HEPATIC MESENCHYMAL HAMARTOMA: A CASE REPORT AND RADIOLOGICAL FINDINGS

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ABSTRACT

Primary tumours of the liver are uncommon in childhood. Of these, more than two-thirds are malignant. As such, benign hepatic tumours are often not considered in the differential diagnosis of a hepatic mass in childhood. We report a case of hepatic mesenchymal hamartoma, a rare benign tumour, in a 10-month-old infant. This tumour is characterised by an admixture of ductal structures within a copious loose connective tissue stroma. Only approximately 160 cases had been reported in the literature. Awareness of the ultrasound (U/S) and computed tomography (CT) features, although not diagnostic, is helpful in distinguishing it from the more common malignant tumours. A correct preoperative diagnosis is important as surgical excision is often curative.

Keywords: *mesenchymal hamartoma, diagnosis, ultrasound, computed tomography.*

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INTRODUCTION

Hepatic mesenchymal hamartoma is a rare benign lesion arising from the mesenchymal tissue in and around the portal system, with associated cystic changes. It is seen predominantly in children within the first 2 to 3 years of life. Painless abdominal swelling is the usual clinical feature. The tumours are usually solitary and are more commonly located in the right lobe of the liver. We present a case of mesenchymal hamartoma that involves only the left lobe of the liver.

CASE REPORT

A 10-month-old baby boy was referred from a nearby general hospital for progressive abdominal distension over the preceding 4 months with associated loss of weight. His appetite was normal and there were no febrile episodes. Physical examination revealed an enlarged liver measuring 15cm below the right subcostal margin. Except for a palpable occipital lymph node, no other significant findings were noted.

The basic laboratory investigations were normal. Specific investigations however showed an elevated serum alpha fetoprotein (AFP) of 165 I.U. (normal : 0-10). The urine vanillyl-mandelic acid (VMA) was borderline positive on 2 occasions. An abdominal ultrasound (U/S) revealed a large, slightly echogenic intraabdominal mass. The mass appeared to arise from the left lobe of the liver. Cystic spaces without septation, the largest measuring 7cm in diameter, were seen within this mass (Fig 1 and 2). The right lobe of the liver and the gallbladder appeared normal.

On the contrast enhanced computed tomographic (CT)

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Fig 1 - Ultrasound section through the liver showing a solid mass in the left lobe which was more echogenic than normal liver parenchyma. A small cystic lesion with posterior acoustic enhancement was noted in the mass.

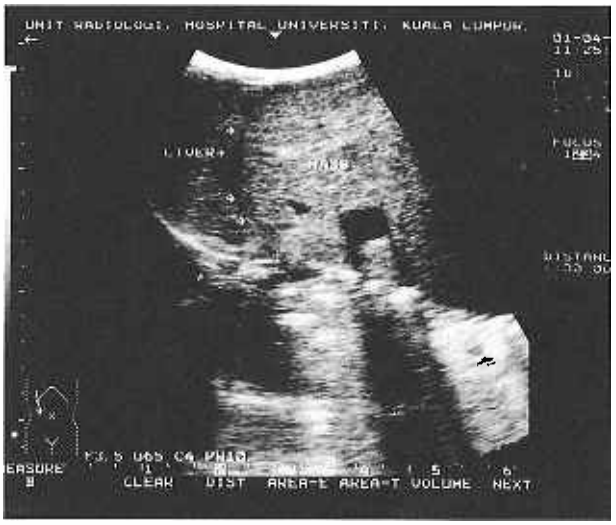


Fig 2 - Ultrasound section through the cystic component of the mass which appeared anechoic.

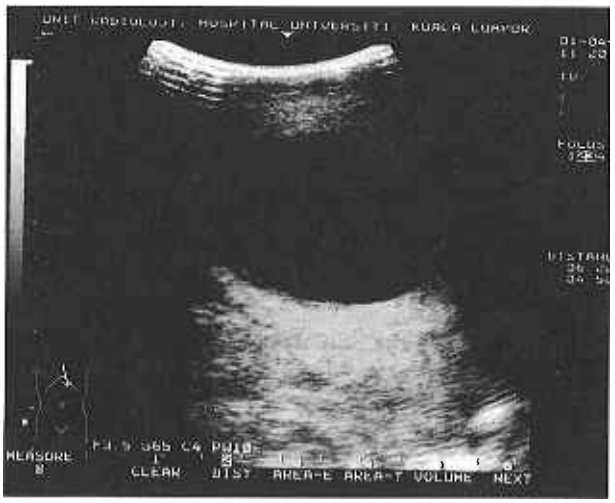
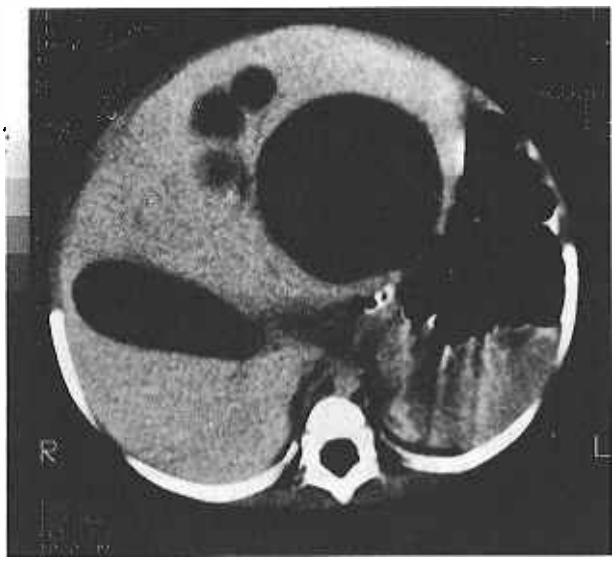


Fig 3 - Axial CT section following iv contrast medium, showing the solid mass with cystic components in the left lobe of the liver. The solid mass appeared more enhanced than the normal liver parenchyma. A dilated gallbladder was seen on the right of this mass.



scan, the mass measured 17 x 14 x 12cm and was predominantly solid with 4 separate cystic lesions seen within it. The largest cyst measured 5cm and the smallest 1cm in diameter (Fig 3). The mass occupied most of the left lobe, medial to the falciform ligament, and extended inferiorly into the pelvis. The bowel loops were displaced inferiorly and to the left of the abdominal cavity by this large mass. No calcification was detected in the mass. The right lobe of the liver appeared normal. No paraaortic lymphadenopathy was noted on the scan.

At laparotomy the large mass was confirmed to arise from the left lobe of the liver and a left hepatectomy was performed. This removed mass measured 19 x 19 x 15cm and weighted 1480gm. Histopathological examination of the mass revealed a hamartomatous lesion with variable mixture of tissues. The predominant mesenchymal component consisted of loose connective tissue with multiple branched and tortuous biliary ducts, lymphatics, blood vessels and irregular islands of liver cells. There were no mitotic activity nor atypia of the liver cells. A few cysts of varying sizes lined by flattened cells were present.

The infant had an uncomplicated post-operative course and had been asymptomatic since.

DISCUSSION

The majority of hepatic masses in the paediatric age group are metastases, most commonly arising from Wilm's tumour or neuroblastoma[1]. Primary neoplasms are less frequent with 72% being malignant and 28% benign. Although mesenchymal hamartoma is generally regarded as the second most common benign hepatic tumour after haemangioma[2], it only accounts for 6% of primary hepatic tumour in childhood[3] with approximately 160 total cases reported in the literature[4]. This lesion usually is found between the ages of 4 months and 2 years with a slight male preponderance[5,6]. It usually presents as an enlarging upper abdominal mass. The progressive abdominal enlargement may be noted over a period of days or weeks to as long as a year. The mass is rarely associated with abdominal pain or tenderness. Decreased appetite, vomiting after meals and respiratory distress due to impaired diaphragmatic excursion had been noted occasionally[5,7,8]. The serum AFP may be normal or increased[5,8,9]. Ito et al found AFP to be markedly elevated in 5 out of 7 cases and suggested that it was produced by proliferating hepatocytes in the tumour[10].

Urinary VMA was not found to be abnormal in the previous reports[7,8]. The borderline positive results in our patient may be related to non compliance in restriction of certain food, such as citrus fruits and food containing chocolate and vanilla, which may cause a false positive urinary VMA result.

Mesenchymal hamartoma of the liver are large tumours, often weighing more than 1000g[6]. In most cases the tumour is located in the right lobe of the liver[1,6], although the left (as in this patient) and in a few instances both lobes may be involved[11,12]. The ratio of right of left lobe involvement had been quoted as 6:1[13].

Histologically, a disordered arrangement of primitive mesenchyme, bile duct and hepatic parenchyma is present. The loose connective tissue stroma has a peculiar propensity to accumulate fluid in large quantities to create microscopic and macroscopic acellular space[1,5,14,15]. Macroscopic cysts, single or multiple, had been found in 80% of cases in a previous report[8].

The U/S and CT findings reflect the spectrum of cystic change observed pathologically. The sonographic appearances may be predominantly solid with multiple small cysts, as was noted in this patient, or multiloculated cystic mass with few solid septa[16,17], the latter being the commoner sonographic finding[6]. In addition, the septa within the cystic component may be mobile[18].

On CT, the hamartoma usually appears as a well-defined mass with low attenuation cystic components. Internal septations are typical although not seen in this patient. Hamartoma without cystic component had also been reported[6]. There is often only minimal or no enhancement of the tumour mass following intravenous contrast medium[6,11,17,19]. No calcification is seen in the mass.

Although the U/S and CT findings are not specific[15], they help to exclude the more common solid liver tumours such as solid hepatoblastoma, infantile haemangio-endothelioma, hepato-cellular carcinoma, adenoma and metastases[5]. The differential diagnoses narrowed down to cystic hepatic lesions which include mesenchymal hamartoma, congenital liver cyst, abscesses, haematoma, teratoma, cavernous haemangioma and cystic hepatoblastoma[5,11,17]. Congenital cyst does not have any solid component or septation[11,19]. Abscess and haematoma have a different clinical presentation and appear more solid on U/S. Teratoma often contains calcifications and can therefore be easily recognised[7,11]. Contrast enhanced dynamic sequential CT scan will reveal the typical enhancement from the periphery towards the centre[11] in cases of cavernous haemangioma[11]. Cystic hepatoblastoma is rare and will be associated with elevated serum AFP[5]. A raised serum AFP however does not exclude the diagnosis of mesenchymal hamartoma as in this case.

In conclusion, mesenchymal hamartoma should be included in the differential diagnosis of a liver mass with cystic component presenting below the age of 2 years, as this is a benign condition and surgical resection is curative.

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