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Case Report

Severe Infantilehepatic Hemangioma or Hepatoblastoma: About a Rare Case and Literature Review

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Received: May 3rd, 2021; **Accepted:** May 8th, 2021; **Published:** May 14th, 2021

Citation: Toughza J, Agadr A , Nejjari M, Al-Ammari I. Severe infantilehepatic hemangioma or hepatoblastoma : About a rare case and literature review. Biom Case Rep Open A Open J. 2021; 2(1): 63-65. doi: 10.33169/biomcase.BACROAOJ-2-117

ABSTRACT

Childhood liver tumors are rare, but two-thirds are of malignant origin. Basically, before the age of 3, the main diagnoses are Hepatoblastoma (HB) and hemangioma. Infantile Hepatic Hemangioma (HHI) is a subclass of infantile hemangiomas that are the most common self-limiting benign tumors of early childhood. Only the diffuse forms or associated with congestive heart failure can be fatal. Careful clinical examination associated with biological and radiological elements does not always allow a diagnosis to be made. The biopsy is still performed frequently for diagnostic or prognostic purposes. Because of the diagnostic difficulties with HB, and the severity of some forms of HHI, we report the case of a newborn with diffuse HHI, observed at the Cheikh Khalifa International University Hospital in Casablanca. This was a newborn male in whom symptomatology of the disease consisted of increased abdominal volume, hepatomegaly and signs of respiratory struggle. The ultrasound described the existence of a multi-hetero-nodular hepatomegaly, and the CT scan a liver increased in size, seat of a large mass, poorly limited with bumpy contours of predominant tissue density with individualization of a central area of fluid density necrosis, containing calcifications. The AFP rate was high. An ultrasound-guided percutaneous biopsy confirmed the diagnosis of HHI. The course was very rapid and fatal in this patient with diffuse HHI with congestive heart failure.

Keywords: *Diffuse Hepatic Hemangioma; Hepatoblastoma, Infantile; AFP.*

INTRODUCTION

Childhood hepatic tumors are rare and represent 2% of tumors. The diagnostic range varies with age, but two-thirds of liver tumors are malignant. Schematically before the age of 3 years the main diagnoses are hepatoblastoma (HB) and hemangioma, while after the age of 3 years, Hepatocarcinoma (HC), sarcomas, and tumors should be considered. Benign such as focal nodular hyperplasia (UFH) and adenoma.[1] The clinical and biological elements with the assay of Alpha-Fetoprotein (AFP) and ß-gonadotropicchorionic hormone (ß-HCG) as well as imaging will play an important role in the orientation of the diagnosis, the assessment of extension, and follow-up under treatment of these tumors.[2] AFP is a serum globulin found in the fetus and synthesized by the fetal liver and by the yolk sac. It appears in the serum of the fetus from 6 weeks. The rate is high at birth and drops rapidly to reach at 2 years, the normal value for adults, which is less than 10 ng / ml. Elevation of AFP is considered a biomarker in some liver tumors.[3]

Ultra sound remains the first-line examination, making it possible to confirm the hepatic localization of the mass, to differentiate solid tumors from cystic tumors (cystic mesenchymal hamartoma and undifferentiated sarcoma), to look for signs of hyper vascularization (hemangioma in the infants, UFH in adults), signs of portal or hepatic thrombosis (malignant tumor), signs of portocava fistula favoring the occurrence of UFH or adenoma.[1] CT scan and Magnetic Resonance Imaging (MRI) with contrast injection are useful for diagnosis and extension workup, and their respective indication will be discussed depending on the clinic. Biopsy is still performed frequently for diagnostic or prognostic purposes for cytogenetic studies. The association of a hepatic mass with an elevation of AFP in a newborn baby is suggestive of the diagnosis of hepatoblastoma, or hepatichemangioma.[4] Hepatoblastoma is the most common malignant liver tumor in children. It mainly affects

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children under the age of 3.[4] Hemangioma is the most common vascular tumor in infants, solitary or multinodular, of very variable severity, asymptomatic or complicated by congestive heart failure, thrombocytopenia, compartment syndrome, pulmonaryarterial hypertension.[5]

The interest of this observation is to know how to evoke the diagnosis of infantile hepatic hemangioma in front of a hepatic mass of the infant, which requires an exploration and a rapid management, because the diffuse form can endanger the vital prognosis of the patient by its cardiac repercussion.[6]

CLINICAL CASE

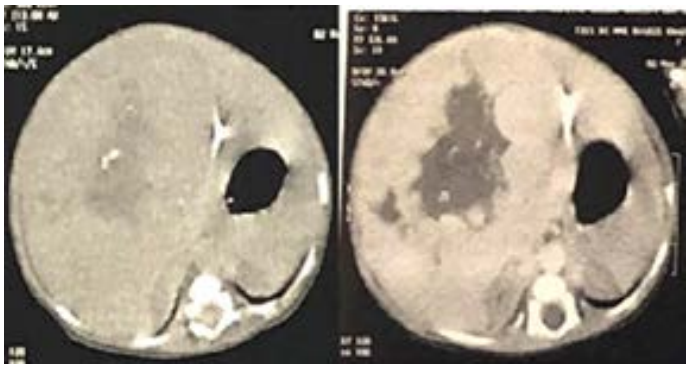
We report the case of the newborn, K., 2 days old, male, born at eutrophicterm, transferred to neonatal intensive care for respiratory distress from birth. The pregnancy was poorly followed, delivery by cesarean section. He is the couple's only child. The parents are not consanguineous. Symptoms go back from birth, with the presence of respiratory distress. He was put on CPAP and transferred to neonatal intensive care for diagnostic and therapeutic management. The general clinical examination noted a fever at 37.9°, poor general condition, a grayish complexion, edema of the lower limbs, polypnea at 65c/min. Physical examination showed a large, shiny abdomen with collateral venous circulation. Abdominal palpation revealed a hardly depressible abdomen with tender hepatomegaly (hepatic arrow at 8cm) occupying the entire right hypochondrium and descending to the right iliac fossa, without splenomegaly. The sucking reflex as well as the archaic reflexes were present. He had no facial dysmorphia or externalized bleeding. The mucocutaneous examination did not reveal any angiomas. Cardiovascular auscultation was unremarkable. Biologically, the full blood count showed thrombocytopenia at 26G/10^3, hemoglobin is 17.7g/ dl, white blood cells at 9.71G/10^3. The prothrombin level (PT) was measured at 36%, the activated partial thromboplastin time (TCA) at 56.5 seconds and the subject TCA to control TCA ratio at 1.93.

The hepatic assessment, showed hepatic cytolysis, relating to the ASAT 136 IU/L, the ALTs were 49 IU/L, associated with cholestasis with a total bilirubin level at 127 mg/l, free bilirubin at 114 mg/l , a conjugated bilirubin at 13 mg/L, GGTs were increased to 223 IU/L, as well as LDH to 1762 IU/L. The blood sugar cycle for hypoglycemia was normal.

Trans-fontalar ultrasound performed at birth did not show cisterno-ventricular dilation, nor any visible mass or fluid collection. An abdominal ultrasound performed at birth showed multi-heteronodular hepatomegaly with some small calcifications.

Faced with the ultrasound appearance, an abdominal computed tomography scan was performed and revealed an enlarged liver with regular contours, the site of a large mass occupying almost the entire liver parenchyma, poorly limited by bumpy contours. This mass was of predominant tissue density with individualization of a central area of necrosis of fluid density, containing calcifications. It was intensely enhanced in early stage and portal stage (Figure 1), without any notion of enhancement in peripheral or centripetal clod. It measured 67x96x85 mm, came into contact with the inferior vena cava which remained permeable, in contact with the abdominal wall and the gastric wall.

Figure 1: A) abdominal CT scan without injection of contrastproduct, B) after injection of contrastproduct



The AFP dosage was 27,144 IU/ml on Day 1 and 30,564 IU / ml on Day 6. The BHCG assay was negative. Faced with this CT appearance more reminiscent of a hepatoblastoma than a solitary angioma, and a high dosage of AFP, an ultrasound-guided percutaneous biopsy was performed, revealing on anatomopathological examination a hepatic parenchyma altered with hyalinized tissue, the seat of hemorrhagic raptus, as well as fibrous tissue harboring rare regular canal structures and congestive vessels, in favor of a vascular malformation or tumor, with a total absence of tumor lesion. The chest x-ray suggested significant cardiomegaly. The patient received transfusions in platelet units and in fresh frozen plasma (FFP), treatment with antibiotics (meronem, amikacin, vancomycin) and vitamin K. The evolution was marked by a very rapid increase in abdominal volume with a significant worsening of respiratory distress and collateral venous circulation. He was intubated ventilated and received hemodynamic support. The evolution was unfortunately fatal.

DISCUSSION

Infantile Hepatic Hemangioma (HHI) is a subclass of infantile hemangiomas that are the most common self-limiting benign tumors of early childhood.[7] HHI is the most common hepatic vascular tumor in the first 6 months of life with a predilection for girls.[8] Its discovery is often fortuitous, following the performance of an abdominal ultrasound requested in the presence of military hemangiomatosis or PHACES syndrome.[9] It has 3 patterns: focal, multifocal or diffuse. Despite the benign nature of HHIs, multiple and diffuse lesions can present with life-threatening complications, including congestive heart failure, and severe hypothyroidism, requiring prompt medical intervention. Therefore, proper diagnosis is of utmost importance.[10]

In the case of diffuse involvement, a reliable diagnosis can be made on the sole basis of radiological characteristics. Indeed, the characteristic radiographic appearance of lesions in contrasted CT includes well-defined margins, hyper-acute peripheral nodular / wavy enhancement (that is to say arterial) with progressive centripetal filling in the portal venous phase without vascular invasion or lymphadenopathy and sometimes associated with a sudden constriction of the subceliac aortic caliber.[11]

However, for some patients, as in our reported case, despite

careful physical examination combined with imaging and tumor marker studies, diagnostic difficulties persist.[12] Indeed, once a liver mass is identified in an infant, the differential diagnosis ranges from vascular malformations to benign and malignant tumors. Although hepatoblastoma is a rare tumor in children, it remains the most common malignant tumor of the liver in newborns.[12] The rate of AFP is just as high for hepatoblastoma as for HHI. In these cases, liver biopsy is of utmost importance to make the correct diagnosis.[8]

Hepatic hemangioma of the fetus and newborn has a poorer prognosis than tumors in the older patient. The prognosis worsens with a higher risk of mortality when it is the diffuse form, or when there is an association with congestive heart failure, severe anemia or thrombocytopenia.[13] A well-monitored pregnancy with a prenatal diagnosis can anticipate and improve the prognosis.[14]

Since the discovery in 2008 of their effectiveness in the treatment of infantile hemangiomas, beta-blockers have been the subject of numerous studies and are now the first-line treatment.[15] Propranolol has a marketing authorization (AMM) for this indication.[9] This oral treatment is initiated between the age of 5 weeks and 5 months according to the Marketing Authorization, for a period of 6 months,[16] and it is more effective if it is started very early.[17]

Treatment should be prompt for symptomatic patients and close follow-up should be reserved for asymptomatic patients to improve mortality.[18]

CONCLUSION

Before any hepatic mass in an infant, with an elevation of AFP, we must always think of an HHI. The management of hepatic tumors in children must be done in a specialized center because it involves the participation of oncopediatricians, pediatric surgeons and pediatric radiologists. However, the role of the pediatrician who first sees the child and its parents is very important. With knowledge of the diagnostic ranges of pediatric hepatic tumors, and careful examination for the relevant clinical, biological and radiological elements, the physician an speed up the diagnostic process, and guide treatment correctly.

CONFLICTS OF INTEREST

None.

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