Multiple Haemangioma/ Haemangioendothelioma

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ABSTRACT

We describe the ultrasonography, computed tomography and magnetic resonance imaging findings of infantile haemangioma or haemangioendothelioma of the liver who presented to us clinically and biochemically suspicious of obstructive jaundice.

Keywords: Diagnostic imaging, Infantile haemangioma, jaundice

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INTRODUCTION

Infantile haemangioma or haemangioendothelioma of the liver, is common and knowledge of their clinical presentation, biochemical results and radiological findings are important in differentiating them from the other more sinister tumours.

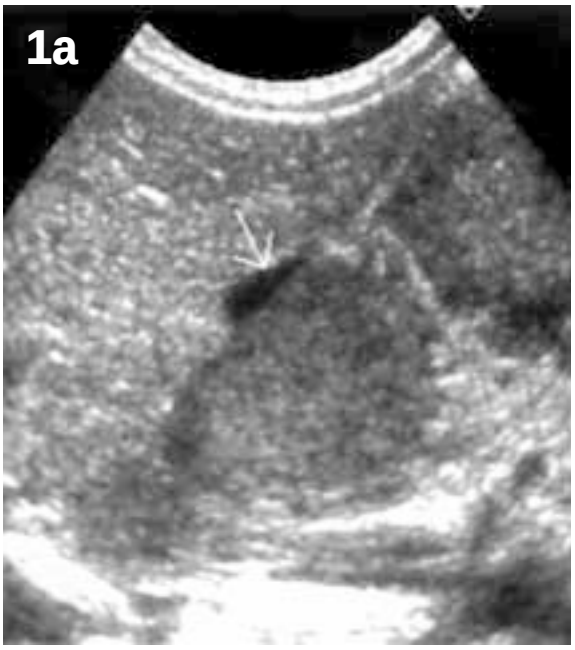
CASE REPORT

Our patient was a full term baby presented at one month of age with history of prolonged jaundice. Apart from a tinge of jaundice and hepatomegaly, he was feeding well with satisfactory weight gain. Both antenatal and perinatal history were unremarkable.

Investigation revealed presence of conjugated hyperbilirubinaemia. Direct bilirubin was 49 umol/l (normal: 1-5 umol/l), indirect bilirubin was 18 umol/l (normal: 0-19 umol/l), Beta Human Choronic Gonadotrophin (HCG) < 2.0 IU/L (normal: < 5.0 IU/L), Alpha-fetoprotein (AFP) 1438.2 ng/ml (normal < 3500 ng/ml).

Ultrasonography of the liver demonstrated three well-defined fairly homogenous hypoechoic nodules in the right lobe and caudate lobe. The largest has a maximum diameter of 2 cm. There was no internal calcification. However, Doppler study reveals presence of both arterial and venous flow within the nodules. The biliary system was not dilated and there was no evidence of obstructive jaundice. Both portal vein and inferior vena cava were normal in calibre. (Fig. 1a, b)

The spleen was not enlarged and adrenal glands were normal. A diagnosis of multiple haemangiomata/ haemangioendothelioma was made.



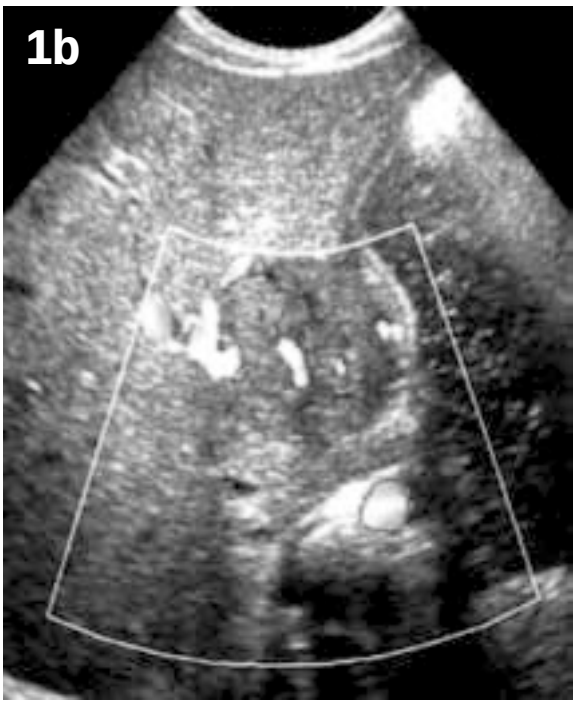


Fig.1a, b Ultrasonography demonstrates a well defined hypoechoic homogenous liver nodule (a). Duplex & Colour Ultrasonography showed presence of both arterial and venous blood flow within the nodule (b).

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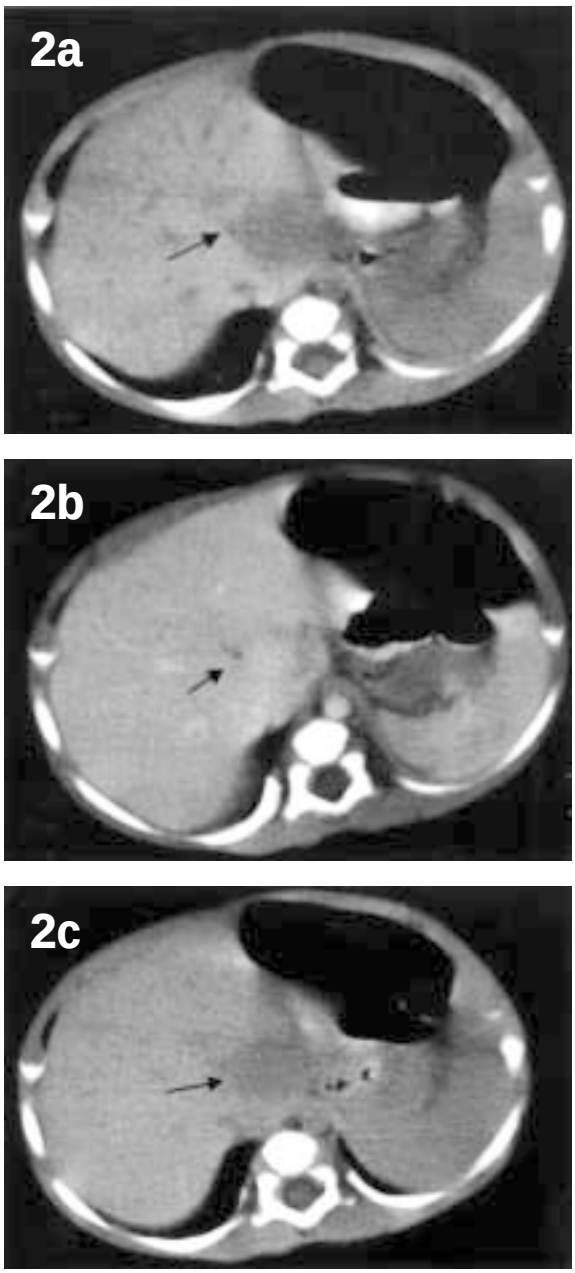

Fig.2a-c Computed Tomography reveal a hypodense (a) that enhances homogenously with intravenous contrast. (b) and becoming hypodense on delayed scan (c).

Computed tomography (conventional) performed showed the nodules to be hypodense relative to the liver. With infusion of contrast there was complete homogenous enhancement in one lesion and peripheral enhancement in the other two.

The delay images at five minutes showed the nodules to be relatively hypodense compared to the surrounding liver parenchyma. (Fig. 2a-c)

MRI was performed which demonstrated the nodules to be hypointense on T1 weight images and hyperintense on T2 weight images. Homogenous enhancement that was found after infusion of gadolinium - DTPA contrast (non-dynamic contrast scan) indicates the presence of solid tissue. (Fig. 3a-c)

Subsequent follow-up ultrasonography performed at six months of age showed disappearance of two of the nodules previously found in the right lobe, while the third in the caudate lobe became smaller.

DISCUSSION

Infantile haemangioendothelioma or capillary haemangioma of the liver is the most common benign hepatic tumours found in infants. They often present early, in the first six months of life and the majority of the patients were less one month of age. Males are more frequently affected than female. However, in another series, there was no difference in the sex distribution[1,2].

Haemangiomas can present as multicentric or as solitary lesion. The frequency of these reported in the literature depended on the population that was being investigated[1,3].

Clinical presentation depends very much on the size of the lesions. Patients may present with relatively mild clinical features as in our patient or more dramatically with high output cardiac failure. Multicentric hepatic haemangioendotheliomas are vascular lesions of the liver that usually present in the infancy with hepatomegaly, high output congestive heart failure and cutaneous haemangiomas[4]. Invariabily, there is some degree of hepatomegaly. Anaemia, hyperbilirubinaemia and increased AST level may also be present[1].

Plain radiographs are not sensitive but may sometimes demonstrate fine speckle or fibrillar type of calcifications in the haemangioma.

Ultrasonographically, these may have variable appearances, ranging from hypoechoic, mixed echogenicity to hyperechoic. Majority of them are often hyperechoic in nature.

Haemangioma tends to be well circumscribed. In addition, anaechoic channels are seen which represent vascular structures within. These sonographic findings correlate well with the pathologic findings of multiple vascular channels separated by fibrous septa[1].

Computed tomography demonstrates features similar to those of cavernous haemangioma where these lesions first appear as focal areas of low attenuation. Then with contrast there is early peripheral enhancement with variable amount of delayed central enhancement or diffuse enhancement. In delayed scans, multinodular tumours appear isodense with surrounding liver, while all solitary ones showed varied degrees of centripetal enhancement and persistent central cleftlike unenhanced areas[5].

Magnetic resonance imaging, haemangiomas appear inhomogenous but often hypointensity on T1WI and of varying degrees of hyperintensity on T2WI. With gadolinium - DTPA contrast, peripheral enhancement with subsequent fill-in towards the centre occurs[6].

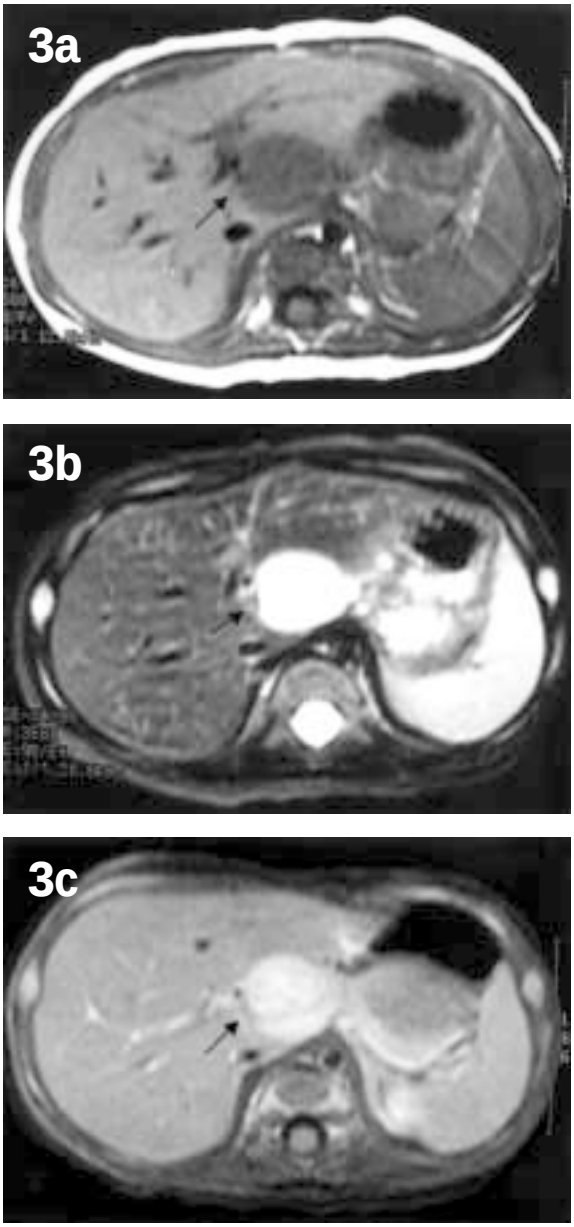

Fig.3a-c Magnetic Resonance Imaging, T1 weight image reveal a hypointense nodule (a) that is hyperintense on T2 weight fat saturation image (b). With gadolinium contrast the lesion enhances homogenously (c).

In radionuclide sulphur colloid scan, haemangiomas appear as cold defects. Technetium-99m labelled red blood cell scan can be diagnostic of haemangioma[7]. Often there is a defect in the early phases that shows prolonged and persistent 'filling in' on delayed scans. Furthermore, this can be used to monitor the progress of the haemangioma[8].

Angiography is seldom performed nowadays unless therapeutic embolisation is considered to control cardiac failure, thrombocytopenia or haemorrhage.

The major differential diagnosis of infantile hepatic haemangioma includes hepatoblastoma and metastatic neuroblastoma. Some of their sonographic features may simulate a haemangioma. In the case of hepatoblastoma, it is often associated with persistent and markedly elevated alpha-fetoprotein levels. In metastatic neuroblastoma, although the primary can be anywhere within the sympathetic neural chain but it is often located in the adrenal glands, which enables this diagnosis to be made. On occasion, histological evaluation is necessary to obtain a definite diagnosis.

Complications that can be associated if these lesions are large and include congestive cardiac failure, disseminated intravascular coagulopathy and thrombocytopenia.

Infantile haemangiomas are often treated conservatively. However, surgery is an alternative when conservative treatment fails. The range of treatment includes steroids, interferon-alpha 2a therapy, radiotherapy, ligation of the hepatic artery and hepatic resection[8-11].

Generally, all haemangiomas eventually involute and this often occurs within six to eight months. As seen in our patient, spontaneous involution occurred in two ofthe lesions while there was significant reduction in the size of the third. The prognosis is often excellent if the effects of shunting do not significantly compromise the patient[8].

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