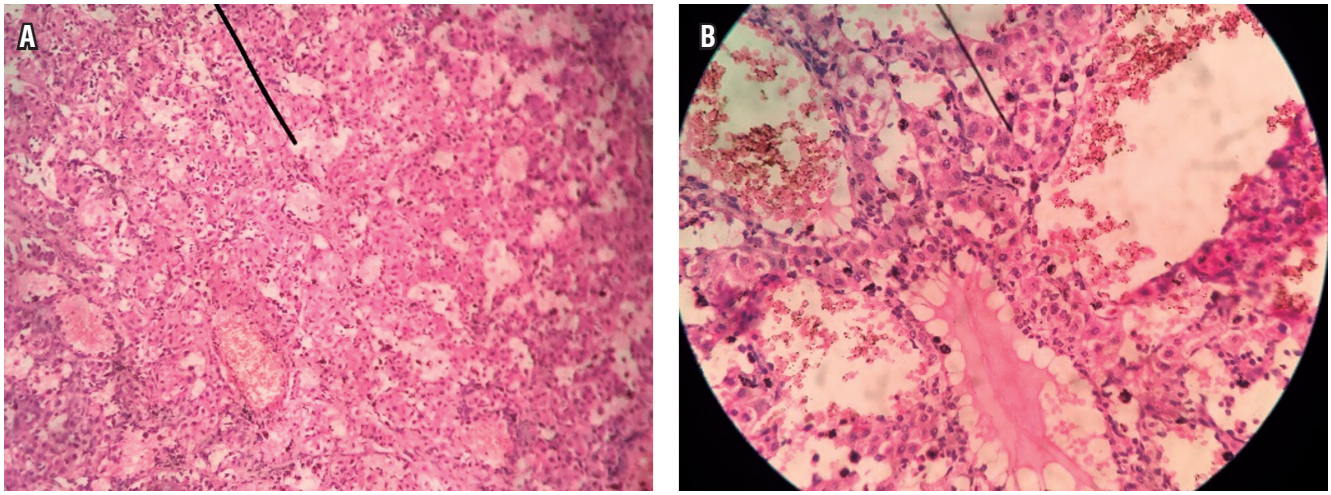

Figure 2A. Microscopic study (HE, 20x) showing vascular spaces covered by an endothelium without atypia. **2B.** (HE, 40x) red blood cells and histiocytes with pigment.

DISCUSSION

Hepatic hemangiomas are benign neoplasms of mesenchymal origin which are usually solitary. According to various dermatological and surgical studies, there are two types of hemangiomas: vascular malformations and vascular neoplasms. (5) They grow rapidly in the first years of life and stabilize at approximately one year of age. Seventy-five percent of cases tend to involute completely between the ages of 5 and 7 years old so that management is based on control of evolution. (6) They have been most frequently described in the left hepatic lobe, as was the case with our patient. (3)

Their etiology remains unknown although there is an association between size and levels of estrogen/progesterone. They have been observed to grow during pregnancy and during use of oral contraceptives and hormone replacement therapies. (7) This would explain the higher prevalence in females. They can also generate hepatic peliosis. (8) In our case, no family history or use of these drugs was found, and there was no relationship with gestational diabetes and chorioamnionitis suffered by the mother during pregnancy.

Among the differential diagnoses are the abdominal tumors typical of childhood, but AFP was normal values for the patient's age, so the possibility of a hepatoblastoma was minimal. This marker should be interpreted according to the age group, and in newborns very high levels are considered normal.

Complications can include thrombosis within the mass, ruptures, uncertain diagnosis, hepatic dysfunction or other abdominal organ dysfunction related to the mass, coagulopathy syndrome also known as Kasabach-Merritt syndrome, and death. In the majority of cases death is due to heart failure. This occurs in approximately 50% of patients with hepatic hemangiomas and is secondary to the arteriovenous shunt of large lesions. (6) In our patient there were clinical and laboratory findings suggestive of hemolytic anemia and severe thrombocytopenia accompanied by consumption coagulopathy. These findings indicated Kasabach-Merritt syndrome which occurs by platelet activation inside of large hemangiomas. (3) Preoperative jaundice with hemolytic characteristics was also observed. This is found in up to 35% of cases of hepatic hemangiomas in children. (6)

Treatment alternatives include the use of corticosteroids or agents with a marked anti-angiogenic effect such as interferon alfa, cyclophosphamide, vincristine and actinomycin D. (9) Treatment using Bevacizumab, a monoclonal antibody capable of inhibiting endothelial growth factor, has been described, but its effectiveness has not been confirmed. There are a small number of cases for which surgical intervention is considered due to the risk of complications which usually occur around the sixth week of life. Surgical management includes segmental resections, lobectomy or removal of the hemangioma by open surgery or laparoscopy. (10) If the need for surgery arises, the procedure of choice is removal. (3) In this case, surgical intervention was the first choice, due to the respiratory compromise secondary to the mechanical effect of the mass which was twice the weight of the liver (250 g vs. 110 g).

It is important to consider hemorrhaging, coagulopathies, infections, biliary fistulas, ascites and liver failure as possible complications during the postoperative period. The direct hyperbilirubinemia of our patient was attributed to liver failure. On the other hand, DIC could be considered a postoperative complication since the patient did not present this anomaly in the preoperative period. This rules out the possibility that it was the product of the Kasabach-Merritt syndrome which commonly is the case. The main consequences of CID are the consumption or loss of hemostatic proteins and platelets in the perioperative period. This causes hemorrhaging and thrombotic obstructions in the microcirculation and compromises the function of various organs. In this case, it led to massive pulmonary hemorrhaging.

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