



Mesenchymal Hamartoma of Liver Mimicking Hepatoblastoma, A Case Report

Aqeel Ahmad^{1*}, Rabiaa Mahmood Ali², Zainab Waseem³, Urfa Mudasar⁴ and Muhammad Atiq⁵

¹Postgraduate Resident, Mukhtar A. Sheikh Hospital, Multan,, Pakistan

²Consultant Radiologist, Department of Diagnostic Radiology, Mukhtar A. Sheikh Hospital, Multan, Pakistan

³Consultant Histopathology, Department of Pathology and Laboratory, Mukhtar A. Sheikh Hospital, Multan, Pakistan

⁴Consultant Radiologist, Department of Diagnostic Radiology, Mukhtar A. Sheikh Hospital, Multan, Pakistan

⁵Consultant Hepatobiliary Pancreatic & Liver Transplant surgeon, Mukhtar A. Sheikh Hospital, Multan, Pakistan

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***Corresponding author:** Aqeel Ahmad, Department of Surgery, Mukhtar A. Sheikh Hospital, Multan, Pakistan

Abstract

Mesenchymal hamartoma is a benign tumor with predominant pediatric epidemiology, and an absence of association with tumor markers. The clinical association of mesenchymal hamartoma with raised serum Alpha-Fetoprotein levels is exceptionally rare [1]. We report a case of mesenchymal hamartoma in a 13-month-old boy, who presented with abdominal distension and hepatomegaly, and had a significant medical history of Hepatitis C positivity. Initial workup showed significantly elevated serum AFP levels (807.9 ng/ml), raised Gamma Glutamyl Transferase (184 U/L). Remaining liver function tests were within normal range. Triphasic CT scan showed a heterogeneous mass with cystic degeneration in segments V and VI of right lobe of liver. Non anatomical resection of segment V and VI with adequate hepatic parenchymal margins of the tumor was performed and sample sent for histopathology. Microscopy confirmed mesenchymal hamartoma. Post-surgery Alpha Fetoprotein Levels were 35.05 ng/ml.

Keywords: Mesenchymal hamartoma; Hepatoblastoma; Alpha Fetoprotein

Introduction

Mesenchymal Hamartoma is currently classified under "Mesenchymal Tumors of Uncertain Cell of Origin/Differentiation" in WHO Classification of Digestive System Tumors, 6th edition [2]. Prognostically, the tumor is considered benign till date, with adequate surgical excision as the treatment of choice. While most cases arise sporadically, a subset are associated with C19MC, Beckwith Wiedemann syndrome and Placental Mesenchymal Dysplasia [3] with emerging evidence of its relation to DICER1 syndrome. Since microRNAs of C19MC are implicated in hepatocyte maturation and differentiation, it is currently postulated that Mesenchymal Hamartomas arise through developmental abnormalities in phases of late embryogenesis.

Mesenchymal Hamartomas are exclusive to liver, with 75% arising in right lobe. These comprise the third commonest hepatic pediatric tumor, following hepatoblastoma and infantile hemangioma. The radiological features are reported as variable,

with a multicystic mass being the most common presentation. However, a solid component is frequently reported [4]. Similarly, clinical presentation ranges from asymptomatic to gross abdominal distension. Large Mesenchymal Hamartomas pose a comorbidity of blood circulation compromise and respiratory distress. Here we present a case of 13 month old baby boy, who underwent abdominal surgery for a liver mass suspected to be hepatoblastoma due to raised Alpha-Fetoprotein Levels, which was microscopically confirmed as Mesenchymal Hamartoma.

Case Report

A 13 months old baby boy, who was a known case of hepatitis C, presented to outpatient department of Hepatobiliary and Pancreatic Department, Mukhtar A Sheikh Hospital, Multan, Pakistan, with complaints of abdominal distension for 3 months, in May 2025. His parents gave a history of abdominal surgery 3 months prior to this visit, where he was operated on clinical

suspicion of Hydatid Liver cyst; no biopsy or organ removal was performed during that surgery. Current physical examination revealed a protuberant abdomen, dull percussion note, pallor and hepatomegaly (liver was palpable up to right iliac fossa). Initial workup including complete blood count, renal function tests, liver function tests and urine analysis were normal. Upon further investigations, serum alpha fetoprotein levels were markedly raised (807.9 ng/ml). His HCV RNA by PCR was not detected. Triphasic CT Scan showed a single large hepatic mass arising from right lobe of liver with heterogeneous enhancement cystic/necrotic areas. Rest of the liver showed normal parenchyma and no other evidence of mass and liver cirrhosis. Radiologic findings were suggestive of hepatoblastoma as serum AFP levels were also raised, recommending hepatic biopsy.

A Non anatomical resection of segment V and VI with adequate hepatic parenchymal margins of tumor was performed.

Intra operative findings included a huge solid and cystic liver tumor arising exophytically from segment V and VI with 600 ml of ascitic fluid present, but rest of liver was normal. There was no other mass detected in the liver (Figure 1a,1b). Post op course remains uneventful. The patient was discharged, with close post op follow up and referred to peads hepatologist for HCV management. Gross findings (Figures 2a,2b,2c) showed an intact liver capsule. On serial slicing a large solid cystic tumor was identified measuring 145x120x85mm with areas of hemorrhage and cystic degeneration. The tumor was well delineated from the surroundings. Tumor was present at the distance of 1mm from the capsule and 10mm from the nearest parenchymal resection margin. Histopathology (Figures 3a, 3b) showed a tumor composed of lobules of loose myxoid connective tissue and angulated bile ducts. The bile ducts showed a variable luminal diameter. There was no evidence of cirrhosis in the background.



Figure 1 (a, b): Intraoperative.

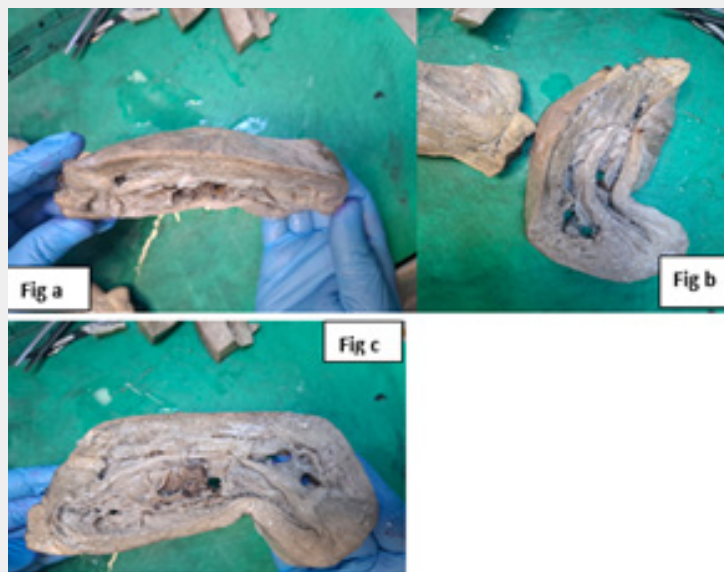


Figure 2 (a, b and C): On Gross, after fixation for 48 hours in 10% pictures were taken and finding shows a predominantly cystic cut surface, with very focal solid areas. Tumor is well delineated from the surrounding parenchyma.

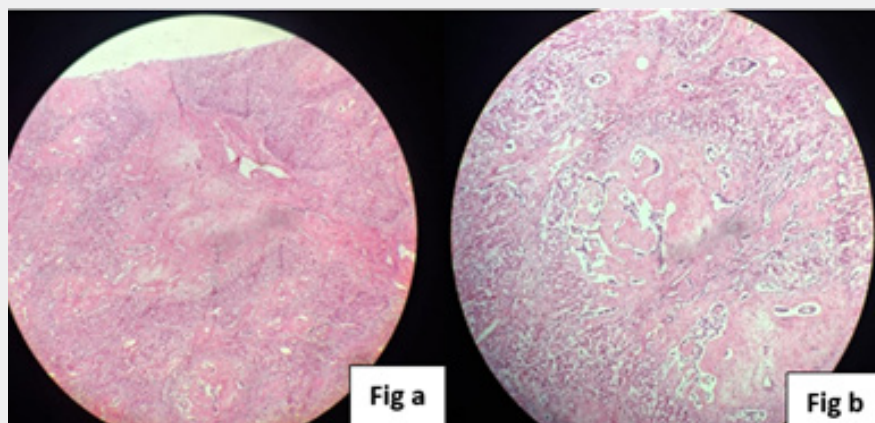


Figure 3 (a, b): Showing lobules of myxoid connective tissue with angulated bile ducts.

Discussion

As per WHO Classification of Digestive System Tumors, 6th edition, Mesenchymal Hamartomas comprise 12% of all hepatic tumors in children younger than the age of 2 years. This fits epidemiologically into the clinical presentation of our case report (13 month old). Although slight elevations in AFP levels are admissible [5], a marked increase, as seen in our case, is still a rarity. Historically, the pathogenesis was considered to be a hamartomata's process; however, recent advances in the last two decades have suggested a neoplastic etiopathogenesis [6] involving C19MC Cluster. Atas E, et al, reported a cystic mesenchymal hamartoma masquerading as hydatid cyst in an infant [7]. It is, however, noted that multiseptation is not as common in a hydatid cyst as it is in mesenchymal hamartoma. We believe that in endemic regions, like Pakistan, this mimicry poses a potential problem for the clinicians. Our patient underwent a laparotomy on the suspicion of hydatid cyst. The variability in radiological findings poses another challenge. Quillin SP, et al, reported that mesenchymal hamartomas, although frequently cystic, often show a solid component [8].

This, coupled with the fact that an AFP level as high as 807.9 ng/ml was noted in our case, makes the possibility of Hepatoblastoma an essential clinical consideration. It is of note here that hepatoblastomas are currently the most common pediatric liver tumors, as per WHO. Furthermore, the AFP levels in our case dropped to 35 ng/ml postoperatively, which served as a proof that the tumor marker was being secreted from mesenchymal hamartoma itself. It is of note that a systemic workup of the patient was unremarkable, since gonadal yolk sac tumors also secrete AFP. Histopathology conclusively showed the tumor to be Mesenchymal Hamartoma. The distinction between the discussed entities is of paramount importance because of the extremely different treatment strategies employed in each of them. Where Mesenchymal Hamartomas [9] are reported to be benign, hepatoblastomas and yolk sac tumors require chemotherapy and aggressive follow ups. We believe that this case report generates an important message, that is, an elevated Alpha Feto Protein

Level can be seen in tumors other than hepatoblastoma and yolk sac tumors. Furthermore, to the best of our knowledge, a case of this kind has not yet been reported from Pakistan.

Conclusion

The masquerading of mesenchymal hamartomas as hepatoblastomas and hydatid cysts is a documented, albeit rare clinical presentation, with dire clinical consequences if not resolved. Histopathology remains the gold standard in distinguishing them. It is imperative to clinically consider all possibilities before deciding radical treatments.

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