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HEPATIC MESENCHYMAL HAMARTOMA: A CASE REPORT

N JAIN[1], S SIDHU[2], K SAGGAR[3]

1. Junior Resident, Department of Radiodiagnosis, Dayanand Medical College & Hospital, Ludhiana (P.B) India.
2. Junior Resident, Department of Radiodiagnosis, Dayanand Medical College & Hospital, Ludhiana (P.B) India.
3. Professor & Head, Department of Radiodiagnosis, Dayanand Medical College & Hospital, Ludhiana (P.B) India.

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Abstract: Mesenchymal hamartoma of the liver is an uncommon entity occurring in infancy and childhood. We report a case of a 1 year 2 month old male child, who presented to our institution with abdominal distension and pain abdomen. Ultrasound and Computed tomography (CT) showed a large multiloculated hepatic lesion. It was surgically resected and histopathology revealed a mesenchymal hamartoma of liver.

Keywords: Mesenchymal, Hamartoma, liver, childhood

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Corresponding Author: DR. NIKHIL JAIN

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INTRODUCTION

A 1year 2month old male child presented with history of nausea, vomiting and abdominal distension since 1 month. Child was afebrile and blood pressure was normal. On physical examination a large non-tender abdominal mass was noted. Serum levels of AST and ALT were elevated, 650 U/L(normal range <40 U/L) and 500 U/L (normal range <56 U/L) respectively. Blood counts and renal function tests were normal.

X ray abdomen was done which showed inferior and left lateral displacement of the bowel loops likely due to a mass in the right hemiabdomen. (Fig. 1)

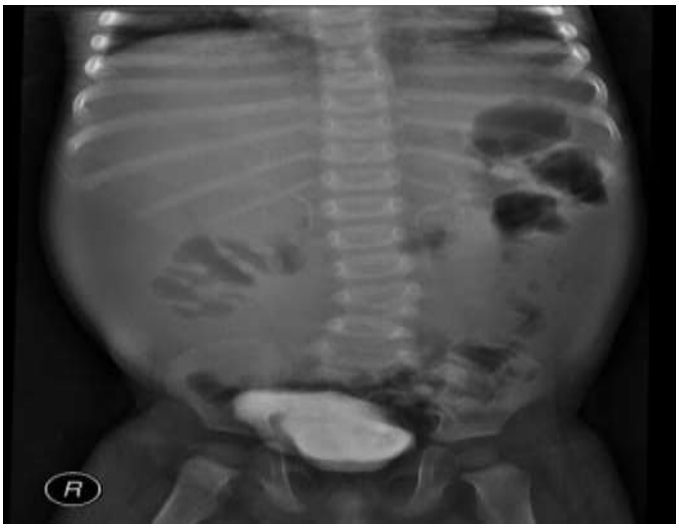


Figure 1: The abdominal radiograph shows the bowel loops displaced inferiorly and laterally to the left side like due to a mass in the right hypochondrium

Ultrasound abdomen was done . It showed a large multiseptated cystic lesion involving the right lobe of the liver. It measured approximately 12 X 11 cm. Portal vein was splayed over by the lesion. Right kidney, pancreas and gall bladder were separately visualized from the mass lesion. On colour doppler, no vascularity was demonstrated. (Fig.2)

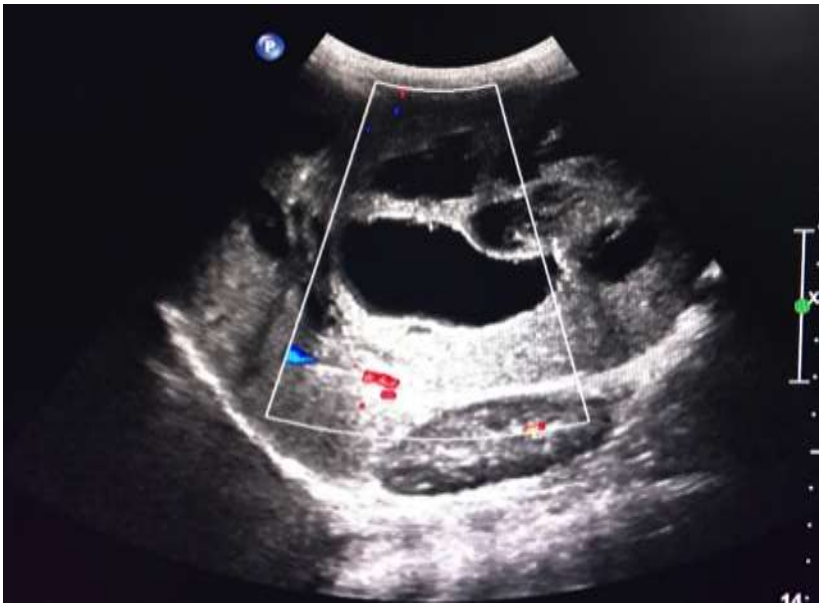
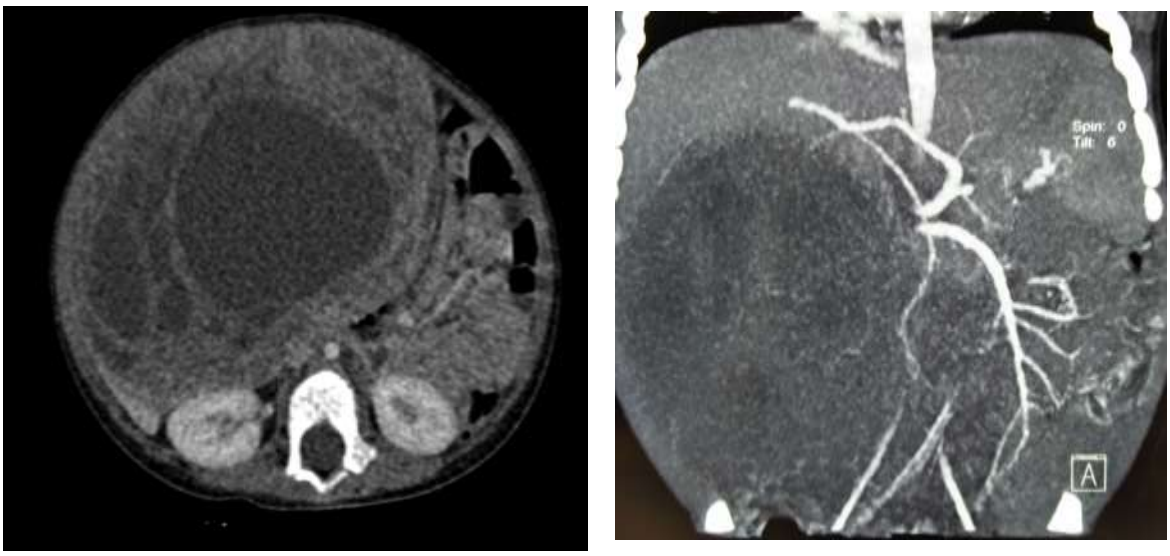


Figure 2: Ultrasound shows a large, complex cystic lesion predominantly in the right lobe of liver. It has multiple thick septations. No obvious vascuraity was noted within the lesion.

Further, a contrast enhanced computed tomography of abdomen was done (Fig 3a,b).It showed a large well defined, exophytically bulging, multiseptated cystic, encapsulated lesion arising from segment IV and V of liver. It measured approx. 10.2(AP) X 12.9(T) X 13.5(CC)cm in size. No evidence of fat component or calcification was seen within it.

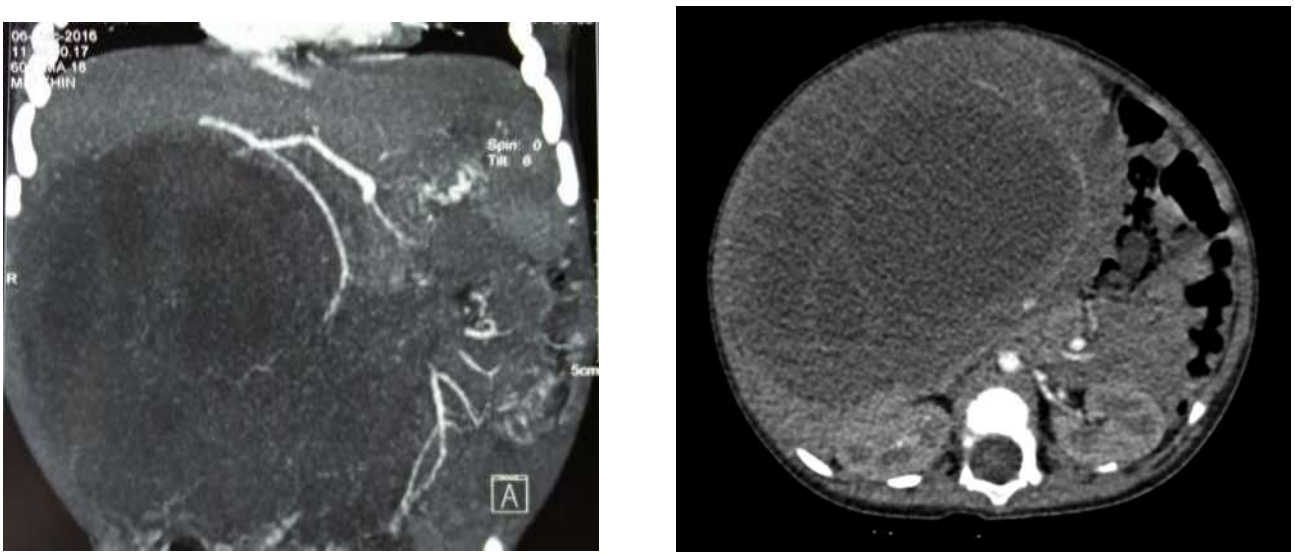
Bifurcation of portal vein was splayed by the lesion. However, no evidence of portal vein thrombosis was seen.

Branches of right hepatic artery were splayed by the lesion and the left hepatic artery was attenuated in caliber. A few branches of the anterior and posterior divisions of right hepatic artery were seen coursing through and supplying the lesion, further suggesting the hepatic origin of the lesion (Fig 4a,b). Pancreas and bowel loops were displaced towards the left by the lesion. Fat planes of the lesion with the bowel loops, colon and right kidney were well maintained.



(a) (b)

Figure 3: Arterial (a) and venous (b) phases of the contrast enhanced CT scan shows a large exophytically bulging multiseptated lesion arising likely from the liver with displacement of the pancreas and bowel loops to the left.



(a) (b)

Figure 4: MIP images of the arterial phase of contrast enhanced CT scan shows that a few branches of the anterior and posterior division of the right hepatic artery are seen coursing through and supplying the lesion further indicating the hepatic origin of the lesion.

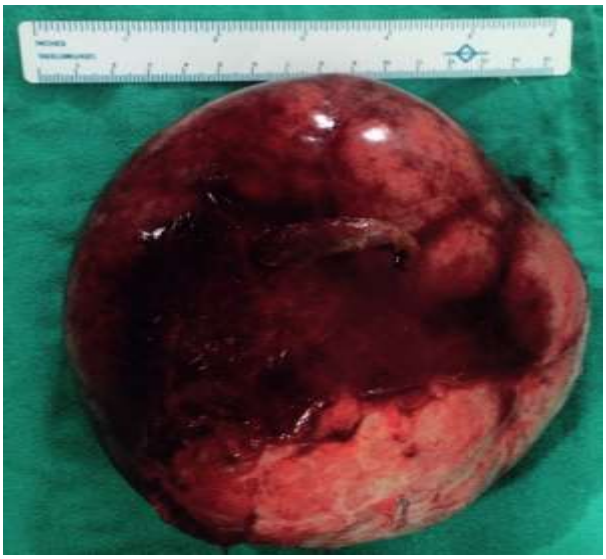


Figure 5: Image shows the surgically resected tumor which was sent for histopthological examination. It was diagnosed to be mesenchymal hamartoma.

The mass was surgically resected and the pathological study of the tissue specimen showed presence of variable sized multiple cysts lined by cuboidal epithelium surrounded by disorganized mass of mesenchymal tissue. At places there were seen entrapped cords of hepatocytes along with few primitive bile ductules. No significant atypia/pleomorphism/evidence of malignancy seen. Histology was consistent with mesenchymal hamartoma.

DISCUSSION

Mesenchymal hamartoma of the liver are uncommon benign lesions usually seen in children younger than 2 years of age. [1] There is a slight male predominance.[2] Its etiology is not well understood.,They represent a developmental abnormality rather than true neoplasm and arise from the mesenchyme of the portal tract.[3-5]. A large abdominal mass is the most common presentation.[6] Vomiting, fever, constipation, diarrhea, and weight loss may occur occasionally . Liver function tests are usually within normal limits.

The tumor may cause respiratory distress and lower extremities edema.[7] These tumors grow slowly and a rapid increase in size suggests fluid accumulation within the cyst. Most of them arise in the right lobe of the liver. [8]

Mesenchymal hamartoma are large masses with average size of 15 cm. Imaging reveals a heterogeneous appearance of these lesions. These may be predominantly solid with multiple small cysts (swiss cheese appearance) or predominantly cystic with thick septations (as in our

case). Calcification and hemorrhage are rare. In larger masses, it is difficult to make out their hepatic origin.[9]

Treatment options include complete surgical resection, enucleation and marsupilization into the abdominal cavity. Out of these, complete surgical resection is the treatment of choice (as done in this case).[10]

Differential diagnosis include hydatid cyst, enteric duplication cyst, mesenteric cyst like lymphangioma, hepatoblastoma, an abscess, cystic teratoma, infantile hemangioendothelioma and metastasis.[11][12] Calcification is usually seen in cases of hydatid cyst and cystic teratomas.[11] Lymphangiomas are generally thin walled unilocular cysts.[12][13] AFP levels are raised in hepatoblastoma.[9] Abscesses are associated with fever and deranged WBC counts. Enteric duplication cysts are attached to bowel and are result of bowel wall duplication.[12][14] Calcification, areas of hemorrhage and necrosis are common in large infantile hemangioendotheliomas. Moreover, the pattern of contrast enhancement is similar to that of hemangiomas.[3]

Hepatic metastases are common from neuroblastoma or wilms tumour however the lesions are mostly solid appearing.[15]

Malignant mesenchymoma is considered a malignant entity of mesenchymal hamartoma. Undifferentiated embryonal sarcoma can also rise from mesenchymal hamartoma.[16][17]

In conclusion, hepatic mesenchymal hamartoma is a developmental anomaly. Ultrasonography is the initial tool for investigation but characterization with CT is also needed. Complete surgical resection is the treatment of choice. No reported cases of recurrence are seen after surgical removal, to the best of our knowledge.

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