Neonatal Mesenchymal Hamartoma of Liver: An Unusual Presentation

Raghu Sampally Ramareddy, Anand Alladi
Department of Pediatric Surgery, Vani Vilas Hospital, Bangalore Medical College and Research Institute, Bangalore, Karnataka, India

ABSTRACT

Mesenchymal hamartoma of the liver is the second most common benign liver tumor in children. Typically, it presents as a large benign cystic, solid or mixed liver mass in a child younger than 3 years and amenable to complete resection. We report a neonate with Mesenchymal hamartoma of the liver presenting as giant intra abdominal cyst and its rare association with malrotation of bowel.

Key words:
Liver, mesenchymal hamartoma, neonate

INTRODUCTION

Clinical presentation of neonatal Mesenchymal hamartoma of the liver (MHL) varies from incidental detection on imaging, asymptomatic mass to symptoms like abdominal distention, pain, respiratory distress, cardiac failure, anorexia, vomiting, poor weight gain, vascular steal and thrombocytopenia, intralesional bleeding, pulmonary hypertension, tumor rupture ascitis, pedal edema, obstructive jaundice, and smooth, large nontender hepatomegaly.[1,2] Prenatal ultrasound usually detects MHL in last trimester with rise in maternal serum alpha fetoprotein (AFP) or human chorionic gonadotropin (HCG) levels and polyhydroaminas.[1,2] Associated anomalies include malrotation, Beckwith-Weidman syndrome, biliary atresia and congenital heart diseases.[1]

We are discussing the rare giant peduncle unilocular cystic variant of MHL presentation, pathological characteristics, diagnosis, and management dilemmas with review of the relevant literature.

CASE REPORT

A 2-week-old boy, one of twins, presented with huge progressive abdomen distention, non bilious vomiting, and respiratory distress since birth. Antenatal ultrasound scan at the 5th month showed diamniotic dichorionic twin fetus. Abdominal examination revealed nontender, cystic mass occupying almost the whole of abdomen. Blood chemistry of serum electrolytes, coagulation screen and liver function tests were normal. Plain X-ray abdomen showed soft tissue homogenous opacity occupying the entire abdomen with displacement of bowel to left upper quadrant [Figure 1]. Ultrasound of abdomen revealed uniloculated, huge midline anechoic lesion extending on either side with clear fluid and of uncertain origin [Figure 2]. Neonate developed severe respiratory distress due to progressive abdominal distention. Hence he was posted for emergency laparatomy and 2000 ml of clear fluid was aspirated from the cyst. The cyst was found to be uniloculated peduncle swelling arising from the inferior surface of right lobe of liver [Figure 3] and did not have any biliary communication. It was excised *in toto*. The cyst had displaced the malrotated bowel. Histopathology confirmed uniloculated cystic structure with wall containing proliferating bile ductules, lymphocytic infiltration, compressed hepatic cords, nodules, and no distinct lining suggestive of MHL [Figure 4]. Postoperative recovery was uneventful and at 1 year follow-up the child is thriving well.

DISCUSSION

Perinatal hepatic tumor comprises 5% of all neoplasia occurring in fetal and neonatal period.[2] Hepatic hemangioma, MHL, and hepatoblastoma are the common primary tumors in order of frequency in the neonatal period.[2] MHL commonly involves right lobe (75%),

Address for correspondence:
Dr. Raghu Sampally Ramareddy,
Department of Pediatric Surgery, Vani Vilas Hospital,
Bangalore Medical College and Research Institute, Bangalore,
Karnataka, India. E-mail: drsrraghu@gmail.com

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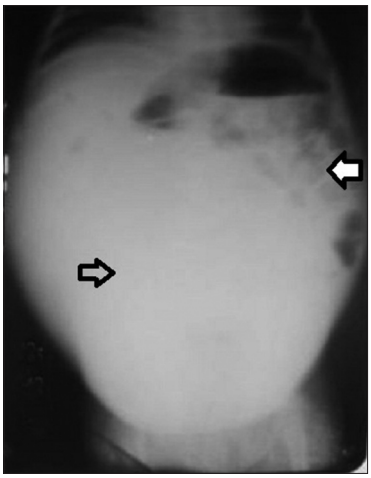

Figure 1: Plain X-ray abdomen: Soft tissues mass (hallow arrow) with bowel displacement (solid arrow)

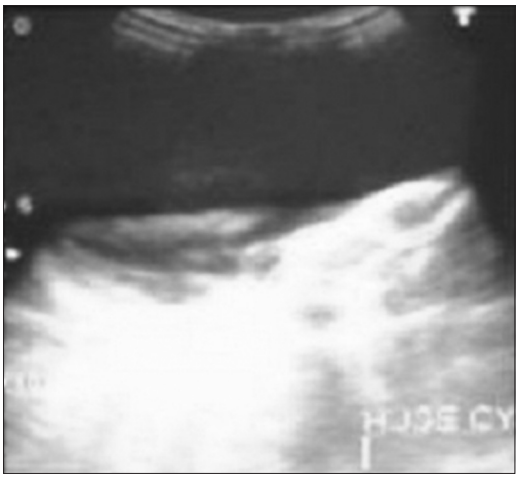

Figure 2: Ultrasound: Giant unilocular intra abdominal cyst of uncertain origin

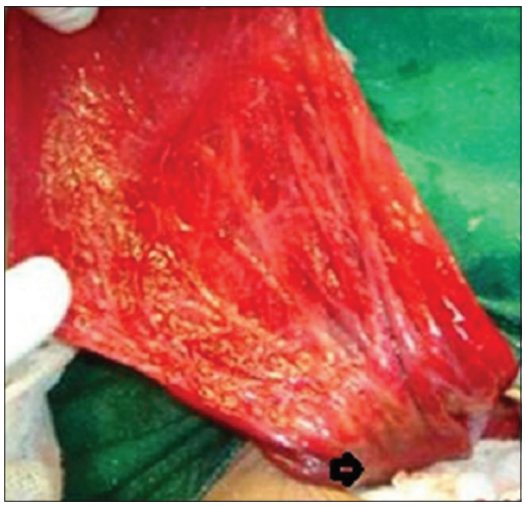

Figure 3: Operative picture: Giant pedunculated unilocular hepatic cyst arising from inferior surface of right lobe of liver (arrow)

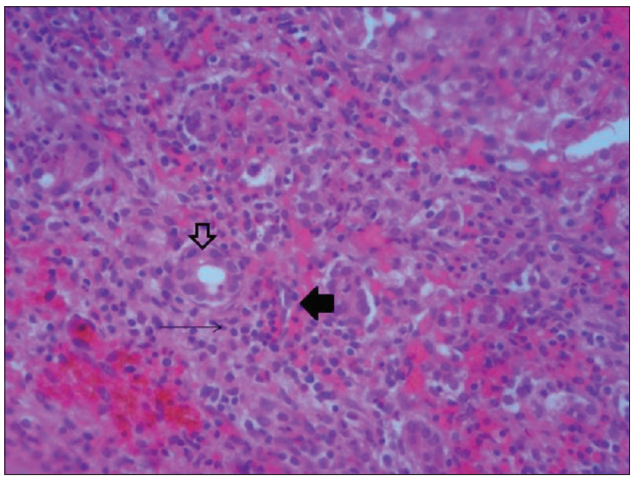

Figure 4: Microscopic picture: H and E Stain shows bile ductules (arrow down), lymphocytic infiltration (line arrow) and compressed hepatic cord nodules (left arrow) suggestive of MHL

usually multiloculated, and often pedunculated[1] (20%). Twenty percent occur in neonates and may have postnatal growth spurt due to rapid accumulation of fluid.[1,2] The rapid fluid collection in the cyst increased pressure on lungs, causing respiratory distress. Tumors may be very large, sometimes reaching 20-30 cm in diameter and poses surgical challenge for complete excision.[1] Spontaneous incomplete regression and malignant transformation are known to occur.[1] Malrotation was probably due to large cyst preventing rotation.[1]

MHL has diverse etiology; it arises from ductal plate malformation, vascular insult,[3] toxic injury, and neoplasia.[1,2] Calcification, significant necrosis and bleeding into lesion are rare.[1] Hepatocytes and bile duct epithelium within the loose myxoid stroma of the tumor sometimes produces AFB and then needs biopsy, and doppler ultrasound to rule out hepatoblastoma.[1,4] Cyst fluid is clear or pale yellow serous fluid or mucoid material and contains protein, cholesterol, and glucose at lower level than plasma. MHL are typically multiloculated, well circumscribed without capsule, do not communicate with biliary tree and are surrounded by an irregular margin of compressed hepatic parenchyma, bile ducts, and blood vessels.[1] Microscopically, they have mesenchymal, epithelial components, and myxoid stroma.[1]

Plain X-ray abdomen may show calcification, and soft tissues mass enlargement with displacement of bowel.[1] Imaging characteristics of MHL are variable.[1] Ultrasound, computed tomography (CT), and magnetic resonance imaging of MHL demonstrate a multiloculated cystic tumor with a variable amount of solid tissue.[1] With ultrasound scan, the presence of thin mobile septae and/or round hyperechoic parietal nodules within the cyst is highly suggestive of MHL.[1] A radiologist will offer differential diagnosis of intra-abdominal cyst depending on epicenter, attachment, and tissue character of mass along with clinical parameter.[3]

The differential diagnosis of abdominal cystic lesion in newborn includes mesenteric cyst, duplication cyst, hepatoblastoma, teratoma, lymphangioma, choledochal cyst, solitary nonparasitic cyst, and renal cyst.[1,4-7] Mesenteric and duplication cyst are mobile but duplication have characteristic inner wall.[5] Predominantly exophytic renal cysts are rare. Choledochal cysts are diagnosed by site of lesion in relation to liver, proximal bile duct dilatation, and jaundice features. Lymphatic cysts are multilocular. Congenital solitary nonparasitic hepatic cysts are congenital or acquired, and arise from the ductal plate. They are rare in newborn,[5] have female preponderance and cuboidal lining epithelium.[5,6] Pre-operative diagnosis of the giant peduncle uniloculated sub-hepatic cystic variant of MHL could be difficult.[1,3-9]

Infection and recurrence are common following per-cutaneous drainage of giant cyst.[1] Complete excision of MHL by laparoscopy/open is the treatment of choice in the symptomatic neonate.[1,2] Fetal hydrops, respiratory distress, abdomen compartment syndrome, and circulatory problems owing to a large space occupying abdominal lesion and sometimes stillbirth are causes of neonatal mortality.[2]

CONCLUSION

Neonatal unilocular pedunculated cystic variant of MHL is one of the rare differential diagnoses of the giant sub-hepatic cystic. Although it is benign, MHL can present as acute surgical crisis in neonatal period due to the rapid postnatal fluid collection in the cyst. Complete surgical excision is the treatment of choice and histopathology helps in the confirmation of diagnosis.

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