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Case Report

# Hepatic cystic mesenchymal hamartoma

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## Introduction

Primary hepatic neoplasms represent only 0.5%–2.0% of all paediatric neoplasms.[1] The most common hepatic neoplasm in children is metastasis. Most primary liver tumours in children are malignant, but one-third are benign.[2] Mesenchymal hamartoma (MH) of the liver, though rare is the second most frequent benign liver mass in children after infantile haemangioendothelioma and is characterized by cystic hamartomatous mesenchymal proliferation.[2] This article discusses the aetiopathogenesis and pathologic features, and describes the role of imaging in the diagnosis and management of this unusual entity.

## Case report

A 10 month old male infant patient, an issue of non-consanguineous marriage and born normally at full-term, was brought to hospital with a painless right upper abdominal lump. This lump was noted over the last one month. There was no history of fever, vomiting, jaundice or haematuria. The baby's weight gain and achievement of milestones had been normal. On examination, the baby was active, playful and weighed 9.0 kg. He had a protuberant abdomen. There was hepatomegaly with a span of 13.0 cm. A firm, non-tender mass with a smooth surface was evident arising from the right lobe of the liver. There was no splenomegaly. The liver function tests and enzymes including serum alpha-fetoprotein (AFP) and gamma-glutamyl transpeptidase (GGT) were normal. All haematological and other biochemical tests were also normal. Test for echinococcal antigen was negative. Ultrasonography (USG) revealed a large well-circumscribed multicystic mass measuring 10.0 cm × 10.0 cm × 11.0 cm in the right lobe of the liver. The cysts measured 2.0 cm–5.0 cm in size. The cyst walls were 2.0 mm–4.5 mm thick with areas of irregularity; however there were no mural nodules or calcific foci. Debris was noted in the dependent part of the larger cysts (Fig. 1). A few small solid areas that were heterogeneously hypoechoic were noted between the cysts. On colour-Doppler flow imaging, there was no evidence of increased vascularity. A subsequent non-contrast and contrast-enhanced computerized tomography (CT) of the

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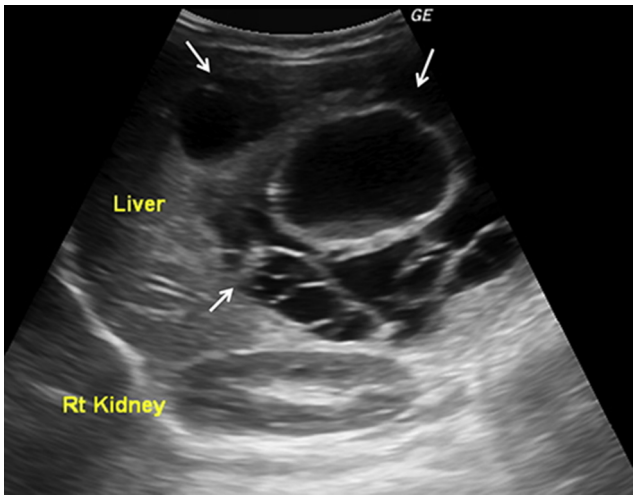


**Fig. 1 – Ultrasonographic image showing a large multiloculated cystic hepatic mass (white arrows). Cysts of variable sizes and echogenic debris noted within the large central cyst.**

abdomen revealed a well-defined multicystic mass measuring 10.0 cm × 11.0 cm × 12.0 cm involving almost the entire right hepatic lobe. The central and peripheral cysts had an attenuation of 4 Hounsfield Units (HU) and 17 HU, respectively. The cyst walls revealed contrast-enhancement. A few small solid areas of enhancement were also noted between the cysts (Fig. 2). The rest of the abdomen was normal. Based on the clinical presentation and imaging findings, a diagnosis of hepatic MH was made. The infant underwent surgical intervention in the form of marsupialization with uneventful post-surgical recovery. Biopsy from the lesion at surgery confirmed the imaging diagnosis.

## Discussion

MH is the second most common benign liver mass in children. A PubMed search revealed about 260 reported cases till 2013.

MH occurs due to uncoordinated proliferation of periportal primitive mesenchymal tissue. It had hitherto been considered a congenital lesion, attributed to factors like ductal plate malformation, bile duct obstruction, ischaemia or degeneration of Ito cells, and lymphatic duct obstruction.[2] However, cytogenetics and flow cytometry have revealed balanced translocations at 19q13.4 and aneuploidy that suggest a neoplastic process.[1,2] The mass grows along the portal tracts, compressing the adjacent parenchyma resulting in atrophy, degeneration and subsequent intralesional fluid accumulation. MH shares several histopathologic, immunohistochemical, and cytogenetic features with undifferentiated embryonal sarcoma (UES).[2,3] UES has also been reported in a background of MH.[2]

MH generally occurs in children less than 2 years of age with a male preponderance (male: female, 2:1). The right lobe of liver is more frequently involved (right: left = 6:1). It has also been reported in foetuses, older children and adults. There is no racial predilection.[2,4] The usual presentation is a large (>10 cm), progressively increasing, painless abdominal mass.[5,6] Others may present with respiratory distress, fever, vomiting, high-output cardiac failure or lower limb oedema.[1] Serum AFP and GGT may be elevated.[5,6] AFP levels are higher in patients with solid MH than cystic MH.[7] The natural history of MH is to initially enlarge and then stabilize or continue to grow. Some spontaneously regress and calcify.[8] Intranatal complications include polyhydramnios, foetal hydrops, abdominal dystocia and foetal demise.[9]

On gross pathology, MH appears as a large, well-defined, unifocal cystic mass. 85% are multilocular and composed of variable sized cysts containing clear amber fluid or gelatinous material. Others have a mixed solid-cystic or solid appearance[2,5,6] and these are immature forms of the former.[7] Solid lesions tend to occur in younger patients.[2] Histologically, MH reveals mesenchymal stellate cell proliferation in a loose mucopolysaccharide-rich matrix and hepatocyte cords surrounding vessels and bile ducts. Mesenchymal collagen fibrils separate to form pseudocysts. The bile ducts constitute the proliferating component and displace the hepatocytes peripherally; however some immature hepatocytes develop as a component of the lesion.[2,7] Necrosis, haemorrhage and calcification are rare.[2,6,10]

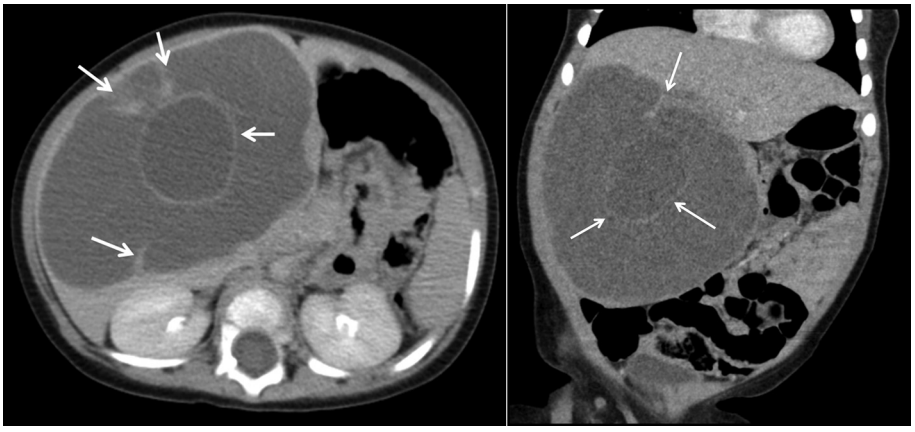

**Fig. 2 – Axial and coronal CT images show a large multicystic hepatic lesion. The attenuation of the central cyst is lower than the peripheral cysts. Contrast-enhancement is noted anteriorly and in the septae (white arrows).**