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Case Report

Mesenchymal Hamartoma – A Case Report

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Abstract
We report a thirteen-month-old female with history of abdominal distension for two months. She presented to the outpatient pediatric department having pallor, massive abdominal distension, and hepatomegaly. She was initially diagnosed with liver abscess and admitted to pediatric ward. All baseline labs including liver aspirate culture and staining did not reveal any infectious etiology. Patient responded initially to empirical antibiotics, but abdominal distension continued to increase during admission. Patient was referred to gastroenterology where the investigations showed solid cystic liver mass. Patient was operated for mass excision and biopsy revealed mesenchymal hamartoma.

Keywords: Abdominal mass, Mesenchymal hamartoma, Abdominal cystic mass, Mesothelioma

Introduction
Mesenchymal tumors of the liver are a common occurrence in the pediatric age group and are third in order following Hepatoblastoma and hepatocellular Carcinoma. Mesenchymal Hamartoma (MH) is the second most common benign liver tumor (next to Hemangioma) in the age group of 5–10 yrs and constitutes 30% of benign liver tumors and 5–8% of all the liver tumors in children. Fewer than 200 cases are reported in the literature. Several options for the management of these tumors including watchful waiting, enucleation, marsupialization, and liver resection are described. We propose that complete excision of the tumor is safe and provides the best long-term results.

Case Report
A thirteen-month-old female patient presented with abdominal distension for a month. She was in usual state of health a month prior to presentation, when she developed rapidly progressive abdominal distension not associated with any aggravating or relieving factor. There was no history of body swellings at other sites, diarrhea, vomiting, constipation, pain abdomen, jaundice, acholic stools, itching and fever. No urinary symptoms altered conscious level or fits were reported. Mother also noticed pallor but there was no history of bruising, rash or bleeding from any site.

Workup was done on outpatient basis including ultrasound and CT Abdomen (Figure1) which showed provisional diagnosis of hepatic abscess. Patient was admitted in children ward of a tertiary care hospital for eight days where she received antibiotics and supportive care and managed as a case of liver abscess. During the stay, she further developed rapid progression of abdominal distension and ultrasound guided liver drainage was done. 1000 ml of fluid was drained, which was dark yellow in color; 20ml was sent for analysis(Figure 2). One packed cell transfusion was given due to low hemoglobin.

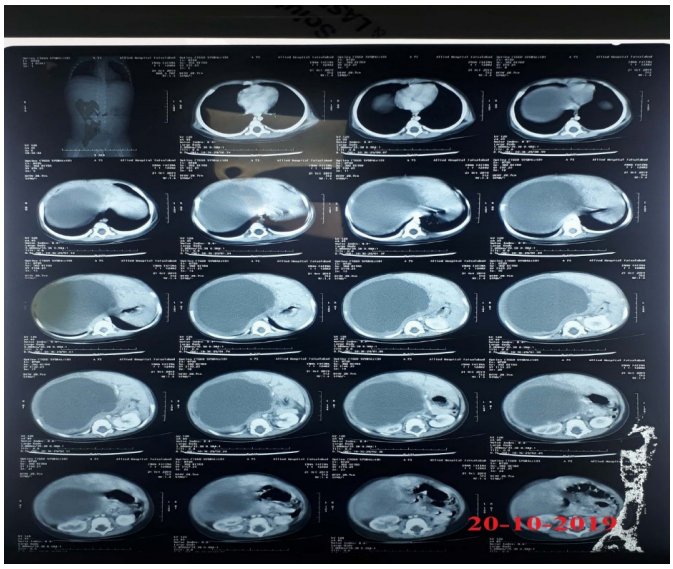

Figure 1: A fluid filled mass in liver.

Patient was referred to Pediatric Gastroenterology and Hepatology ward. During the stay Intravenous antibiotics were given and further workup, including MRCP (Magnetic Resonance Cholangiopancreatography)(Figure 3) and MRI were done(Figure 4).

Baseline labs showed high total leukocyte count in the beginning which became normal after antibiotics. Electrolytes, renal function, liver function tests and coagulation profile were normal. Antibodies for Echinococcus were negative. Indirect hemagglutination test for amoebiasis was negative. Fluid aspirate from liver showed proteinaceous background against few inflammatory cells. Gram staining, ZiehlNeelsen staining and culture sensitivity of liver aspirate did not reveal any organism. Ultrasound abdomen revealedenlarged liver measuring15

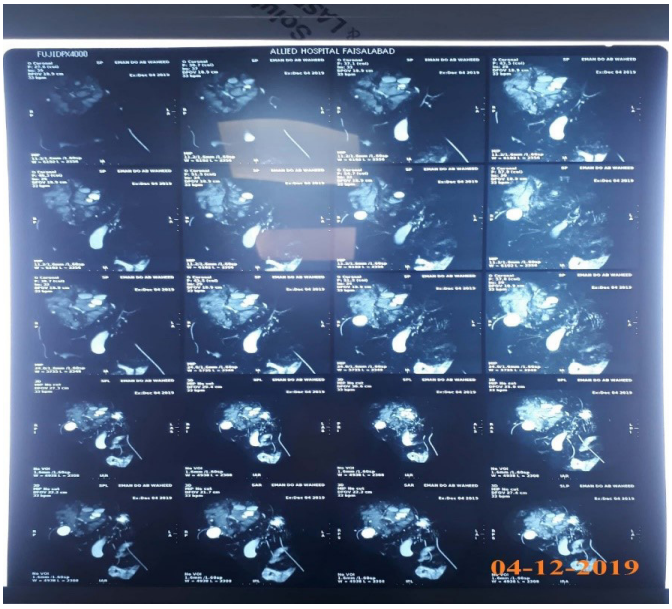

Figure 2: MRCP.

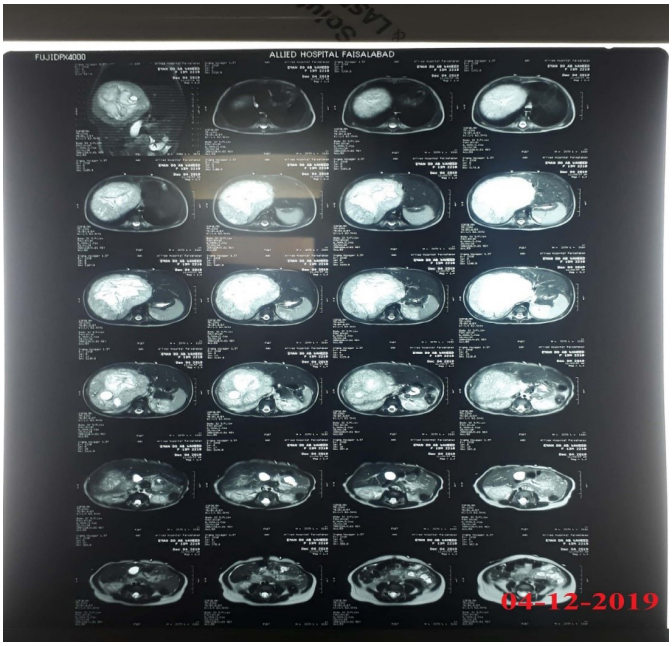

Figure 3: Solid cystic mass in liver.

Figure 4: Fluid drained before pigtail catheter (1000ml).

cm, large fluid collection of about 500-600ml having internal debris and well-defined wall present in segment V, VI and VII of the liver. No solid mass seen. MRI abdomen showed mixed solid cystic lesion in right lobe of the liver with internal hyper-intense T2W signals.

Abscess was drained after consultation with pediatric surgeon, with the help of a pigtail catheter, which was kept in place. Daily fluid output was about 200ml /24 hours, which initially was bile stained (and tested positive for bilirubin on dipstick). Later, it was bile free, with total amount being reduced to 150ml/24 hour. Abdominal distension has slightly improved over the course of time with no other symptoms. Patient underwent surgery for excision of solid cystic mass in right lobe of liver. Biopsy of specimen turned out to be Mesenchymal Hamartoma.

Post-surgical course was uneventful. Ultrasound after surgical excision showed no mass in liver. Child is following up in gastroenterology clinic and is thriving well.

Discussion

Mesenchymal Hamartoma is considered a ductal plate malformation, wherein mesenchymal rests become isolated from the normal portal triad architecture and differentiate independently [3,4]. The biologic behavior of these tumors varies with the relative predominance of blood vessels and bile ducts within the loose connective tissue stroma (mesenchymal) that surrounds them. Other theories gaining acceptance are regional ischemia, toxic-metabolic, and a true neoplastic etiology with most cytogenetic analysis pointing to chromosomal abnormalities involving the region 19q13.4 [1,4].

Grossly, mesenchymal hamartoma is a well circumscribed, unencapsulated mass that can be very large with a soft, myxoid, and cystic cut surface. The tumor presents as a cystic structure and enlarges rapidly because of fluid accumulation. Microscopically, there is a mixture of epithelial and stromal components. The epithelial component consists mainly of tortuous and dilated bile duct elements. Cystic changes in the bile ducts could present as ectasia, pseudocysts, or lymphangiomatous malformation [5]. The stromal component is formed of spindle cells in a background that ranges from myxoid in about 50% and collagenous or hyalinized in 47% [5]. As a minor component of MH, vascular proliferation and hepatocytes were identified. Vessels consisted of small to medium sized veins or capillaries. Hepatocytes were found in cords, islands, or lobules. The hepatocytes were largely located in the periphery of the hamartoma. However, in few cases, they were widely distributed within the entire hamartomas. Transition between hepatocytes and bile ducts was also identified focally in about a third of the cases [5].

About three-quarters of MH occur in the right lobe of the liver. The rest are found in the left lobe or involve both lobes. Up to 20% of MH are pedunculated, arising from the inferior surface of the liver. Painless RUQ mass noticed by the mother or an incidental imaging finding is the usual presentation. Mechanical compression of adjacent viscera causing pain, vomiting, jaundice, or poor weight gain is described. There is no tumor marker (AFP, β-HCG) or liver function test that is specific for a diagnosis of MH. Normal laboratory parameters are useful in excluding other diagnoses. The usual imaging finding is a large, well-defined, heterogeneous solitary mass containing

cysts of varying sizes, ranging from a few millimeters to more than 15 cm. The characteristic ultrasonography findings are multiple echogenic cysts with thin septa. On a noncontrastCT the stromal elements appear hypoattenuating and the cystic components are of near water attenuation. The mesenchymal component enhances with contrast administration [1,2]. MR imaging appearance of mesenchymal hamartoma depends on the relative cystic versus stromal components. Solid areas may appear hypointense to adjacent liver both on T1- and T2-w images owing to fibrosis. The cystic areas are generally close to water signal intensity on T2-weighted images and demonstrate variable signal intensity on T1-weighted images, depending on the protein content of the cyst fluid. After intravenous administration of gadolinium, enhancement is mild and limited to the septa and stromal components [1,2].

The current standard of care of these tumors is complete resection with clear margins. A formal hepatic resection or nonanatomical resection could accomplish these goals. Besides symptoms a strong indication for resection of all these tumors is a well proven association with Undifferentiated Embryonal Sarcoma (UES), an aggressive liver tumor with a median survival of less than 1.5 years [1,6,7]. UES after incomplete excision of MH, coexistence of the two entities in the same tumor and similar features on gross pathology, immunohistochemistry, and cytogenetics suggest a strong association between the two and the plausible theory that MH can degenerate into UES [6,7]. If deemed unresectable orthotropic liver transplantation or LDLT should be considered for a long term survival [8].

The practice of watchful waiting, enucleation, and marsupialization is strongly discouraged for the above-mentioned reasons [1,6,7,9]. If a nonoperative management is chosen a close surveillance protocol including clinical examination and imaging (USG) is the best possible strategy to identify any malignant conversion at the earliest.

The overall mortality after liver resection for primary liver malignancies in children is 3.7% [10]. In series that specifically reported outcomes after resection for MH, the mortality rate is higher [11,12]. Mortality is mainly from an acute event like blood loss and air embolism and from unrelated causes. Morbidity rates tend to be substantial at 30–35% [1,10,12]. Major morbidity stems from bile leaks and bile duct strictures, postoperative infections, and incisional hernias. Most morbidity is treatable and is with no long term consequences. Operations performed at high volume centres tend to have lower mortality and morbidity rates [8].

Parental involvement in treatment decisions, heightened awareness of malignant transformation, and an expert surgical management are essential elements of high value care in treating these tumors.

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