



Case Report

Mesenchymal Hamartoma of Liver-A Rare Case Report

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ABSTRACT

Hepatic mesenchymal hamartoma(HMH) is a rare liver tumor in childhood. Fewer than 200 cases have been reported. We present a case of a 1 year old male infant, who was admitted in our institution with the history of abdominal distension. Computed tomography(CT) showed a 12-cm multiloculated mass in the right lobe of liver. The patient underwent surgical enucleation of the tumor. The pathological report confirmed a benign cystic mesenchymal hamartoma of the liver. We present this case for its rarity.

KEYWORDS: Mesenchymal hamartoma, Liver tumor, Cystic.

INTRODUCTION

Mesenchymal hamartoma of liver is a benign tumor of infancy characterized by admixture of epithelial structures in a loose connective stroma with fluid accumulation suggestive of lymphangiomatous channels[1]. It is a rare tumor of infancy and eighty percent of the tumors are found within the first 2 years of life and the remainder are detected by 5 years of age[1,2,3]. Rare cases have been reported in adults[4]. Approximately 140 cases have been reported[2]. First case was reported in 1903[4].

MHL may affect either lobe of the liver and presents as a cystic or solid mass, and on occasion both components may be present. Alkaline phosphatase, β -human chorionic gonadotropin, serum transaminases and α -fetoprotein are usually within normal limits in patients with MHL. Hematoxylin and eosin staining as well as immunohistochemical studies have described MHL as having spindle cells positive for vimentin and smooth muscle actin and negative for CD31, CD34 and S100 proteins, while the ducts stain positive for cytokeratin 7 and negative for cytokeratin 20[2,3].

CASE REPORT

A one year old male infant was referred to our hospital for abdominal distension that was noticed incidentally by his

mother since one week. His vitals were stable and no other findings were made out on general examination. Investigations were done and USG revealed a 105×75 mm multi-septated cystic lesion in the right lobe of the liver. CT scan showed a large well encapsulated cystic lesion with enhancing wall and multiple septae in the right hepatic lobe. Excision was done and sent for histopathological examination.

Gross examination revealed a single gray brown cystic mass measuring 9.5×7.5×5cms. On cut section, multiloculated cyst filled with gelatinous, mucinous material were seen along with foci of haemorrhage (Figure 1).Imprint cytology from the fluid showed occasional strips of fibrous connective tissue and tiny clusters of hepatocytes are seen in the background of several scattered lymphocytes. On microscopic examination, the cystic mass lesion showed multiple cysts with myxomatous connective tissue containing scattered bland stellate-shaped mesenchymal cells and branching bile ducts surrounded by loose mesenchyme itself. Some cysts were devoid of epithelial lining and surrounded by loose mesenchymal tissue. Remaining cysts were lined by cuboidal epithelium (Figure 2). Groups of hepatocytes were seen focally along with few dilated and congested vascular spaces admixed with muscle fibres (Figure 3).

Figure 1: Multiloculated cysts filled with gelatinous material



Figure 1: Multiple cysts with myxomatous connective tissue containing scattered bland stellate-shaped mesenchymal cells and branching bile ducts

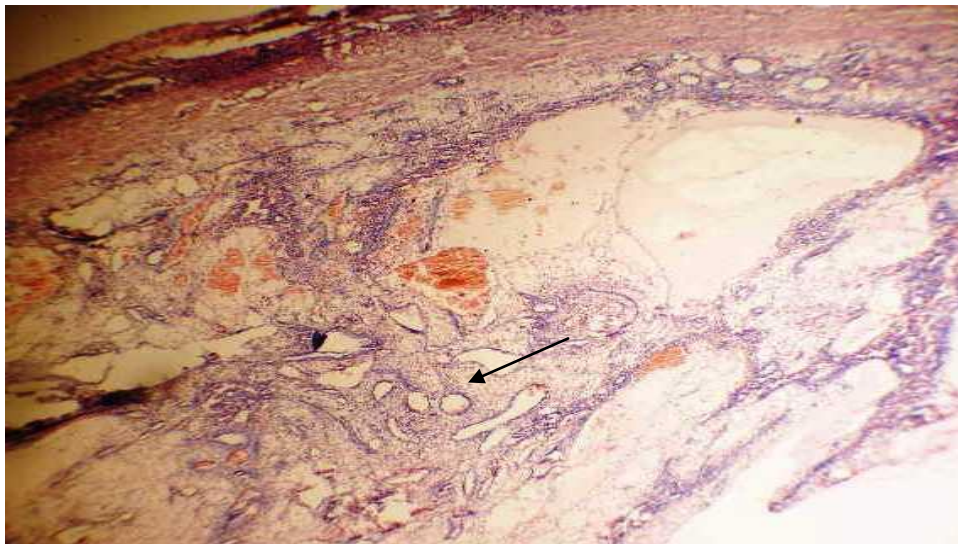
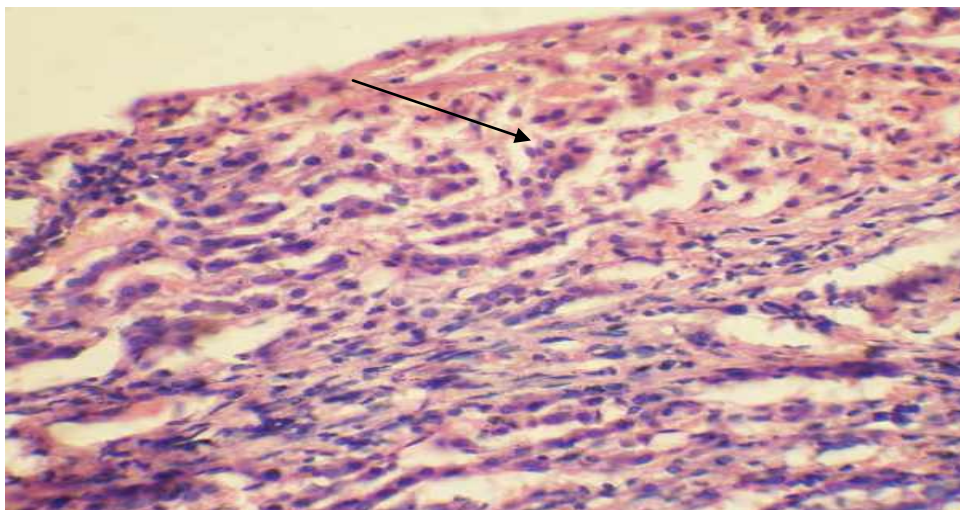


Figure 2: Bland stellate-shaped mesenchymal cells along with hepatocytes.



DISCUSSION

Mesenchymal hamartoma of liver is benign tumor that occurs exclusively during infancy and childhood although few cases in older age groups have been reported[5]. Majority of the cases present at mean age of 16 months, the range being from newborn to 5 years. The origin is mostly from right lobe of the liver[2]. Most of the cases remain asymptomatic while the others are detected incidentally when they present with right upper quadrant mass, respiratory distress, fever and raised right hemidiaphragm[2,3]. Liver function tests usually remain within normal limits. The most helpful diagnostic tests are ultrasono-graphy, computerised tomography and histopathology[3]. On CT scan, it presents as a complex mass containing areas of low attenuation separated by solid septae and stroma which enhance with intravenous contrast administration[6].

Benign liver tumors in children may be divided into two major groups: those of epithelial derivation, including simple cysts, focal nodular hyperplasias and adenomas and those of mesenchymal derivation including hamartomas and hemangiomas. Benign mesenchymal tumors of liver are more common than their epithelial counterparts[2]. The hamartoma is characterized by marked overgrowth of mesenchymal or connective tissue of varying maturity and marked tendency to cyst formation. The mesenchyme may contain minor remnants of liver cells, duct cells and even portal tissue, the most characteristic feature being overgrowth of connective tissue. The lobular architecture is maintained. The cysts may be without lining or may be lined with flattened cells thought to be mesothelial cells[7].

Comparison with adult HML:

Since adult MHL can also occur rarely, it has to be differentiated with pediatric MHL. Radiographic appearance of MHL represents a continuum from predominantly cystic to mostly solid tumors[8]. In pediatric patients the literature is unclear in regards to the cystic or solid nature of MHL. Early studies by Stocker and Ishak[9] describe nearly all pediatric cases as having cysts, however Chau et al. reported that children more commonly have solid lesions[10]. Interestingly, Chau et al.[10] have also reported that adults have predominantly cystic lesions while Hernandez et al. have published that both pediatric and adult cases are primarily cystic[5]. In the adult population, when solely cystic or solid tumors are present, females were more likely to have a cystic tumor, while males showed no predilection. When both cystic and solid portions of MHL were encountered, females were predominantly affected[11].

Location of the tumor in the liver appears to be different between pediatric and adult patients. MHL in children is primarily localized to the right lobe, while in adults, 12 cases (40%) were localized to the left lobe, 12 (40%) to the right lobe and in 6 (20%) tumors crossed into both lobes[10]. Interestingly, adult males have a predilection for the right lobe of the liver, while females primarily have MHL in the left lobe. In situations when MHL was found in both lobes, females were more commonly affected than males. Hepatic mesenchymal hamartomas are best treated by complete excision. Progressive abdominal distension may be fatal in untreated infants[6].

Excision may be by conventional hepatic resection or by nonanatomical excision with a small margin of normal liver pedunculated lesions are amenable to laparoscopic resection[12].

CONCLUSION

Mesenchymal hamartoma of the liver may be found incidentally on physical examination or imaging, but typically it presents with abdominal distension and/or an upper abdominal mass. Laboratory results are noncontributory and radiographic imaging is variable and inconclusive. Thus MHL with its indolent behavior, carries an excellent prognosis and therefore it should be diagnosed correctly to avoid over treatment. Proper imaging interpretation, histologic typing & optimal surgical planning is essential for its detection & treatment.

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