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Pediatric Surgery

A GIANT PEDIATRIC HEPATIC MESENCHYMAL HAMARTOMA: CASE REPORT

Dr. Apurva Arora*	Department Of Pediatric Surgery, Sri Ramachandra University, Chennai *Corresponding Author
Dr. Prakash Agarwal	Department Of Pediatric Surgery, Sri Ramachandra University, Chennai
Dr. Madhu Ramasundaram	Department Of Pediatric Surgery, Sri Ramachandra University, Chennai
Dr. S Jegadeesh	Department Of Pediatric Surgery, Sri Ramachandra University, Chennai
Dr. Selvapriya	Department Of Pediatric Surgery, Sri Ramachandra University, Chennai

ABSTRACT Mesenchymal hamartoma of the liver is the second most common benign liver tumor in children, yet its etiopathogenesis are poorly understood. Typically, it presents as a large benign multicystic liver mass in children younger than three years, amenable to complete resection. Most tumors gradually increase in size, some reaching enormous proportions, some undergo incomplete spontaneous regression, and rarely, few have shown malignant transformation to undifferentiated (embryonal) sarcoma. Here, we report a 10 month-old male child who presented with abdominal distension and respiratory distress. Ultrasonography and Computed Tomography (CT) of the abdomen were suggestive of a Mesenchymal Hamartoma of the liver. The patient was managed with a Right Hepatectomy.

KEYWORDS :

Introduction

Hepatic Mesenchymal Hamartoma (HMH) is an uncommon benign tumor of childhood. It makes up approximately 8% of all pediatric liver tumors. Eighty percent are found within the first 2 years of life and the remainder are detected by 5 years of age [1,2] with anecdoctal case reports in adults.

Hepatic mesenchymal hamartoma is a hamartomatous growth of mesenchymal tissue in the liver of uncertain etiology. It is a space-occupying lesion that can potentially compress adjacent organs resulting in various complications. Rarely it can be fatal due to rupture or congestive heart failure (CHF). Hepatic mesenchymal hamartoma is characterized by proliferation of variably myxomatous mesenchyme and malformed bile ducts. The differential diagnosis includes other pediatric hepatic masses like Hepatoblastoma, Infantile Hepatic Hemangioendothelioma (IHH). The diagnosis is typically made during infancy, and complete resection is invariably curative.

Case report

A 10-months-old male child presented with gradual distention of the abdomen over a period of 25 days with fever, cough, and respiratory distress since four days. On examination patient was afebrile and anicteric. There was visible fullness in the right upper quadrant of the abdomen. There was gross nontender firm hepatomegaly. Serum bilirubin, SGPT, SGOT were normal, Hb – 7.4 gm/dl, Serum Alfa-fetoprotein (AFP) - 722 ng/ml (normal range - 0 to 13.4 ng/ml). Bleeding time, clotting time, Prothrombin time and platelet count were also normal.

An ultrasound of the abdomen revealed a large 10.5 × 9 × 8 cm, sized solid and cystic mass occupying almost the entire right lobe of the liver. There were multiple internal septations within. No portal or retroperitoneal lymphadenopathy was seen nor were there any ascites. A plain and contrast-enhanced CT scan of the abdomen showed a large well-defined cystic mass with few internal septations in the right lobe of the liver, measuring approximately, 10 × 8 × 10 cm, occupying segments V, VI, and VII. There was no calcification, nor was there any soft tissue component within. The mass was seen to displace the middle hepatic vein, the right hepatic vein was not visualized and the left hepatic vein was normal [Figure 1].

Patient underwent a Trucut biopsy under USG guidance of the mass before excision to rule out Hepatoblastoma (due to elevated AFP). Histopathology confirmed it to be a Mesenchymal hamartoma. The patient underwent Right Hepatectomy. [Figure 2] The mass was excised completely with a healthy wedge of segment VIII of the liver [Figure 3]. Postoperatively, the patient recovered well. On examination, the cut section of the specimen showed areas of multiple, thin-walled, cysts containing straw-colored gelatinous material. [Figure 4] On Histopathological examination, sheets of stromal spindle cells, dilated cystic bile duct spaces, large cystic spaces and thickened veins were seen [Figure 5]. The cells were seen embedded within it were bile ducts and hepatocytes.

Patient recovered well and was discharged without any post-operative complications. The child has completed one year of follow up and the size of left lobe of liver on sequential ultrasonography screening is increasing with age and LFT remains normal.

Discussion

Mesenchymal hamartoma (MH) usually presents as a painless right upper quadrant abdominal mass in children younger than 2 years. [3,4,5] Some patients may have evidence of CHF at diagnosis. Similar to Infantile Hepatic Hemagioendothelioma (IHH), MH can be diagnosed prenatally. [6,7] Edmondson proposed that MH arises from a mesenchymal rest that becomes isolated from the normal portal triad architecture and differentiates independently. [8] The tumor grows along bile ducts and may incorporate normal liver tissue. Because the blood vessels and bile ducts are components of the mesenchymal rest, the biological behavior of the tumor varies with the relative predominance of these tissues within the loose connective tissue stroma (mesenchyme) that surrounds them. Thus, the tumor may present as a predominantly cystic structure that enlarges rapidly because of fluid accumulation, or it may be predominantly vascular and present with CHF.

Von Schweinitz et al suggested that fat-storing (Ito) cells of the immature liver may be involved in the development of MH. [9]

Serum AFP levels are usually normal in children with MH, but they may be mildly elevated. The radiographic features of these tumors are consistent and distinguishing; abdominal ultrasonography and CT demonstrate a single, usually large fluid-filled mass with fine internal septations and no calcifications. [10,11,12]

Management must be tempered by the understanding that MH usually follows a benign course, although there have been reports of malignant transformation. In general, complete operative resection is the procedure of choice, if it can be accomplished safely. Huge lesions or those that involve both lobes may be treated by unroofing and

marsupializing the cysts, although the lesion may recur after incomplete resection. [13,14,15]

In summary, mesenchymal hamartoma is one cause of a cystic liver mass in pediatric age group; the lesion is benign. Management depends on the location of the lesion and assessment of resectability as depicted by the imaging. We are reporting this case due to its rarity.

Figure :1

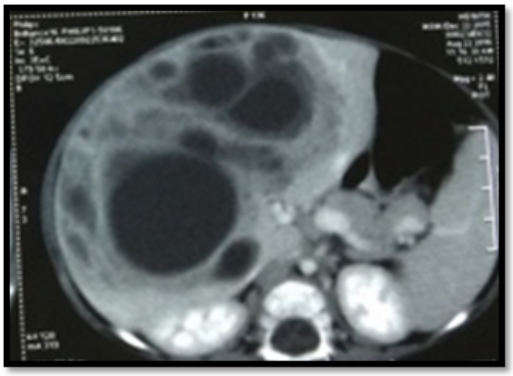


Figure 2

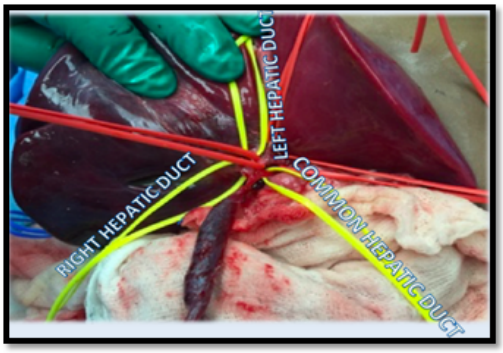


Figure 3

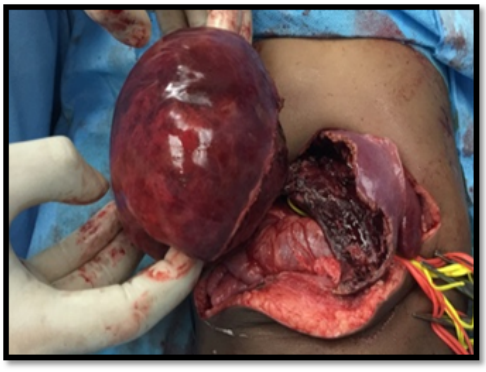


Figure 4

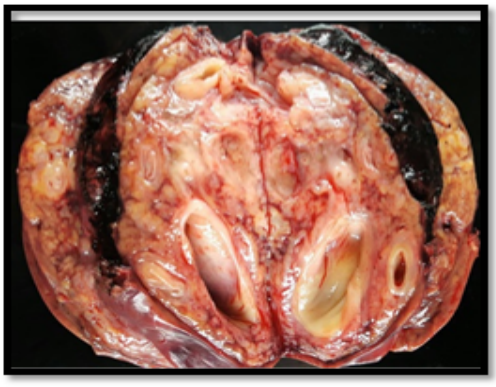
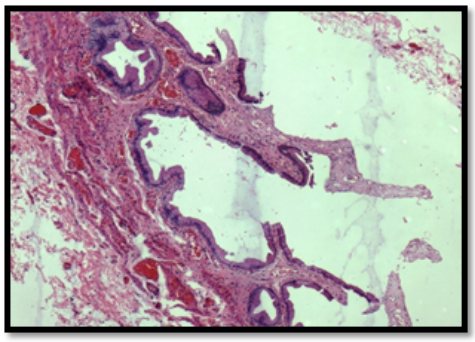


Figure 5



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