



Case Report

Hepatic Mesenchymal Hamartoma.

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Abstract

Hepatic mesenchymal hamartoma is a rare benign tumor arising from the mesenchyma of the portal tract. Hamartoma usually presents before the third year of life. Remarkable abdominal swelling is the most frequent clinical feature. We herein present a new case with a comprehensive literature review. The aim was to highlight the clinical, radiological and histopathological characteristics of this entity.

Keywords:

Mesenchyma; Hamartoma; Liver tumor; Children.

Introduction

Hepatic mesenchymal hamartoma (HMH) is an uncommon tumor arising from the mesenchyma of the periportal tract. It accounts for 8% of all pediatric hepatic tumors. However, its etiopathogeny is still unclear and the management remains controversial [1]. Abdominal enlargement and respiratory distress are the most common clinical presentations [2]. HMHs are benign lesions and must be considered in the differential of liver malignancies.

Observation

A 17-month-old boy was referred to us for progressive diffuse abdominal enlargement started 2 months ago. His medical history was unremarkable. The physical examination showed a large abdominal mass palpable on the right upper abdominal quadrant. All the laboratory tests performed including alpha-fetoprotein (AFP) were within normal range. The abdominal radiograph showed a large non calcified intra-abdominal soft tissue density in the right upper quadrant with a displacing mass effect on the gastrointestinal tract (figure 1). Chest radiograph showed an elevated right hemidiaphragm. Abdominal ultrasound examination identified multiloculated cystic mass occupying the right hepatic lobe with multiple thin echogenic septae (figure 2).

CT scan of the abdomen and pelvis was performed for further assessment. It revealed large 13x7x7cm cystic tumor with heterogeneous enhancement of its solid component. The cystic portion contained multiple enhancing septae (figures 3).

The patient underwent a laparotomy. A huge, encapsulated tumor developed between the hepatic segments IV and V was noted. Tumorectomy was performed. Frozen sections confirmed its benign character. The postoperative courses were uneventful. The patient was discharged on the fifteenth postoperative day.

Microscopically, the tumor contained hepatocytes, abnormal bile ducts and immature mesenchyma in variable portions. Most of the cysts were areas of degeneration with fluid accumulation separated by thin strands of connective tissue (figure 4). The histopathology concluded to mesenchymal Hamartoma of the liver. On the six months follow up, the child was asymptomatic with normal physical examination.

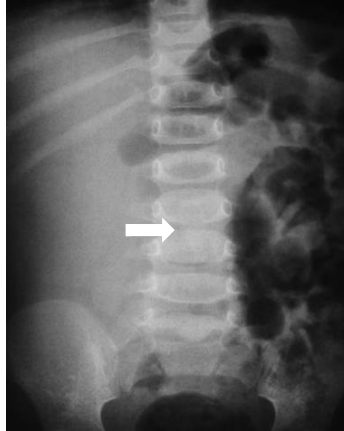


Figure 1: Displacing mass effect on the gastro-intestinal tract

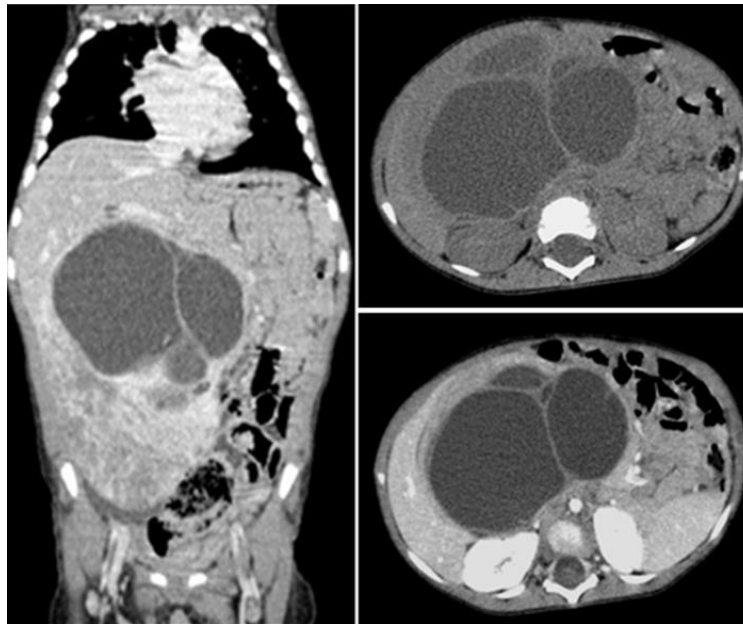


Figure 2: CT features of liver mass with heterogeneous enhancement of its solid component. The cystic portion contains multiple septae.

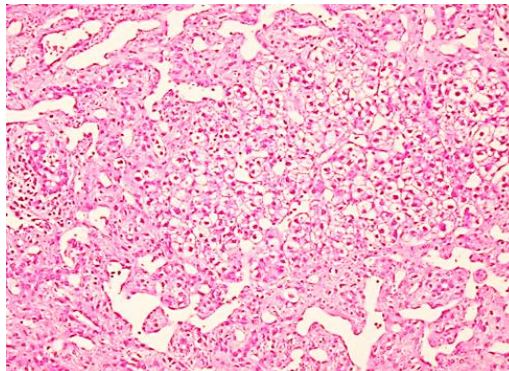


Figure3: histopathological features

Discussion

Described since 1903, Hepatic mesenchymal hamartoma (HMH) is one of the most common benign liver tumors in children. The lesion is usually diagnosed within the first 2 years of life [3]. However, prenatal cases diagnosis has been described [4]. The most frequent clinical presentation is upper abdominal palpable mass [5]. The tumor is in the right lobe in more than 90% of cases. The liver function remains usually normal. Mild elevation of the alpha-fetoprotein is occasionally observed and has no malignancy significance [6]. Histologically, HMH is characterized by a lobular growth of myxomatous connective tissue containing scattered bland stellate-shaped mesenchymal cells [7]. The etiopathogenesis involves mutations and genetic alterations in both sporadic and syndromic HMHs [8]. Imaging features are typically of complex cystic mass with multiple solid enhanced septae [9]. Complete surgical excision is the only curative option. Malignant transformation on the residue of incomplete excision has been described [10]. Ultrasound guided aspiration and monitoring are proposed for unresectable tumor cases.

Conflict of Interest: None

References

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