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Mesenchymal Hamartoma of the Liver: About A Case

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Abstract

Case Report

Mesenchymal hamartoma of the liver is a rare benign tumor, diagnosed mainly during the first two years of life. Its pathogenesis is poorly understood. Imaging plays an essential role in diagnosis and post-therapeutic follow-up. The diagnosis of certainty remains histological and the treatment is essentially surgical. We report a case of hepatic mesenchymal hamartoma diagnosed in a one-year-old infant with isolated abdominal distension. The diagnosis was evoked on imaging and confirmed after histological examination.

Keywords: Mesenchymal hamartoma, diagnosis, Hepatic tumors, heterogeneous echostructure.

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INTRODUCTION

Hepatic tumors represent 1 to 4% of solid tumors in children and are generally represented by hepatoblastoma, hepatocellular carcinoma and hemangioendthelioma [1, 2].

Liver hamartoma is a rare condition, which corresponds to 5 to 8% of primary liver tumours [2].

It is a benign lesion which is most often discovered before the age of 2 years, exceptionally in adults. Its origin and pathogenesis are still obscure [2].

Imaging plays an essential role in the positive diagnosis as well as in the post-therapeutic follow-up [2, 3], nevertheless the diagnosis of certainty is histological [1].

The therapeutic management of mesenchymal hamartoma of the liver consists of surgical excision of the mass, per and post-operative complications are rare and its prognosis isgenerally good [2].

OBSERVATION

We report the case of a one-year-old male infant with abdominal distension evolving for two months in a context of apyrexia and conservation of general condition. Abdominal examination revealed a huge abdominal mass in the right hypochondrium and right flank.

Biological assessment was unremarkable and blood alpha-feto-protein assay was normal.

An abdominal ultrasound objectified a large mass that appears to be at the expense of the left liver, of heterogeneous echostructure, vascularized on color Doppler, site of multiple anechoic cystic formations (Figure 1).

To better characterize this mass and study its relationships, further exploration by abdominal CT scan with injection of PDC was done and showed a heterogeneous liver, increased in size, with regular contours, seat of a voluminous tissue mass at the expense of the lobe. left extended up to the FIG, measuring 17x15.5x9.6 cm (T x CC x AP), well limited, regular contours, spontaneously heterogeneous isodense, seat of hypodense areas related to areas of necrosis, heterogeneously enhanced after injection of PDC. It pushes back the abdominal aorta and the IVC which remained permeable as well as the pancreas with loss of fatty border of separation by place. It includes the left hepatic artery which remains permeable as well as the left portal branch (Figure 2).

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H. El Madkouri *et al.*, Sch J Med Case Rep, Oct, 2022; 10(10): 996-998 diagnosis of mesenchymal hamartoma of the liver.

An echo-guided biopsy of the mass was performed and histological examination confirmed the

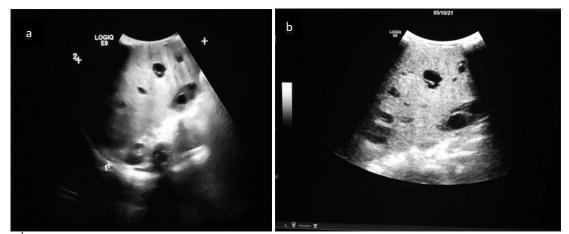


Figure 1 (a and b): Abdominal ultrasound shows a mass at the expense of the multi-cystic heterogeneous left liver

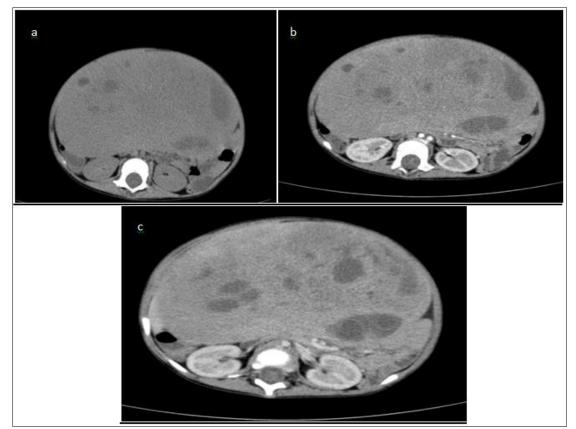


Figure 2: Abdominal CT scan without (a) and with PDC injection (b and c) shows the presence of a large hepatic mass at the expense of the left lobeHeterogeneous isodense, seat of hypodense zones, heterogeneously enhanced after injection of PDC

DISCUSSION

Mesenchymal hamartoma of the liver is a very rare condition, represents 5 to 8% of hematic tumors in children and ranks second among benign liver tumors in children after infantile hemangioendothelioma [4]. It is usually observed during the first two years of life in 80% of cases [1, 4], exceptionally in adults [4, 5]. A mesenchymal hamartoma is defined as a tumor-like malformation in which there is an abnormal mixture of normal constituents of the organ involved. Its origin and pathogenesis are still obscure. Some authors believe that it is a malformative lesion developed in utero from the primitive portal mesenchyme; others evoke a reaction origin to an ischemic process or a biliary obstruction. Recently the tumoral origin was suggested in front of the identification of a balanced translocation involving the chromosome 19q13.4 [1, 4, 6, 7].

The tumor is often discovered following palpation of an abdominal mass or by progressive distension of the abdomen. Other signs have also been reported, namely abdominal pain, anorexia, vomiting, weight loss and digestive transit disorders.

Ultrasound, abdominal CT and MRI play a key role in the positive diagnosis as well as in posttherapeutic follow-up [3]. On ultrasound, it is generally a voluminous, essentially cystic lesion, well limited and comprising fine septa, the presence of hyperechoic parietal nodules inside the cyst is strongly suggestive of hamartoma. In some cases the tumor takes on the appearance of a solid echogenic mass as in the case of our observation [2, 3].

The scanner allows a better anatomical definition, to confirm the hepatic origin of the mass and to plan the possible surgical approaches. Its appearance is that of a moderately enhanced hypodense cystic mass after injection of PDC [1, 7, 8].

In MRI, the solid part of the lesion is generally in hyposignal on the T1 and T2 weighted sequences. The cystic part has a variable signal depending on the protein concentration but can appear frankly liquid [3].

In most cases, the diagnosis is suggested by imaging. However, the diagnosis of certainty is histological and it is usually easy because of the typical microscopic characteristics of the tumor [4].

Surgical excision and the treatment of choice [4] complete resection of the lesion can sometimes be difficult or even impossible due to the involvement of vital structures. Per and postoperative complications are rare [2]. After complete resection, clinical and ultrasound monitoring is recommended for at least 5 years, but generally the prognosis is immediately excellent [2].

There are several non-surgical options, especially in asymptomatic infants, namely percutaneous drainage, chemotherapy and radiotherapy, but there are no studies proving their effectiveness [2].

CONCLUSION

Mesenchymal hamartoma of the liver is a rare benign tumor in children. Its pathogenesis remains a matter of controversy. Imaging plays a key role in the positive diagnosis and in the post treatment follow-up; however the diagnosis of certainty remains histological. The treatment of choice is surgery and the prognosis is immediately excellent.

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