Case Report

Mesenchymal Hamartoma of the Liver Incidentally Discovered in a 4 Year Old Girl: Management and Treatment

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Abstract

Mesenchymal hamartomas of the liver is considered a benign tumour, although it provides significant challenges in the diagnosis and operative management. Clinical features, laboratory results and radiographic imaging are often non-specific and inconclusive. Thus, it is still difficult to differentiate from malignant lesion without pathological examination. While this tumour is usually diagnosed in the first 2 years of life, few cases in older children have been reported and most of them are case reports. In this article, we want to report our clinical evaluation, results of image diagnosis, surgical procedure and the follow-up simultaneously we discuss the possible differential diagnosis of such a case at this age.

Keywords: Hepatic mesenchymal hamartoma, paediatric liver tumours, paediatric surgery

Introduction

Mesenchymal hamartoma of the liver (MHL) is the second most common benign hepatic tumour in childhood and represents about 5% of all the primary hepatic tumours.[1] It was first reported by Edmanson in 1956. This tumour is usually diagnosed in the first 2 years of life (80% younger than 2 years), although few cases in older children have been reported and most of them are case reports.[2] The exact pathogenesis of MHL is unclear, but some recent studies on this tumour have found neoplastic features such as genetic anomalies and malignant transformation.[3] These findings support that the conventional approach of completely resecting the tumour whenever possible remains the best treatment. In this article, we report on our experience treating this patient.

Case Report

We report a 4-year-old girl who was initially followed up at the paediatric department for idiopathic thrombocytopenic purpura. She was referred to our institution for further evaluation of an abdominal mass discovered incidentally on ultrasonography (USG) performed to assess abdominal pain. There was no history of abdominal discomfort or nausea and recurrent abdominal pain.

Upon physical examination, the patient was found to be slightly pale without jaundice, fever or weight loss. Abdominal examination found a huge, non-tender and firm mass with a smooth surface occupying the right lumbar reaching the right iliac fossa.

On admission, laboratory evaluation showed normal liver function tests with a prothrombin time (PT) of 100% tricarboxylic acid: 39"/32"; alanine aminotransferase 13 U/L; aspartate aminotransferase 23 U/L and gamma-glutamyl transferase 24 U/L with normal alkaline phosphatase and bilirubin levels. Complete blood count shows: Haemoglobin: 9 g/dl, haematocrit: 27, 5%, white blood cell: 10,950 and platelet count: 446,000. Her alpha-fetoprotein (αFP) was normal (1.45 IU/mL).

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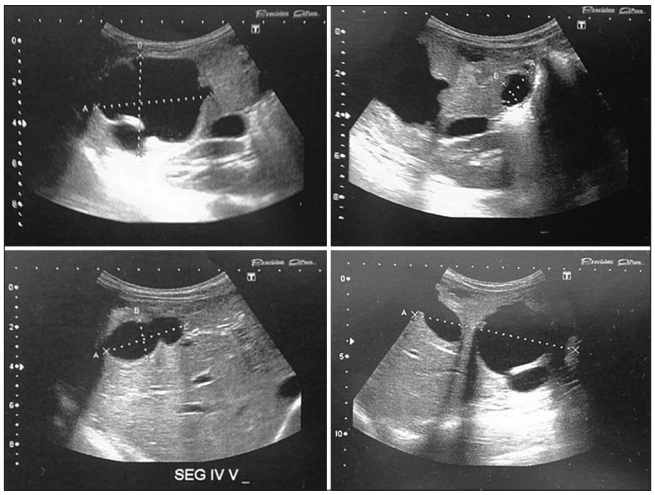


Figure 1: Ultrasonography shows a huge, mixed solid and cystic tumour. The solid portion of the tumour is slightly hyperechoic; No calcification was seen

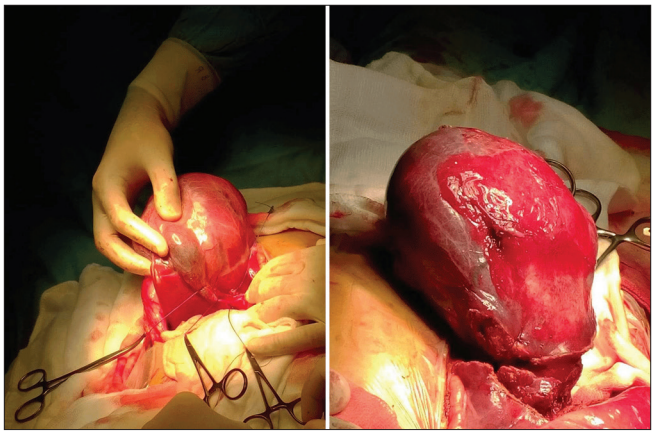


Figure 3: Per-operative photo of the mesenchymal hamartoma of the liver showing the gallbladder adherent to the tumour

Ultrasound revealed a well-defined solid cystic mass involving the right hepatic lobe. The cysts were anechoic, avascular and variable in size (largest 58 mm × 50 mm) with posterior enhancement. Thin septae and peripheral solid components were noted. The solid areas were slightly hyoerechoic to the normal liver parenchyma [Figure 1]. An X-ray of the chest showed the elevated dome of the right diaphragm. A magnetic resonance imaging (MRI) examination of the abdomen was performed which revealed the presence of an exophytic large mass measuring 100 mm × 60.8 mm × 111 mm occupying segments IV, V and partially segment VI with clusters of multiple T2-bright circumscribed cystic lesion with displacement of the bowel loops and the gallbladder [Figure 2]. There was no calcification. Hydatid serology was negative.

However, since malignancy of the lesion could not be ruled out by pre-operative imaging diagnosis and to clarify the diagnosis, a liver biopsy was performed and the pathology department diagnosed mesenchymal hamartoma.

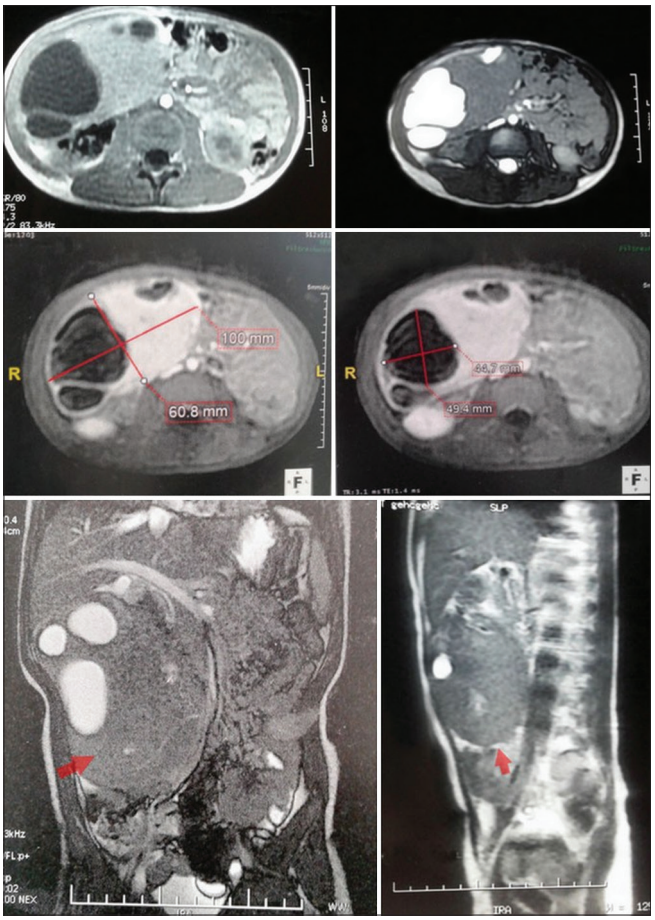


Figure 2: Axiale coronale and sagittale magnetic resonance image showing the cystic solid mass with displacement of adjacent structures and Clusters of multiple T2-bright circumscribed cystic lesions suggestive of mesenchymal hamartoma

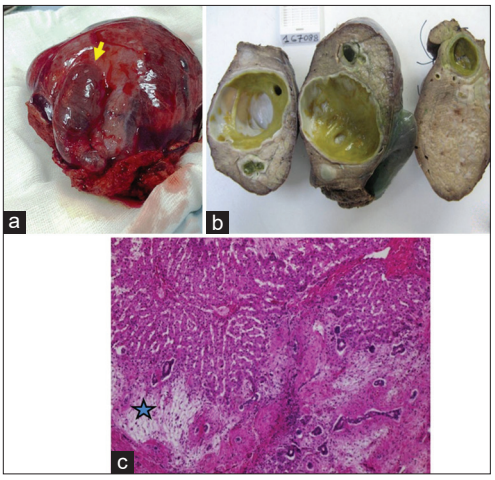


Figure 4: Pathological findings of hepatic mesenchymal hamartoma: (a) Tumorectomy specimen, Note the gallbladder adherent to the tumour (yellow arrow). (b) Gross finding of MH. Cut section of the specimen showing areas of multiple cysts and solid portion in the tumour. (c) Mesenchymal hamartoma of the liver (HE × 200): Mixture of mature hepatocytes and bile ducts Surrounded by an immature mesenchyme (blue star); Hepatocytes were found in cords, islands or lobules

The patient was planned for surgical intervention and the abdomen was explored through a right reversed L-shaped incision. There was a tumour measuring approximately 15 cm arising from the right lobe, to which the gallbladder was adherent [Figure 3]. The mass was excised completely with a healthy wedge and was measuring 500 gr in weight. The operative time was 145 ms. The post-operative course was uneventful and the child was discharged in a good general condition at the 7th day after surgery.

On examination, the cut section of the specimen showed areas of multiple cysts. Histopathopathological examination of the specimen confirmed the diagnosis of cystic mesenchymal hamartoma [Figure 4].

A 5-year follow-up ultrasound revealed a normal liver with no residual or recurrent tumour.

Discussion

Mesenchymal hamartoma of the liver (MHL) is the second most common benign liver tumour in children after infantile hemangioendothelioma; yet, its pathogenesis remains poorly understood. This lesion was described by various names including: bile cell fibroadenoma, bile duct fibroadenoma, lymphangioma, cavernous lymphangiomatoid tumour, pseudocystic mesenchymal tumour, benign mesenchymoma and cystic hamartoma.[4]

Clinically, patients with mesenchymal hamartoma of the liver present in the first 2 years of life with a median age of 10 months. This lesion is two time more common in male gender, the right lobe being affected more frequently than the left. The characteristic of our case was that this patient was female and more than 2 years old.

Mesenchymal hamartoma of the liver (MHL) may be found incidentally by the parent while bathing the child or palpated unexpectedly on routine physical examination or discovered on abdominal imaging especially with the widespread use of USG, but usually, it presents with abdominal distension and/or an upper abdominal mass. Occasionally, the mass can expand rapidly, most likely because of rapid accumulation of fluid within cystic spaces and complications including ascites, jaundice and even congestive heart failure can occur. Findings during physical examination show a non-tender right upper quadrant mass with firm smooth borders. Abdominal pain, anorexia, vomiting and poor weight gain have also been reported.

Routine laboratory test results are usually normal. Serum αFP levels are typically normal; however, elevated αFP serum levels have been reported in some cases confounding thus the diagnosis.[5]

Plain radiography may shows calcification within the hepatic tumour, but this is uncommon.

In most patients, the diagnosis of MHL is suggested by imaging and confirmed by histological examination of the resected specimen. If there is doubt about the diagnosis, a percutaneous or open tumour biopsy can be performed. The role for biopsy prior to resection of these lesions has a place in selected lesions. This would include patients who have a predominantly solid lesion or those patients with inappropriately elevated age specific αFP.[6]

USG of the abdomen, computerised tomography (CT) scan and MRI have been used to confirm the diagnosis. At unenhanced CT, mesenchymal hamartoma usually has a heterogeneous appearance. The stromal elements appear hypoattenuating, whereas the cystic component has water attenuation. The appearance of the mass depends on whether it is predominantly stromal (mesenchymal) or cystic. MRI features of MHL are still emerging. MRI reflects the underlying pathology, exquisitely demonstrates the cystic and solid components and identifies the sharp outlines of the lesion separate from rest of the tissues. The MRI appearance of mesenchymal hamartoma varies depending on the presence of stromal elements and the protein content of the fluid.[7] Haemorrhage occasionally occurs but is not typical. After intravenous administration of contrast material, the stromal component enhances.

A cystic MH may need to be distinguished from several other cystic and solid-cystic lesions of the liver such as congenital liver cysts, hydatid cyst, liver abscesses, haemangiomas and haemangioendotheliomas, teratomas, hepatoblastomas and undifferentiated embryonal sarcoma of the liver.[7]

Conditions reported in association with mesenchymal hamartoma include adrenal cytomegaly, small bowel malrotation, neonatal hyperbilirubinemia, fetal hydrops, Beckwith Wiedemann endocardial syndrome, fibroelastosis and diffuse endocrinopathy.[8] Haematologic disorders associated to this tumour such as idiopathic thrombocytopenic purpura and disseminated intravascular coagulation, also known as Kasabach-Merritt phenome, have also been reported in older children.[9]

Mesenchymal hamartoma of the liver has traditionally been regarded as a benign tumour with no malignant potential, but several reports have highlighted an association between MHL and undifferentiated embryonal sarcoma.

The surgical approach to mesenchymal hamartomas is controversial. Non-operative management has been proposed by some authors in selected cases (e.g., infants with a biopsy-proven MHL) since spontaneous regression of these lesions has been reported.[10] Rarely enucleation may be an option if the tumour can easily be dissected from the surrounding tissue; This approach minimises the loss of normal liver tissue and decreases blood loss. In case of large lesions, percutaneous aspiration or drainage of larger cysts may temporarily control tumour size, and marsupialisation of the cysts into the peritoneal cavity may be employed.[11] However, because of the risk of recurrence and malignant transformation into embryonal sarcoma, complete surgical excision remains the mainstay of treatment and the most widely used techniques include hepatectomy, segmentectomy, and expanded resection.[3,11]

Other alternative therapy such as hepatic artery embolisation, chemotherapy and radiotherapy have been proposed but did not avert the need for surgery. Our preference has been total excision of the huge tumour with a surrounding rim of normal liver tissue.

After complete resection, occasional clinical and US review is advisable for at least 5 years, but an excellent prognosis can be anticipated.

This is an interesting case since few cases of mesenchymal hamartoma of the liver are diagnosed at this age. We want to show that this lesion can still clinically silent for a long period.

Conclusion

Mesenchymal hamartomas while considered uncommon and benign provide significant challenges in the diagnosis and operative management. Clinical features, laboratory results and radiographic imaging are often non-specific and inconclusive. Biopsy is rarely recommended to establish the diagnosis. Complete surgical excision whenever possible remains the treatment of choice. Early diagnosis and resection is needed otherwise severe complications can ensue. In most cases, prognosis is excellent. Mortality is related to the surgical complications. All patients should be followed up, even in cases of complete surgical resection, for the potential of local recurrence and malignant transformation.

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.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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