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Case Report
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Intantile Hepatic Hemangioendothelioma with Kassabach Merrith Syndrome: A Rare Cause of Acute Liver Failure in Children

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Abstract

Most hemangiomas have a distinctive ultrasound (US) appearance, and even with those with atypical appearances on conventional gray-scale US, specific diagnoses can be made using pulse-inversion harmonic US with contrast agents. In this essay, we review the spectrum of US appearances of hepatic hemangiomas on conventional gray-scale, power doppler, and pulse-inversion harmonic US with contrast agents.

Keywords: Hemangiomas; Power Doppler; Pulse-inversion

Introduction

Hemangiomas are the most common benign tumor of the liver, and are not infrequently encountered incidentally during U/S screening. Because the vast majority of hepatic hemangiomas are asymptomatic and require no treatment, they must be differentiated from hepatic malignancies. Most have a distinctive US appearance [1-4] but some, known as atypical hemangiomas, show various US patterns [5-8]. For the differentiation of the atypical variety, dynamic contrast-enhanced studies have been widely employed. Using microbubble contrast agents, it has been shown that pulse-inversion harmonic US can effectively depict the typical enhancement patterns of hepatic hemangiomas [9], enabling on conventional gray-scale, power Doppler, and pulse-inversion harmonic US with contrast agents. Infantile hepatic hemangioendothelioma (IHH), a type of primary mesenchymal hepatic neoplasm, comprises 1-2% of all pediatric hepatic tumors and is seen particularly during the first year of life. There are two different histopathologic subtypes, according to cell size and vascularity: Type1 has benign histopathologic specific diagnosis. In this essay, we review the spectrum of US appearances of these hemangiomas as seen characteristics, although rarely may display malignant clinical behavior, while Type 2 has more atypical cellular properties. Thrombocytopenia and consumption coagulopathy due to hemangioma or large vascular lesion, called "Kasabach Merritt syndrome" (KMS), may develop in some cases. In the current literature, data about IHH are usually represented as single cases. Therefore, there is difficulty in clinical assessment and therapeutic approach in IHH.

Case Presentation

An eight months old female infant admitted with the assessment of severe pneumonia, hepatic hemangioma with kassabach merrith syndrome after she presented with dry cough of one week duration associated with grunting , fast breathing, high grade intermittent fever and vomiting of ingested matter. For this complaint, she was taken to private clinic and was given unspecified po medications and was then referred to Tikur Anbessa Specialized hospital because she didn't show any improvement.

She has been experiencing progressive abdominal swelling noticed at the age of 02 months. For this U/S of the abdomen was done and showed multiple liver masses, then She was started on predinsolone daily with the impression of hepatic hemangioma and appointed to come back after a month for the next follow up.

When she came for her follow up, she had bleeding diathesis (epistaxix, petechial rash, evidences of anemia and jaundice.) This time, repeat U/S showed significant increment in size of the masses, HCT= 13% and platelet= 15,000.

Physical examination

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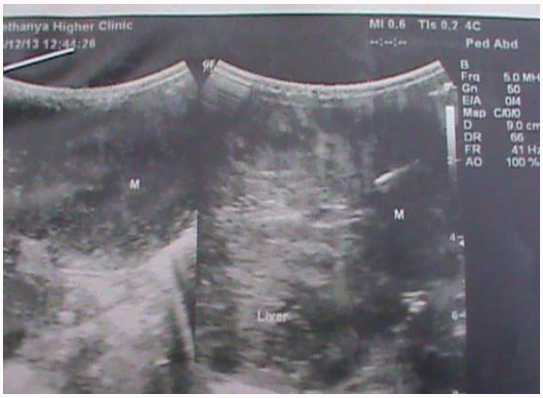

Figure 1: U/S showing multiple hypodense masses in this infant.

V/S-PR=150, bounding RR= 60, T=38oc, BP= 110/50 mmHg (wide pulse pressure).

Anthropometry- Not affected

Pale conjunctivae and icteric sclerae.

Chest: Signs of respiratory distress with coarse crepitation.

CVS: Ejection systolic murmur at pulmonary area with S3 gallop.

Abdomen: Grossly distended. Liver is hugely enlarged which is firm, has nodular surface with blunted edge which measures 11cm BRCM with TVLS=15cm. Spleen measures 3cm.

Integumentary: Purpuric rash scattered all over the body.

Investigations: WBC=14,200 with N=59% and L=41%, Hb=4g/dl Hct=13.5%, PLT=15,000→17,000 (repeat), ESR=32, MCV= 76, MCH=30, MCHC= 30.7, Retic count=1.8%, Alpha feto protein (tumor marker)=9iu/ml (normal), **Bleeding profiles:** PT= 33(12.6-18) ,INR=0.96, aPTT= 37(14.5-28.6).

Coombs' test: Negative, TORCH screening = non revealing, DNA PCR for HIV negative, hepatitis viral markers were negative.

First Abdominal ultrasound: Liver is enlarged in size 12.7cm with bumpy smooth contour and multiple well defined hypoechoic masses seen. There is background of normal liver in between. The masses have significant increment in sizes, and increased vascularity relative to the normal liver.

Index= Hemangioma of the liver.

Second Abdominal ultrasound: Liver is enlarged in size 12.7cm with bumpy smooth contour and multiple well defined hypoechoic masses seen. There is background of normal liver in between. The masses have significant increment in sizes, and increased vascularity relative to the normal liver (Figure 1).

Index: multiple liver masses secondary to Infantil hemangioendothelioma? Hemangioblastoma?? Metastasis.

Abdominal CT: There are multiple different sized well defined nodular mass lesions which shows intense contrast enhancement similar to abdominal aorta. It measures in the range of 0.5x4.7cm in diameter. The larger lesions showed peripheral enhancement on the early arterial contrast phase with centripetal filling.

Index: Multiple liver masses which have patterns of contrast enhancement similar to hemangioma; multiple liver hemangioendotheliothelioma (Figure 2).

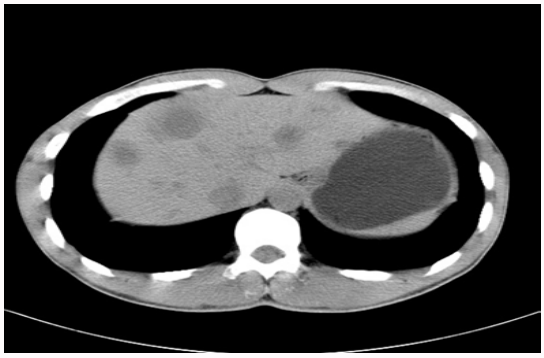

Figure 2: CT of the abdomen showing multiple hypodense masses.

Discussion

Infantile hepatic hemangioma is a proliferative endothelial cell neoplasm that involves the liver. The lesion is composed predominately of endothelial cells and has characteristic phases of rapid growth caused by cellular proliferation and spontaneous involution (which can be accelerated by the use of angiogenesis inhibitors). It is frequently referred to as hepatic hemangioendothelioma type 1 or type 2. However, it differs from epithelioid hemangioendothelioma and adult hepatic hemangioma.

The former is a proliferative tumor that has malignant potential and does not involute, and the latter is believed to represent a vascular malformation and also does not involute. The term "infantile hepatic hemangioma" is preferred to "hemangioendothelioma" because the clinical and biologic behavior is similar to infantile hemangiomas that affect the skin and other parts of the body [1–3].

A subset of hemangiomas produces clinical symptoms because of lesion size, location, or hemodynamic effect [4]. This subset is often referred to a tertiary care facility for confirmation of diagnosis and treatment planning. Therapeutic options include steroids; interferon alfa-2a, embolization; and, less frequently, chemotherapy, radiotherapy, surgery, or liver transplantation. In the diagnosis of hepatic hemangiomas, the most important aspect is non-invasive differentiation from other tumors. The diagnosis and treatment of infantile hepatic hemangiomas have received much attention in recent years as a result of advances in diagnosis and treatment [5–7].

Conclusion

Infantile hepatic hemangiomas are the most common hepatic vascular neoplasms of infancy. Most are asymptomatic lesions that are incidentally discovered during imaging of the abdomen, but some are associated with severe symptoms such as high output cardiac failure, hepatic dysfunction, and abdominal compartment syndrome. U/S and CT of the abdomen are important investigative modalities for visceral hemangiomas. Our 8 months old infant had clinical evidences of hepatic hemangioendothelioma with Kassabah. Merritt syndrome (rapidly increasing multiple hemangioma of the liver with anemia and thrombocytopenia) with U/S and CT- evidences, confirming that the patient was real case of IHH with KMS (Infantile hepatic heangioendothelioma with Kassabach Merritt Syndrome).

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