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MESENCHYMAL HAMARTOMA OF LIVER – A RARE BENINGN TUMOUR IN CHILDREN

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KEYWORDS :

INTRODUCTION :

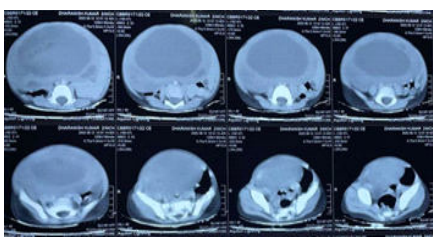
Mesenchymalhamartomas of liver is a rare primary hepatic tumour composed of a complex mixture of immature mesenchyme interspersed with small abnormal bile ducts ,aberrant blood vessels,small hepatocyte islands,stromal nodules .Apart from solid component ,which may be associated with elevated serum alphafetoprotein levels ,many tumour show extensive cystic change.The presentation of the tumour is highly variable depending upon the lesion size ranging from small asymptomatic to large tumour with life threatening complications.we present the rare occurrence of an large mesenchymalhamartoma of liver in a 2 year old children and the clinical approach and management of the case .

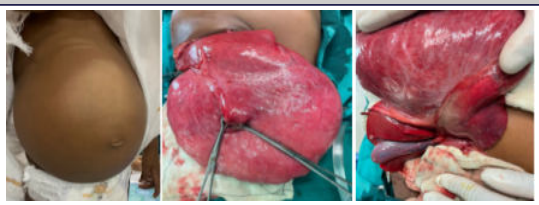
CASE REPORT :

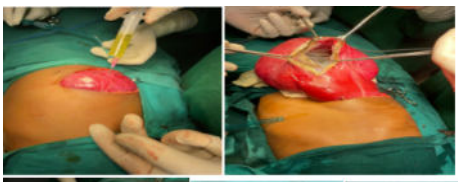
- 2 yrs Male child brought by the mother presented to the opd with abdominal distension since 1 year of age ,Initially smaller ,progressed to attain the present size .there is No h/o recent increase /rapid increase in size of the swelling . no h/o abdominal pain,constipation,jaundice ,respiratory distress.on examination a visible fullness noted in the abdomen occupying predominantly in the right hypochondrium ,epigastrium, right lumbar ,rt iliac ,umblicus ,hypogastrium size 18*15 cm , Round , Smooth surface , Moves with respiration,firm in consistency .liver was palpable 5cm below the right costal margin in the midclavicular line,non tender ,firm in consistency.the superior margin of the liver in the 5th intercostal space .haemoglobin 11.6 gm%, serum bilirubin 0.7 mg/dl, SGOT,SGPT ,PT INR were within normal limits.serum alpha protein was normal .

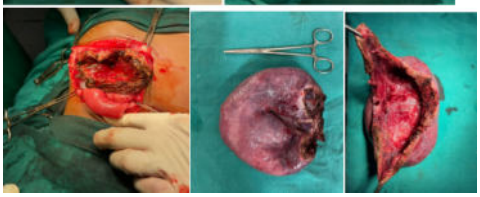
- CECT ABDOMEN - Welldefined large hypodense lesion of size 4.5*10.5*8cm noted in left lobe of liver with predominntexophytic component extending uptoopelvis displacing bowel loops laterally and IHBR dilatation in left lobe of liver .multiple septations with no solid component seen.

- The abdomen was explored through right upper transverse incision and found to have a mass of 12x8 cm in the segment VIII ,IV,II of the left lobe of liver .marsupilisation was done .postoperatively patient recovered well.on examination cut section shows thick walled multiloculated cyst with straw coloured fluid .histopathologicallyfew compressed ,tortuous and dilated bile ducts lined by cuboidal to atrophic biliary epithelium embedded in a collaginised and myxoidmesenchymalstroma composed of stellate to spindle cells.blood vessels are seen within the stroma.on immune histochemistry CD 31 and CD 34 demonstrated the blood vessels and bile duct elements.









DISCUSSION:

Mesenchymalhamartoma (MH) is an uncommonhamartomatous growth of mesenchymal tissue, bile ducts, hepatic cords and blood vessels in the liver .MH represents 5-8% of paediatric hepatic tumours and is the second most common benign hepatic tumour in childhood . Eighty percent are diagnosed before the second year of life and the remainder are detected by 5 years of age.Symptoms of MH depend on the patient's age, tumour size and growth rate. Due to its rapid increase in size, it is often misdiagnosed clinically as a malignant tumour or as a hepatic cyst because of its cystic appearance.The pathogenesis of MH is not fully understood and is still debated. There are many theories about pathogenesis of MH. Some theories point to developmental abnormalities, regional ischaemia or biliary obstruction as a possible cause . Aberrations involving the chromosomal region 19q13.4 are probably very important in pathogenesis and have been described in many reported cases with cytogenetic analyses . Translocations t(11;19) (q13;q13.4), t(11;19)(q13;q13.3), t(15;19)(q15;q13.4) have been reported, as well as interstitial deletion del(q13.1q13,4) and complex rearrangements involving 11q2, 17p11, 19q13.3 [19-23].Conventional radiograph Although nonspecific, radiographs may show a large, non-calcified mass in the right upper quadrant . Ultrasound - It usually appears as a multiseptated cystic lesion interspersed with solid components. Detection is difficult for pedunculated lesions. In some lesions may be the predominance of solid structures . CT- On unenhanced CT, it usually has a heterogeneous appearance. The stromal elements often appear hypoattenuating, whereas the cystic components have water attenuation . The appearance of cystic and solid portions has been likened to Swiss cheese.

On a postcontrast CT scan, solid portions or thick septa of the tumours can show heterogeneous enhancement .MRI prominent cystic components multifocality is uncommon MR imaging appearance of

mesenchymalhamartoma can also vary depending on the presence of stromal elements as well as the protein content of the fluid . Angiography (DSA) While not being a standard diagnostic imaging modality of choice, angiography may show peripheral hypervascularity to the lesion with a septated avascular centre .

MH is a benign tumour and many authors recommend complete resection of the tumour as sufficient therapy [28]. After incomplete resection recurrent lesions are also benign.Unroofing ,Marsupilization ,sclerotherapy , Laproscopic fenestration are the other treatment modalities for mesenchymalhamartoma of liver . Differential diagnosis simple liver cyst , duplication cyst , choledochal cyst ,hepatic abscess , hepatoblastoma: often predominance of the solid component and persistently elevated or rising AFP undifferentiated (embryonal) sarcoma of the liver: older age at presentation (6-10 years)infantile haemangioendothelioma of the liver: associated with midaortic syndrome .

Conclusion :

Hepatic mesenchymalhamartomas are rare conditions primarily seen in the pediatric population .complete surgical resection or marsupilisation of the tumour is preferred to prevent the complication .histopathological diagnosis helps in cionfirmation and specificity of mesenchymalhamartoma of liver .

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