CASE REPORT

Hepatic hemangioma


Abstract

Objective: to describe a case of a congenital hepatic hemangioma treated with surgery.

Methods: we report the case of a 6-day old male newborn who presented a giant hepatic hemangioma and its evolution.

Results: the child developed hemodynamic instability secondary to consumption coagulopathy and respiratory failure. The image studies were inconclusive. The patient was submitted to surgery with complete resection of the tumor. Anatomopathological study confirmed hemangioma. The child was dismissed from hospital after 15 days, and is presently asymptomatic.

Conclusions: hepatic hemangiomomas should be treated conservatively, with surgery reserved for intractable cardiac failure and/or refractory consumption coagulopathy.


Introduction

Hemangioma is the hepatic tumor most frequently seen during the 1st year of life, and it is the most common benign neoplasia of the liver in childhood. Hepatic hemangioma is rarely seen in children above 3 years of age. There is practically no predominance in relation to sex or race.1

Hepatic hemangioma grows fast in the first years of life, and becomes stable around the age of 1; it recedes spontaneously with time. The period of involution varies. Seventy-five percent of the lesions tend to heal completely around the age from 5 to 7 years. The most significant complications, however, occur around the 6th week of life. Death, in most cases, is due to cardiac failure.2

The most usual clinical finding in infants with hepatic hemangioma is the hepatomegaly, which is often associated with a cutaneous hemangioma and with a cardiac failure secondary to arteriovenous shunting.1-3

Vascular lesions are divided in hemangiomas and vascular malformations. The main difference between them is that, in hemangiomas, proliferation of endothelial cells and angiogenesis are observed, and lead to a fast growth of these lesions.4,5
Usually, advanced imaging exams, such as computed tomography, magnetic resonance imaging, and ultrasonography are sufficient for diagnosing hemangiomas.1,6-10

A conservative approach to the disease is the treatment of choice in most cases; however, surgical intervention may be eventually needed.1,2,6

We describe the case of a neonate with a voluminous hepatic mass, diagnostic tests, the patient’s evolution and treatment, in addition to a review of the literature.

Case report

Newborn, 6-day old, male, white. His 37-year old mother was hypertensive. She received prenatal care since the first gestational trimester, presenting two normal ultrasonographic exams. The child was born by a C-section due to the presence of meconium in amniotic fluid. Apgar 7 at the first minute and 7 at the fifth, 3,380 g of weight, gestational age of 38 weeks. The patient presented abdominal distension and a tumor at the right hypochondrium on the first days of life. He was admitted in good general status, eupneic, with jaundice in Kramer's zone III, globose abdomen with evident collateral circulation, and palpable mass at the right hypochondrium.

Laboratory exams showed HB at 11 mg/%; HT at 33mg%; leukocytes at 6,500/mm³; platelets at 36,000/mm³; total bilirubin at 10.29 mg%; and indirect bilirubin 8.6mg%.

Abdominal computed tomography showed a large heterogeneous hepatic mass (8.0 x 6.5 x 9.5 cm) occupying the left lobe and part of the right lobe, with a heterogeneous concentration of venous contrast. The scintigraphy with labeled red blood cells was negative for hemangioma.

The patient evolved with exacerbation of the abdominal distension and restrictive respiratory distress, as well as decrease in the levels of hemoglobin (from 9.4 to 8.0 mg%) and plateletopenia (20,000/mm³).

Due to the fast development of the hepatic lesion, with progressive increase in the abdominal volume, evident respiratory failure, and absence of diagnosis through the imaging exams, we decided to perform a surgical intervention.

Laparotomy with left hepatectomy and exeresis of the lesion were performed.

The macroscopic anatomopathological study revealed a right hepatic lobe with 10.0 x 4.0 x 8.5 cm and weighting 220 g. The sections showed an intraparenchymal nodular lesion, port-wine hued, with spongy aspect, surrounded by fibrous trabeculae. The lesion measured 7 cm on its longest axis. Microscopic examination showed badly delimited lesion, with irregular outlines and compressing the adjacent hepatic parenchyma. The lesion was characterized in its central area by large vascular spaces, with thin walls and endothelial covering, filled with blood (Figure 1). The peripheral area of the lesion showed a significant proliferation of biliary ducts in sometimes myxoid, sometimes fibrous stroma, with several foci of extramedullary hematopoiesis. Immunohistochemical study with endothelial cells markers (CD34 and factor VIII) revealed multiple capillaries and vessels with an hamartomatous aspect in the solid peripheral areas, surrounded by the proliferated biliary ducts (Figure 2).

Figure 1 - Lesion characterized by large spaces with thin walls and extramedullary hematopoiesis foci (HE x 400)

The identification of proliferating cells with Ki-67 clone MIB-1 demonstrated extended positivity in both vascular and mesenchymal elements, and also in the proliferated biliary ducts, which indicates a high fraction of cellular proliferation. The diagnosis of hemangioma was established due to the high amount of vascular elements and their proliferative character.

Figure 2 - The immunohistochemical positivity for CD34 characterizes the nature of the vascular lesion (CD34 x 100)
In the postoperative period, the patient presented a septic status with positive response to broad-spectrum antibiotic therapy. Jaundice and the symptoms of cardiac failure were attenuated, and the child was dismissed at 38 days of life.

In the routine clinical follow-up, at 3 months of life, we observed an hemangioma-like nodule on the right thoracic wall. We chose an expecting conduct.

Discussion

Although vascular tumors of childhood are considered benign lesions, often presenting spontaneous resolution, hepatic hemangiomas may be found in large extensions, with a significant hemodynamic disorder. Cardiac failure occurs in around 50% of the patients with liver hemangiomas, and it is secondary to the arteriovenous shunt of large lesions. This complication leads 70% of the cases to death when the disease is not properly treated. The literature describes that the younger the child is, the more severe the cardiac symptoms will be.

Another severe symptom in these patients is the Kasabach-Merritt syndrome, which consists of the trap of platelets by the lesion, microangioplastic hemolysis, thrombosis, and consumption coagulopathy. Madan et al. reported a 75% incidence of Kasabach-Merritt syndrome in 16 children with hemangioendothelioma. The mortality rate was set between 30% and 40% for patients presenting this syndrome. Clinical and laboratory findings of anemia, plateletopenia, and coagulopathy, which were observed in the case here reported, characterize the presence of Kasabach-Merritt syndrome. Jaundice, which was also observed in our patient, is seen in approximately 35% of the cases of hepatic hemangioma in children.

Cutaneous hemangioma is present in 40% of the patients with liver hemangioma. Our patient did not present cutaneous hemangioma at the time of diagnosis; however, 3 months later, we observed a lesion on the right thoracic wall, which was diagnosed as cutaneous hemangioma.

In the majority of cases, advanced imaging techniques are sufficient for the establishment of a diagnosis of hepatic hemangioma. Computed tomography, especially with contrast, is highly sensitive in locating and delimiting vascular lesions. The computed tomography and the ultrasonography-Doppler are not only capable of showing, but also of quantifying intrahepatic shunts. The indications for arteriography have increased significantly over the last years. The study of the Tc-99m-labeled red blood cells may also be used, especially when the mentioned exams leave some doubts about the diagnosis. This exam presents an up to 78% of sensitivity in tumors larger than 2 cm in diameter. However, regardless of the high sensitivity of noninvasive diagnostic methods, vascular tumors with high rates of cellular proliferation may produce high density images, simulating a solid tumor. In these cases, surgical procedures are required for establishing the diagnosis.

Treatment must be directed to the correction of systemic manifestations of the tumor. Heart failure has been treated with digitalis and diuretics. Steroids have been used in order to try to reduce the immature hemangioma tissue, as first proposed by Touloukian et al. Several centers have used steroids as a first-line treatment, with a response rate around 45% in symptomatic cases. Chemotherapeutic drugs, such as cyclophosphamide, have also been used associated or not with steroids. However, the few reports about the use of these drugs do not present positive results.

Due to its action over angiogenesis, inhibiting the locomotion and proliferation of endothelial cells, the alpha-2 interferon has demonstrated to be efficient in reducing large hemangiomas. Using alpha-2a interferon in 20 patients with hemangioma, Folkman observed a reduction of 18 tumors. Nowadays, alpha-2a interferon is considered the treatment of choice for cases of Kasabach-Merritt syndrome.

The association of interferon and corticoid did not present advantages, and increased the toxic effects on the patient.

Radiotherapy is also used in some centers; however, its indication has significantly decreased due to low response and to the damaging effects to the liver and adjacent organs.

Surgical resection of hepatic hemangiomas is indicated when the conservative procedures are not capable of controlling the disease, and in cases in which the diagnosis is uncertain. Devenport et al. indicate early surgery in cases of cardiac failure secondary to arteriovenous shunt. Surgical treatment consists of lobe resection in cases of focal tumors confined to an area; in cases of bipolar tumor, a ligation of the hepatic artery is indicated. Embolization has also been performed using gel-foam, polyvinyl alcohol and steel coils, with good results; however, a renal failure secondary to the passage of embolic agents may occur. Surgery was indicated in our case due to the nonconfirmation of our previous diagnosis, and also based on the unfavorable clinical development, with consumption coagulopathy and signs of hepatic, cardiac, and respiratory restrictive failure with the growth of the tumor, which represented a threat to our patient’s life.

The Kasabach-Merritt syndrome was described as the association between capillary hemangioma and plateletopenia. Nevertheless, recently, Enjolras et al. discuss that the vascular lesions seen in the Kasabach-Merritt syndrome are not true hemangiomas. Vascular anomalies described in this syndrome may present different aspects, with a varied spectrum of histological presentations. In our case, the anatomopathological study did not confirm a classical capillary hemangioma of childhood.
References

Correspondence:
Dra. Cecília M.L. da Costa
Departamento de Pediatria
Rua Prof. Antônio Prudente 211
CEP 01509-000 – São Paulo, SP, Brazil
Fax: +55 11 242.5088
E-mail: cmldcosta@sti.com.br