Case Report

Mesenchymal hamartoma of the liver

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Abstract

A case report of a 7-year-old girl who presented with a 4-year history of painless progressive abdominal swelling that became painful 2 weeks before presentation. Examination revealed a girl in painful distress with distended abdomen and a tender palpable firm right upper quadrant abdominal mass measuring 28 cm × 22 cm with well-defined borders. Abdominal ultrasound scan showed a cystic liver mass filling the abdomen which was multilocular. Liver function tests show mildly elevated liver enzymes and mild prolongation of the prothrombin time. She had repeated aspiration of the cyst content in various peripheral hospitals with transient relief of symptoms. She thereafter had abdominal exploration through a right upper transverse incision and was found to have a cystic mass involving segments V, VI, and VII of the right lobe of the liver measuring 30 cm × 26 cm. Marsupialization of the cyst was done and an incisional biopsy of the cyst wall was taken. Histology of the cyst wall showed a mesenchymal hamartoma of the liver (MHL). The aim of this study is to highlight the clinical features, diagnosis, treatment, and differential diagnosis of MHL

Keywords: Abdominal mass, liver cyst, mesenchymal hamartoma

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Submitted: 14-Mar-2021 **Revised:** 16-May-2021 **Accepted:** 18-May-2021 **Published:** 30-Jun-2021

INTRODUCTION

Mesenchymal harmatoma of the liver (MHL) is a rare benign liver tumour, although the second commonest benign liver tumour in children after hemangioma. It has a wide range of clinical presentation, radiological features and rarely, the ability for malignant transformation. Tumour resection in resectable tumours is curative.[1,2] However, neglected or poorly managed tumours may grow huge in close proximity to the porta hepatis and may become unresectable completely, predisposing the patient to the risk of malignant transformation of partially excised residual tumour. Early clinical suspicion, histological diagnostic confirmation and complete surgical excision is therefore key in the management of MHL.[3-6] In this report, we present a case of a lately diagnosed huge MHL that complete tumour resection was difficult due to close apposition to major portal vessels.

Access this article online	
Quick Response Code:	Website: www.njghonweb.org
	DOI: 10.4103/njgh.njgh_2_21

CASE REPORT

The patient was a 7-year-old female who presented with a 4-year history of progressive painless abdominal swelling that became painful 2 weeks before presentation. The swelling which was more at the right hypochondriac region was insidious in onset but gradually increased in size over 4 years to about 12 times its initial size. There was no history of the leg or facial swelling or swelling in any other part of the body. The pain was located at the right upper quadrant (RUQ), does not radiate to other parts of the body, was dull in nature and had no known aggravating

How to cite this article: Mashor MI, Shehu H, Abobarin O, Shehu M, Mosugu O. Mesenchymal hamartoma of the liver. Niger J Gastroenterol Hepatol 2021;13:23-6.

factors but it was relieved occasionally by analgesics such as paracetamol, diclofenac tablets, and sometimes injections. There was no history of vomiting, constipation, jaundice, or bleeding from any orifices. There was no history of cough or breathlessness. There was no associated history of fever. There was no significant history of weight loss. She had repeated aspiration of the cyst content in various peripheral hospitals with transient relief of symptoms and reduction in the size of swelling but rebound distension results with associated pain. Physical examination revealed a young girl in painful distress. She was afebrile with a temperature of 36.9°C, moderately pale, anicteric, and acyanosed. There was no significant peripheral lymph node enlargement or pedal edema. There were no peripheral stigmata of chronic liver disease. The abdomen was distended with marked fullness at the right hypochondrial region and mild tenderness at the RUQ. There was a palpable firm RUQ abdominal mass measuring 28 cm × 22 cm with well-defined borders, with cystic and solid components. The mass could not be demarcated from the liver. The liver span was 22 cm and there were no demonstrable ascites. An X-ray of the chest showed an elevated right dome of the diaphragm. Abdominal ultrasound and computerized tomography scans showed a huge cystic liver mass filling the abdomen. The mass occupied most of the right lobe of the liver with very little calcification, the left lobe was thinned out by the compression of the mass on the right lobe. The enlarged liver was seen to significantly compress the right kidney, right adrenal, and adjacent bowel. There were no intraabdominal lymph nodes enlargement and no cyst in the spleen and pancreas. Liver function tests showed mildly elevated liver enzymes alkaline phosphatase of 249 u/L (36–126 μ/L), aspartate transaminase of 52 μ/L (8–39 μ/L), alanine transferase of 43 μ/L (3–42 u/L). Electrolytes, urea, and creatinine showed sodium of 125 (135–145 mmol/L), chloride of 91 mmol/L (96–106 mmol/L), urea of 1.5 (2.5–6.1 μm/L) and creatinine of 25 μm/L (60–110 μm/L). Hepatitis B surface antigen, hepatitis C virus, retroviral screening, and venereal disease laboratory test results were nonreactive. Total white cell count was 8.0 × 10^9/L and the differential white blood cell count showed neutrophils 70%, lymphocytes 20%, monocytes 8%, eosinophils 2%, and basophils 0%. The packed cell volume was 36% and the random blood glucose was 5.3 mmol/L. There was a mild prolongation of the prothrombin time. She had abdominal exploration through a right upper transverse incision and intraoperative findings revealed a huge cystic mass [Figure 1a and b] involving segments V, VI, and VII of the right lobe of the liver measuring 30 cm × 26 cm. The left lobe of the liver appeared normal. Right partial hepatic resection was done with deroofing and marsupialization of the residual cyst due to its proximity to the porta hepatis. The cyst wall was taken for histopathological examination. The gross examination showed fragments of greyish white cyst wall tissue measuring 3 cm × 1 cm × 0.5 cm and weighing 2 g. The histology of the cyst wall showed a cyst wall devoid of an epithelial lining, composed of disorderly arranged nests and cords of hepatocytes, bile ducts, blood vessels, and varying degrees of maturing fibro-collagenous tissue [Figure 1c and d] in keeping with a mesenchymal hamartoma of the liver (MHL). The patient was discharged and has been on 5 years followed up plan at the Pediatric Outpatient Clinic with no symptoms, and the abdominal ultrasound done has been normal so far. Her 5-year follow-up plan is as follows; 3-monthly for the first 1 year, twice yearly for the next 2 years and then once yearly for the 4th and 5th years.

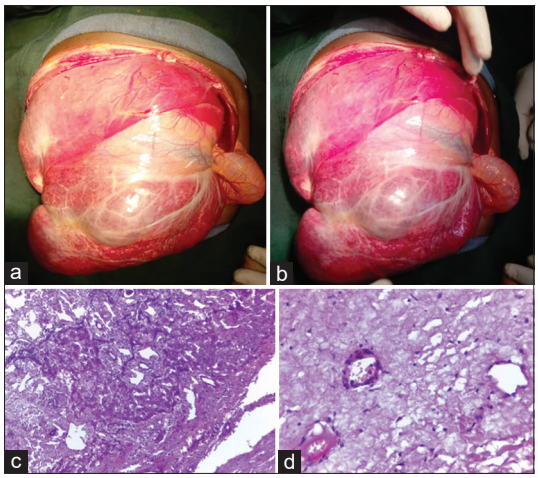

Figure 1: (a and b) Intraoperative gross morphology of hepatic cystic mass. (c) H and E stained cyst wall composed of disorderly arranged nests of hepatocytes, bile ducts, and blood vessels. (d) H and E stained cyst wall composed of myxoid immature fibro-collagenous stroma and multiple vascular channels some of which are dilated

DISCUSSION

Although MHL is generally rare, it is the second most common pediatric benign liver tumor after hepatic hemangioma, accounting for 18%–29% of these tumors.[1,2] This tumor was first named MHL by Edmondson in 1956, although it was first described by Maresh in 1903.[3] In this report, the patient first developed symptoms at the age of 3 years old. This was similar to the age of presentation of 1–4 years reported by Gupta *et al*., Rosado *et al*., and Koganti *et al*.[4-6] However, a wider age range of presentation of 0–19 years was reported in a case series by Stocker and Ishak.[7] Although this case was that of a female, there is generally a male preponderance with a male to female ratio of 1.5–2:1.[7,8]

This index case presented with the classical clinical features of progressive abdominal swelling due to a palpable RUQ abdominal mass abdominal with associated pain and tenderness similar to most other reports.[4-6] However, other symptoms such as loss of appetite, nausea, postprandial vomiting, and poor weight gain as reported by Stocker and Ishak were not present in this patient.[7]

The liver function test in this patient showed some derangement in liver function as evidenced by the elevated liver enzymes and prothrombin time. This is contrary to findings by Rosado *et al*. and Koganti *et al*. who reported normal liver enzymes and function in their patients.[5,6] They also found that cases with deranged liver enzymes had huge tumor sizes >12 cm in widest diameters, suggesting that the huge tumor compressive effect on the hepatocytes and hepatobiliary tracts may be responsible for the deranged liver function in this patient.[5,6] The ultrasound and intraoperative finding of a cystic mass in this report is the commonest imaging and intraoperative finding. These cysts are more commonly multilocular as in this present case or unilocular in some other cases. These cysts are thought to occur as a result of cystic dilatation and degeneration of the bile ducts and lymphovascular channels. However, few cases are entirely solid.[7]

MHL is grossly a well-circumscribed but unencapsulated mass with the cut sections showing cystic and myxoid appearance as seen intraoperatively in this case. The histology classically showed a cyst wall devoid of an epithelial lining, composed of disorderly arranged nests and cords of hepatocytes, bile ducts, blood vessels, and varying degrees of maturing fibro-collagenous tissue with areas of myxoid degeneration. This disorderliness or loss of the normal lobular liver architecture is thought to result from ductal plate malformation during development. Hence, there is the isolated development of mesenchymal rests outside the normal portal triad architecture leading to independent differentiation.[9,10]

Intraoperatively, this tumor was seen in the right lobe of the liver similar to the finding in the review by Stringer and Alizai who reported that 75% of cases involved the right lobe of the liver with about 20% involving both lobes and it is limited to the left lobe in only 5% of cases.[11] Rarely, MHL may be pedunculated. The treatment option is mainly surgery with resection and clear margins, enucleation, marsupialization, and sometimes watchful waiting in asymptomatic cases. However, enucleation, marsupialization, and watchful waiting were discouraged by some authors except for marsupialization in unresectable cases, due to the risk of transformation of the remnant hamartoma to undifferentiated embryonal sarcoma.[12,13] In this index case, complete resection was impossible due to the proximity of the residual cyst to the portal vein

Differential diagnoses included other cystic lesions which ranged from inflammatory to neoplastic conditions.[14] The inflammatory conditions include hydatid cysts and amoebic liver abscesses which are characterized by typical parasitic morphologies and mixed inflammatory cell infiltrates composed of eosinophils, lymphocytes, plasma cells, macrophages, and some neutrophil polymorphs. Neoplastic conditions include benign lesions such as bile duct adenoma characterized by absence of islands of hepatocytes, bile duct hamartoma characterized by multiple bile ducts within a fibrous stroma, infantile hemangioma, and hemangioendothelioma characterized by multiple variable caliber vascular channels.[14] The malignant mimickers of MHL include embryonal sarcoma and hepatoblastoma characterized by markedly pleomorphic cells with increased mitotic activity and mixed mesenchymal and epithelial cells characterized by embryonal and fetal hepatocytes which may be admixed with mesenchymal components such as bone, and cartilage.[14]

CONCLUSION

MHL is a rare benign liver tumor in children with variable clinical, laboratory, imaging, and histological features. There are many mimickers of MHL ranging from inflammatory cystic lesions to benign and malignant neoplasms. It is essential to make the early histological diagnosis or rule out MHL in clinically suspected children with hepatic masses to allow for early treatment and prevent its transformation to malignancy.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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