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CASE REPORT | PEDIATRICS

Solitary Septated Simple Liver Cyst in a Newborn Infant

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Abstract

Simple liver cysts (SLC) are generally rare and are typically symptomatic when detected in infancy. We present a case of a newborn infant in whom fetal ultrasound and MRI revealed a cystic structure. Postnatal imaging revealed a septated, single cystic structure causing mass effect on the common bile duct and partially obstructing the inferior vena cava. Treatment of a solitary septated SLC was successful by laparoscopic total excision. The infant had an uncomplicated postsurgical course and has done well.

Introduction

Simple liver cyst (SLC) is a rare nonparasitic cyst of congenital origin thought to derive from aberrant bile ducts, microhamartomas, or peribiliary glands isolated from bile ducts.[1,2] They are categorized as solitary or multiple; the latter is associated with polycystic kidney disease and Caroli's disease.[3-5] There are fewer than 10 reported solitary cystic cases described in fetuses.[6] Most are seen in the fourth and fifth decades of life.[5] SLC has a prevalence of 0.1–2.5% and a male to female ratio of 1:1.5.[7-9]

Case Report

A 26-year-old obese woman with hypertension and anemia presented for a routine prenatal ultrasound at week 17 of gestation. Fetal ultrasound revealed a pelvic cystic mass measuring 3.4 x 2.9 x 3.7 cm. Fetal MRI at 22 weeks gestation showed a large cystic structure within most of the abdominal cavity measuring 7.8 x 5.3 x 6 cm, displacing the right kidney posteriorly with compression of the bladder and lungs, possibly originating from the left kidney (Figure 1). Abdominal circumference measured >97%.

The pregnancy was uncomplicated. A male infant was born following an uneventful spontaneous vaginal delivery at week 38 of gestation. On physical examination, the newborn infant had an increased abdominal circumference, systolic murmur, and an undescended right testicle. The abdominal wall musculature was hypotonic, and a mass was palpable in the right abdomen. In the nursery, the infant remained comfortable on room air and tolerated feeds. On day 2, abdominal magnetic resonance imaging (MRI) revealed an 8.6 x 5 x 8-cm fluid filled mass in the right upper quadrant extending downward with thin margins. The mass had decreased T1 signal with increased T2 signal without solid enhancement after contrast administration (Figure 2). A small septation was seen anteriorly, with a surrounding thin rim of enhancement and multiple rounded T2 hypointense foci in the gallbladder without intra- or extrahepatic ductal dilations. Other findings included compression of the right kidney, gallbladder stones, and sludge secondary to bile duct compression, and inferior vena cava (IVC) flattening concerning for obstruction (Figure 3).

The differential included a mesenteric cyst, lymphatic malformation, hepatic malignancies, choledochal cyst, or liver cyst. Complete blood count with differential, a basic metabolic profile, and total bilirubin were normal.

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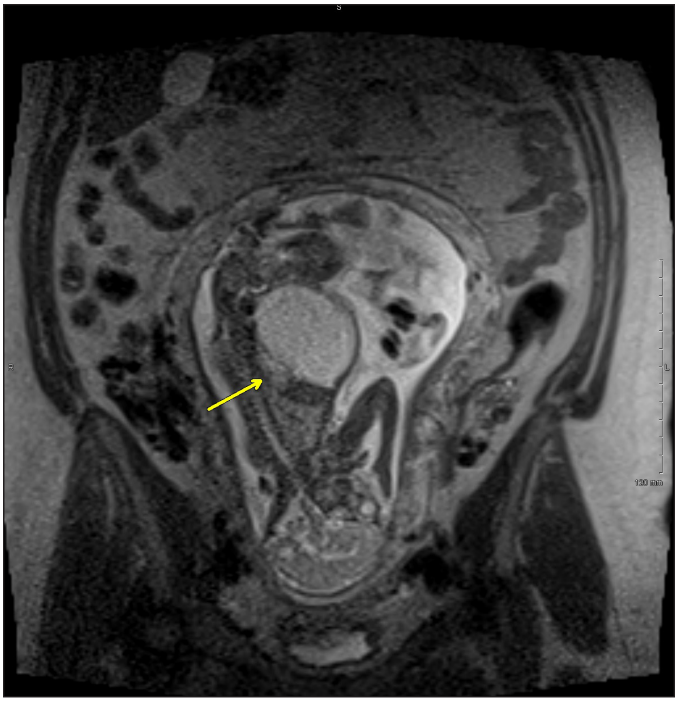

Figure 1. Prenatal MRI at week 22 of gestation showing coronal image of the mother and sagittal image of the fetus. A T2 hyperintense structure (arrow) appears within the fetal abdominal cavity inferior to the liver and anterior to the kidney.

Exploratory laparotomy with excision of a cystic liver mass attached to the inferior portion of the right hepatic lobe revealed a benign, simple epithelial cyst lined by flattened, squamoid-appearing epithelium (Figure 4). The wall was composed of fibrovascular tissue and contained small nests of benign hepatocytes. These findings supported the diagnosis of a SLC. The infant had an uneventful postoperative course and was discharged home shortly after.

Discussion

SLC are generally rare and not symptomatic, especially in the postnatal period. Most require no intervention if found prenatally.[3] However, large cysts (>3 cm) may obstruct adjacent tissues and organs, leading to atrophy and complications like spontaneous hemorrhage, bacterial infection, biliary obstruction, rupture, and neoplastic degeneration into squamous cell carcinoma.[4,6,10-15] There is a reported neonatal case of IVC obstruction from a SLC treated with surgical intervention.[16]

Successful diagnosis is challenging due to the inability to perform invasive techniques. Accurate diagnosis is made primarily by radiographic studies often requiring histopathological verification. On ultrasonography, a SLC appears as an anechoic unilocular fluid-filled space with a posterior acoustic enhancement.[17] Septations are usually absent, but when present, signify bridging of major bile ducts and vessels.[18]

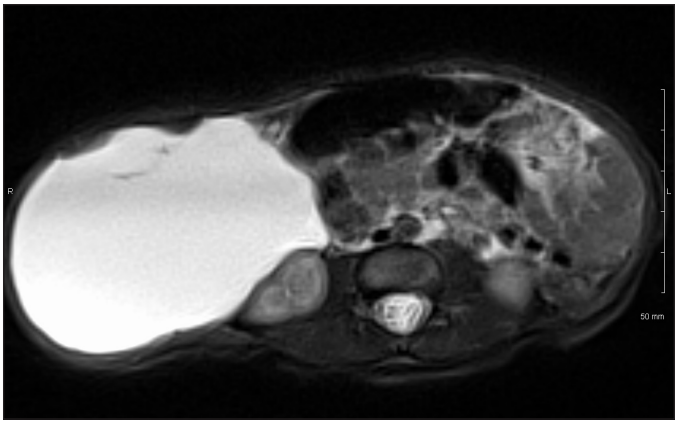

Figure 2. Postnatal MRI axial T2 image demonstrating a T2 hyperintense structure anterior to the right kidney. A small septation can be seen within the cyst anteriorly.

If congenital in origin, ultrasonography may show a large cyst, extension from the right hepatic lobe, cyst progression from midgestation into the neonatal period, and inferior liver wall flattening.[5] MRI typically reveals a well-defined water-attenuated lesion without enhancement after gadolinium. T1-weighted images show a low-intensity signal, while T2-weighted images show a high-intensity signal.[4] SLC typically cannot be differentiated from cystic malignant tumors on radiological studies; tissue diagnosis is necessary even in asymptomatic patients.[3] Histological criteria for an SLC include an outer layer of thin dense fibrous tissue, an inner epithelial lining (cuboidal or columnar), and lack of mesenchymal stroma or cellular atypia.[2,3]

Treatment of SLC by aspiration or surgery is usually not required; in the prenatal period, intervention is reserved only for extreme circumstances such as hydrops and instances where aspiration by percutaneous drainage of the cyst may help with symptoms.[19,20] In the neonate with a large lesion, surgical treatment consists of partial or total excision, with

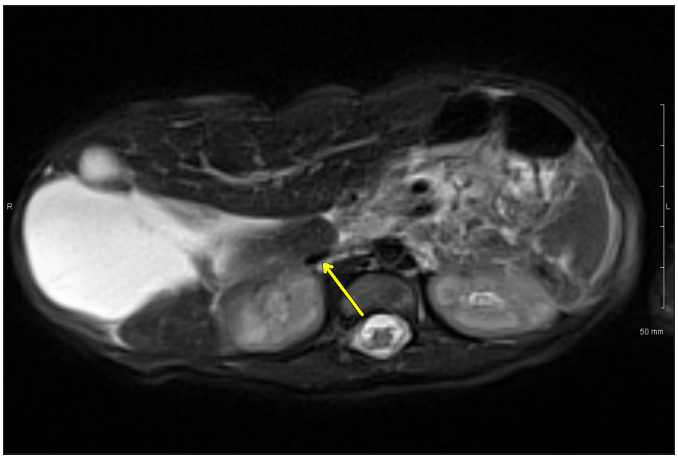

Figure 3. Postnatal MRI axial T2 image demonstrating a compressed IVC (arrow) and liver parenchyma anterior, posterior, and medial to the cyst.

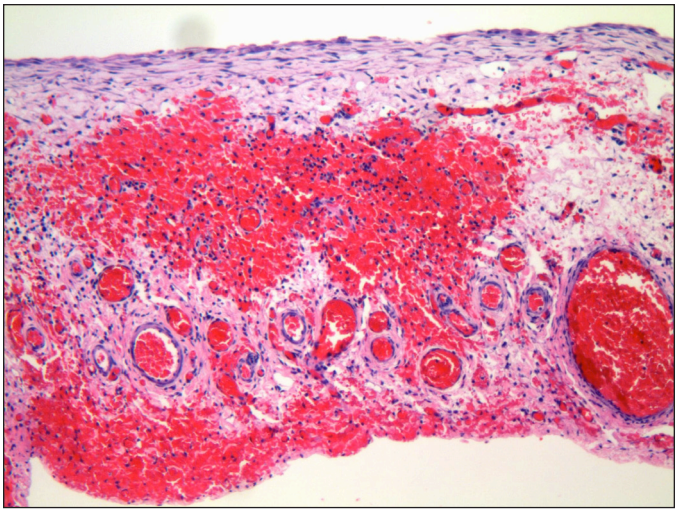

Figure 4. H&E stain (magnification 10x) showing the lining of the cyst composed of flattened squamoid epithelium, and the wall of the cyst composed of fibrovascular tissue.

a goal to excise as much of the cyst wall as possible without damage to the surrounding vital organs.[2,21] Laprascopic resection is safe and necessary to prevent further compression effects, especially IVC obstruction; total resection is preferred to avoid recurrence.[22] Percutaneous needle aspiration with a sclerosing agent injection is a safe and effective alternative treatment.[23-25] Cyst unroofing is associated with minimal complications.[17] Perioperative cholangiography should be performed in all cases to detect connections between the SLC and the biliary tree.[22] The postoperative course may be complicated by infection, cholangitis, and recurrence.[22] The infant must be watched closely after cyst aspiration for signs of re-accumulation, as this is common.[26] Total excision has the lowest recurrence and failure rate, making this the treatment of choice.[26]

Disclosures

Author contributions: E. Ropp gathered information, drafted and approved the manuscript, and is the article guarantor. N. Alviedo helped draft and approve the manuscript. A. Kent and I. Cohen gathered the radiographic images and drafted the manuscript.

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