

CASE REPORTS

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Giant solid mesenchymal hamartoma of the liver in a neonate: case report



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Abstract

Background: Mesenchymal hamartoma is the second most common benign liver tumor in children, with 20% of the cases diagnosed during the neonatal period. The exact etiology is still unclear, and most investigators believe that it is a developmental anomaly rather than a true neoplasm. The presentation of these tumors is highly variable depending on the lesion's size, ranging from small asymptomatic lesions to very large tumors with life threatening complications. Radical surgical excision, whenever possible, is the gold standard for treating these lesions to avoid the problems of local recurrence and possible malignant transformation. We present the rare occurrence of an entirely solid, giant hepatic mesenchymal hamartoma in a 3-week-old male newborn and discuss the mode of presentation, as well as the diagnostic and therapeutic approach.

Case presentation: A 3-week-old male newborn was referred to our institution with huge abdominal distension and respiratory distress. Imaging studies confirmed the presence of a very large solid intraabdominal mass occupying the majority of the abdominal cavity and abutting the inferior aspect of the right lobe of the liver, but did not reveal the diagnosis. At laparotomy, a huge solid mass was found attached to the right lobe of the liver. Complete excision was done, and histopathological examination confirmed the diagnosis of mesenchymal hamartoma.

Conclusion: Although rare, mesenchymal hamartoma of the liver can present as a neonatal surgical emergency. Emergency intervention is required in symptomatic patients. Radical surgical intervention is possible and is the treatment of choice to relieve the patient's symptoms and avoid future complications.

Keywords: Mesenchymal hamartoma, Liver, Newborn, Case report

Background

Mesenchymal hamartoma of the liver is reported to be the second most common benign liver tumor in children, with 80% of these lesions diagnosed within the first 2 years of life [1, 2]. The exact etiology behind the development of this pathology is still unclear and is thought to be a developmental anomaly of the portal connective tissue rather than a true neoplasm [3]. The presentation of hepatic mesenchymal hamartoma is variable depending on the lesion's size. Small lesions are usually asymptomatic and often diagnosed incidentally on imaging studies. In many cases, these lesions present as giant tumors filling almost the entire abdominal cavity with a

diameter of up to 20–30 cm [4]. Large mesenchymal hamartomas usually present with abdominal distension, vomiting, poor weight gain, respiratory distress, cardiac failure, thrombocytopenia, intralesional bleeding, ascites secondary to tumor rupture, obstructive jaundice, and hepatomegaly [5]. Twenty percent of these lesions are diagnosed during the neonatal period as they attain large sizes secondary to their postnatal growth spurt from the rapid accumulation of fluid within them [5]. Several associated anomalies have been reported including malrotation, Beckwith-Wiedemann syndrome, biliary atresia, and congenital heart diseases [1, 5]. Radical surgical excision, whenever possible, is the gold standard for treating these lesions to avoid the problems of local recurrence and possible long-term malignant transformation [2].

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Case presentation

A 3-week-old male newborn was referred to our institution with huge abdominal distension and respiratory distress, with an abdominal ultrasound that revealed the presence of a huge, ill-defined heterogeneous, hyperechoic, solid-appearing soft tissue mass occupying the majority of the abdominal cavity and abutting the inferior aspect of the right lobe of the liver with difficulty to determine its origin. Complete blood count revealed almost normal blood counts and morphology apart from many target red blood cells. Serum sugar, urea, and creatinine levels were within normal limits. Liver function test revealed mild indirect hyperbilirubinemia, with normal levels of liver enzymes. Serum alpha-fetoprotein was significantly elevated (> 2000 ng/ml). Abdominal computed tomography revealed the presence of a very large ($10 \times 8.5 \times 7.5$ cm) well-defined, homogeneous, hypovascular solid mass filling almost the entire central abdominal cavity with anterior displacement of the bowel loops, with the suggestion of teratoma or connective tissue tumor. There were no signs of major vascular encasements or significant effect upon intraabdominal structures, no abdominal or pelvic lymphadenopathy, and no focal lytic or sclerotic bony lesion (Fig. 1a, b). No signs of pulmonary collapse, infiltrates, or pleural effusion were evident on CT imaging of the chest.

The presence of severe respiratory distress from diaphragmatic compression resulting from the presence of a huge intraabdominal space occupying lesion mandated surgical intervention. The patient was admitted to the neonatal intensive care unit, initially stabilized, central line access was secured, two units of packed red blood cells were prepared, and a full set-up of monitoring and possible ventilatory support was arranged. Laparotomy was done through a transverse supraumbilical incision, and a huge solid mass ($15 \times 9.5 \times 8$ cm) was found attached to the inferior aspect of the right lobe of the liver with a broad thick peduncle. The mass was completely resected intact without difficulty after dividing the peduncle and separating the remainder part from the liver capsule using diathermy. A few millimeters of normal liver capsule where the remainder part of the tumor was attached were included with the resection as a safety margin (Fig. 2a, b, c).

Histopathological examination revealed a well-vascularized mature connective tissue within myxoid stroma intermixed with few branching bile ducts consistent with the diagnosis of mesenchymal hamartoma of the liver.

The patient experienced uneventful postoperative recovery and discharged well from the hospital on the 6th postoperative day with no signs of respiratory distress. One week later, the patient had been checked, with excellent wound healing and quiet respiration. Thereafter, the patient attended a regular follow-up at 3 months interval for 1 year with serial abdominal ultrasound examinations that

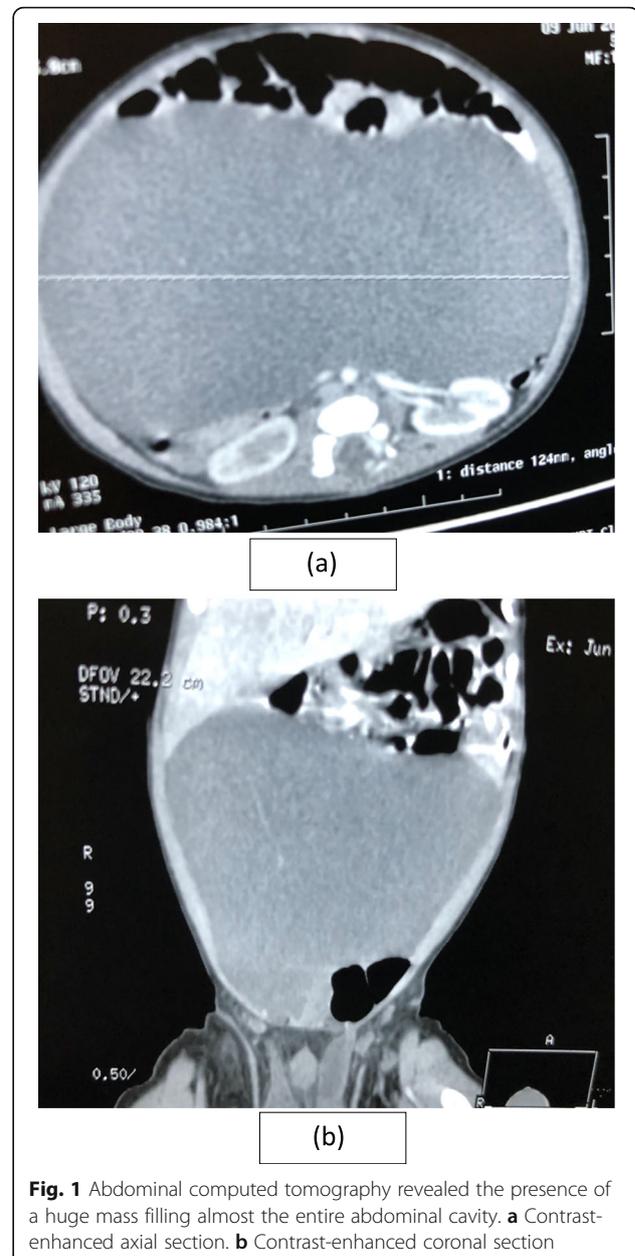


Fig. 1 Abdominal computed tomography revealed the presence of a huge mass filling almost the entire abdominal cavity. **a** Contrast-enhanced axial section. **b** Contrast-enhanced coronal section

revealed no residual mass. No significant respiratory or other postoperative complications were recorded. Serum alpha-fetoprotein level has significantly reduced to < 100 ng/ml in 3 months postoperatively.

Discussion

Mesenchymal hamartoma of the liver is a rare benign tumor that is believed by most authors to be a developmental anomaly rather than a true neoplasm. Histologically, the tumor is made up of loose or mucoid connective tissue containing different amounts of blood vessels, cystic lymphatic spaces, bile ducts, and normal hepatocytes [4]. Developmental theories include exposure



(a)



(b)



(c)

Fig. 2 Intraoperative findings. **a** Mass attached to the liver with a thick broad peduncle. **b** Dissecting the mass off the liver proceeded. **c** Mass is completely excised

of otherwise normal hepatic parenchyma to regional ischemia or toxic injury during its development [6]. The term “mesenchymal hamartoma” was first used by Edmondson in 1956 who distinguished the pathology from other cystic and tumor-like lesions of the liver [2, 7].

Hepatic mesenchymal hamartomas usually present as asymptomatic lesions; however, they can grow to attain large sizes with the potential to compress adjacent organs resulting in a wide spectrum of symptoms and complications ranging from respiratory distress to death [8].

In this study, the patient was a newborn who was presented with severe respiratory distress due to diaphragmatic compression that results from the presence of a giant tumor filling almost the entire abdominal cavity.

Mesenchymal hamartoma of the liver presents most frequently as a multiseptated cystic or mixed solid and cystic tumor; however, it can rarely occur as a solid tumor [3]. Twenty percent of these tumors can be diagnosed during the neonatal period [5]. In this study, the patient was a 3-week-old newborn with a giant solid hamartoma. Some authors believe that the solid tumor is a “younger” form that will be transformed into a cystic form at a later date. This is confirmed by the more frequent detection of solid forms at younger ages. However, other researchers do not support this finding [4].

Laboratory tests in children with hepatic mesenchymal hamartomas are usually within normal limits. Increased levels of serum alpha-fetoprotein along with young patient’s age may lead to the misdiagnosis of hepatoblastoma [4]. Chang et al. reported increased levels of serum alpha-fetoprotein in patients with solid lesions [9]. In this study, the patient was a newborn with solid form of the tumor. His serum alpha-fetoprotein level was significantly increased.

The majority (75%) of mesenchymal hamartomas develop in the right lobe of the liver, with the remainder found in the left lobe or involving both lobes. Up to 20% of these lesions are pedunculated, arising from the inferior surface of the liver [6]. In this study, the tumor was attached to the inferior surface of the right lobe of the liver with a broad thick peduncle making its complete excision easy after dividing that peduncle.

Various therapeutic strategies have been reported for the management of hepatic mesenchymal hamartomas, including enucleation for small lesions and marsupialization into the peritoneal cavity for larger lesions [10]. However, complete surgical excision with a margin of normal tissue liver is the gold standard of treatment of hepatic mesenchymal hamartomas to avoid the problems of local recurrence and long-term malignant transformation [2]. Liver transplantation is considered controversial for the treatment of this pathology; however, it is a viable therapeutic option in giant unresectable bilobed lesions [2]. In this study, radical surgical excision of the tumor was achieved without complications. The patient attended regular follow-up for 1 year

with serial abdominal ultrasonography. No significant post-operative complications or findings were recorded.

Conclusion

Although rare, mesenchymal hamartoma of the liver can present as a neonatal surgical emergency and must be considered as a possible diagnosis in newborns with huge abdominal masses related to the liver. Emergency intervention is necessary in symptomatic patients. Radical surgical excision is possible and is the treatment of choice to relieve pressure symptoms, confirm the diagnosis with histopathology, and avoid future complications.

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Availability of data and materials

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Ethics approval and consent to participate

Written informed consent to participate was obtained from the parent.

Consent for publication

Written informed consent for publication of this case report and accompanying images was obtained from the parent.

Competing interests

The author declares that he has no competing interests

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