

An unusual congenital hepatic cyst in an adolescent and review of differential diagnoses of complex liver cysts

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ABSTRACT

The diagnosis of a simple hepatic cyst is not difficult, but diagnostic confusion occurs when atypical features such as intracystic debris or extremely large size are present. In children, simple liver cysts are described as small, asymptomatic, and rarely hemorrhagic. We report an adolescent male presenting with an unusually large hepatic cyst that did not have typical imaging characteristics. The imaging findings and histology are displayed along with the differential diagnoses of complex liver cysts.

CASE REPORT

A 17 year old African American male presented to an ambulatory clinic with a large, uncomfortable, abdominal mass. The patient reported a remote history of trauma 4 months prior to presentation, when a cross bar with end weights had fallen on his upper abdomen. However, he stated that he did not have any significant abdominal pain or bruising at that time.

On physical exam, a non-tender bulging mass was palpated in the right upper quadrant of the abdomen. Laboratory results showed a slightly elevated AST of 51U/L, (normal 0-35U/L), an ALT of 25U/L (normal 0-45U/L), an elevated serum alkaline phosphate of 241U/L (normal 38 and 126 U/L), a total bilirubin of 0.9mg/dL (normal 0-1mg/dL), a direct bilirubin of 0.4mg/dL (normal 0-0.4mg/dL), hemoglobin level of 12.3g/dL (normal 13-16g/dL), and a slightly increased Prothrombin Time of 16.19 seconds (normal 1-15 seconds). Serum antibodies for Echinococcus and Entamoeba histolytica were negative.

A CT scan of the abdomen showed a large, smoothly marginated, hepatic cystic mass with punctuate calcifications along the periphery (Fig. 1 and Fig. 2), without enhancement. The abdominal ultrasound demonstrated a large, cystic, well-circumscribed mass. The posterior acoustic shadowing could not be evaluated due to the enormous size of the mass that measured 20.5 cm x 20.4 cm x 19.1 cm. This cystic mass contained floating debris and dependent solid like material (Fig. 3 and Fig. 4). The liver MR depicted a similar size, unilocular cyst within the right lobe of the liver, causing mass effect on the internal liver structures and displacement of the

inferior vena cava without parenchymal invasion. By MR, the cyst content was layered with intermediate signal on T1 and T2 sequences (Fig. 5-8).

Even though, the primary diagnosis was a simple liver cyst complicated by internal hemorrhage, other etiologies, including infection or a benign cystic tumor could not be excluded.

Because of the clinical symptoms, the patient was referred to surgery. 2.5 L of brown fluid was drained and the cyst was unroofed (Fig. 9 and Fig. 10). Histopathology showed a benign, nonparasitic, simple cyst of biliary or foregut origin with regions of mural hemorrhage, along with macrophages engorged with hemosiderin (Fig. 11 and Fig. 12).

DISCUSSION

Simple hepatic cysts are congenital lesions, usually lined with biliary-like epithelium, secreting a fluid similar to plasma (1). Their etiology is unclear, but they apparently arise from maldeveloped bile ducts (1). The prevalence of simple cysts in the general population is reported as 2.5-4.25% (1). They are encountered more often in women and in individuals older than 60 years (2). When found in children, they tend to be small (3). Cysts are classified as 'large' when they are greater than 4 cm in the longest dimension (1). The largest simple cyst in children we encountered in literature was post trauma, measuring 10x15 cm in size (4). Rare complications of simple hepatic cysts are right upper quadrant abdominal pain or

discomfort, early satiety, hemorrhage within the cyst, infection, intraperitoneal rupture (5), torsion of a mobile cyst, inflammation, and obstructive jaundice (1). If these cysts become symptomatic, then treatment includes cyst aspiration with sclerotherapy or “unroofing”.

Imaging of simple hepatic cysts is fairly characteristic. On ultrasound, they are well-circumscribed, echo-free, with good through transmission of sound waves, and indiscernible walls (1). On CT, they are usually round or oval shaped masses with thin, smooth walls and homogenous fluid attenuation (6). Rarely, calcifications may be seen (6). On MRI, simple cysts are hypointense on T1-weighted images and hyperintense on T2 (7).

If there is hemorrhage within the cyst, typically the ultrasound shows hyperechoic fluid. It may depict septations or solid portions (8). CT shows hyperdense or heterogenous fluid and MRI shows high signal intensity on T1 and T2 weighted images. The cyst may contain a thickened wall and a fluid-fluid level (9,10).

Our patient had complex imaging characteristics depicting heterogeneity on US suggestive of a bleed, but a solid component could not be excluded. The cyst lacked the typical appearance on CT and MRI. However, appearance is very variable depending on the age of hemorrhage.

Surgical intervention was necessary for both relieving the patient’s symptoms and to confirm the diagnosis of a congenital simple cyst with internal hemorrhage. We postulated that the hemorrhage occurred due to the episode of abdominal trauma versus the sheer large size of the cyst stretching blood vessels (11).

Differential diagnoses for cystic lesions in the liver include infection with echinococcus organisms, which cause hydatid cysts and echinococcosis. Hydatid cysts are characterized by a large, primary, mother cyst with a surrounding wall of inflammatory material and a germinal center from which arise daughter cysts (12). Patients are usually asymptomatic, but may present with right upper quadrant abdominal pain (12). In addition, infected patients usually show IgG and IgE antibodies to the organism with a specificity of 70% and a sensitivity of 100% (13). Hydatid cysts may at first appear similar to simple cysts on imaging (12). However, protoscolices and scolices in cysts appearing as snowflakes, along with a double-line sign, signifying a laminar structured wall surrounding fluid, may be seen by US (12). The CT usually shows a main cyst with higher attenuating floating protoscolices and scolices and daughter cysts with lower attenuation (14). On MR images, free-floating protoscolices and scolices are hypointense on T1-weighted images and distinctly hyperintense on T2-weighted images (12). Daughter cysts have lower intensity on T1-weighted images than the primary, mother cyst (12).

Entamoeba histolytica, the cause of Entamoebiasis, is another source of hepatic cysts and may be contracted by eating or drinking contaminated food or water. Patients have abdominal pain, fever, and leukocytosis, and have a history of diarrhea and weight loss although they may be asymptomatic. By ultrasound, amoebic abscesses are hypoechoic without wall echoes. They may be spherical or oval, similar to a simple cyst. There may be septae and debris, rarely gas bubbles, and hemorrhage on MR and CT (15).

Pyogenic abscess could present as a hepatic cystic lesion. They are often caused by bowel flora in an episode of ascending cholangitis although they also result from a bacteremia or sepsis, instrumentation, and from direct inoculation secondary to trauma. Presenting symptoms are abdominal pain, fever, leukocytosis, and peritonitis if the abscess ruptures. By ultrasound, abscesses may be hypoechoic and have poorly defined and irregular walls. Posterior acoustic enhancement, if present, is a reliable sign. In addition, there may be foci or diffuse echogenicity or debris within the fluid. Air may be present within the abscess. On CT one may visualize a “double-target” sign of an enhancing circle surrounded by a hypoattenuating zone signifying parenchymal edema. Gas may be present in up to 50% of abscesses and is strongly suggestive of an abscess. By MR, their walls may become thickened and more enhancing with time (15).

Cystic mesenchymal hamartoma of the liver is an uncommon benign tumor which occurs primarily in the first two years of a child’s life (16). It may be cystic or cystic-solid and results from ductal plate malformation (16). It is typically composed of myxoid mesenchymal tissue with tortuous and dilated cystic ducts, and presents as a rapidly evolving, asymptomatic upper abdominal mass (17). US typically shows a well-defined variable size cysts with multiple thin septae in the liver. Cysts may contain internal debris and a fluid-fluid level (18). US may also show a solid heterogenous component (18). Likewise, rounded fluid-filled cysts that range in size with enhancing intervening septae are seen on post-contrast CT. Moreover, on a T2-weighted MR scan solid components of the tumor and septae are seen as an intermediate high signal intensity and cystic portions are seen as high signal intensity (19).

Biliary cystadenoma is extremely rare in children occurring mostly in middle-aged women between the ages of 42–55 (20). It is a benign tumor with a potential for neoplastic transformation. It is usually a multilocular cystic tumor, but can be unilocular and difficult to distinguish from a simple hepatic cyst. On CT scan the tumor has fluid attenuation and may show fine septal calcifications. On MRI, it usually has hyperintense T2 and hypointense T1 signals (21,22). A fluid-fluid layer or variable intermediate signal intensities can sometimes be present depicting hemorrhage within the neoplasm (23).

In conclusion, the list of diagnostic possibilities is long when confronted with the presence of a complex hepatic cyst. Signs and symptoms may be misleading, and imaging features can be variable. In this report, an adolescent patient with an enormous liver cyst did not have the classical appearance of a simple cyst. Ultimately, tissue and fluid analysis was needed to establish the diagnosis of a hemorrhagic simple cyst.

TEACHING POINT

Very large simple liver cysts occur in children. They can be complicated by peripheral calcification and internal hemorrhage. The diagnosis can be difficult in these complex cases with a lengthy differential as described above.

ABBREVIATIONS

AST = Aspartate transaminase
ALT = Alanine transaminase
CT = Computed tomography
MR = Magnetic resonance (imaging)
US = Ultrasound
IgG/E = Immune globulin G/E
H&E = Hematoxylin and eosin

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FIGURES

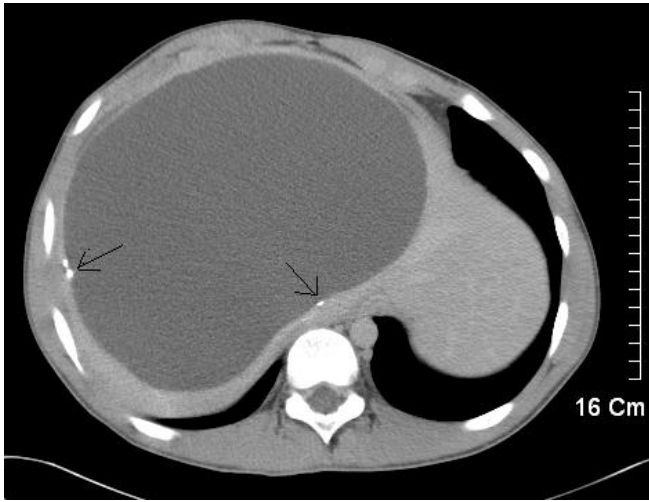


Figure 1: Non-contrast axial CT image though the liver shows a large fluid-attenuation cystic mass with punctuate peripheral calcifications (arrows).

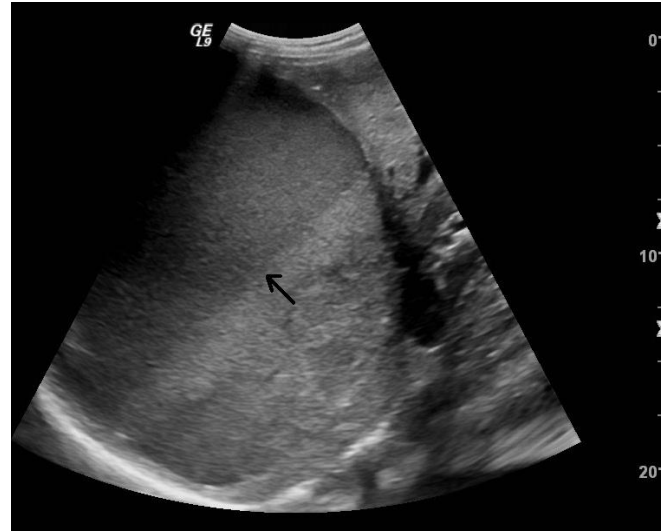


Figure 3: Sagittal grayscale ultrasound image of the liver demonstrates a large 20.5 x 20.4 x 19.1 cm complex cyst in the right liver lobe, containing fluid-fluid level (arrow) and debris.



Figure 2: Portal-venous phase axial image of a contrast enhanced CT of the abdomen shows a large cystic mass with mass effect on the hepatic vascular structures and the pancreatic head.

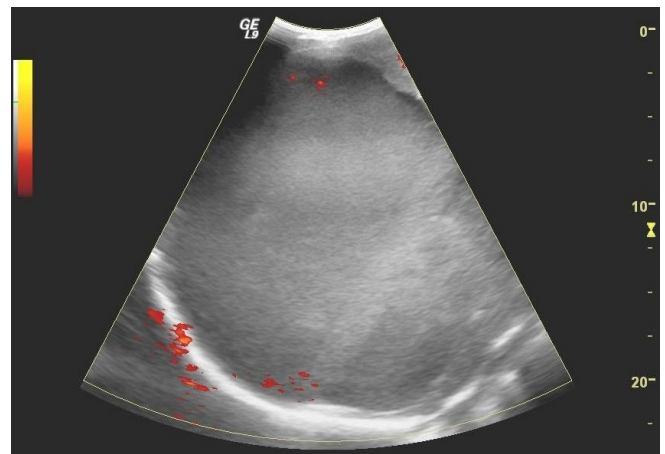


Figure 4: Sagittal ultrasound image with color Doppler demonstrating no vascularity within the cystic structure.

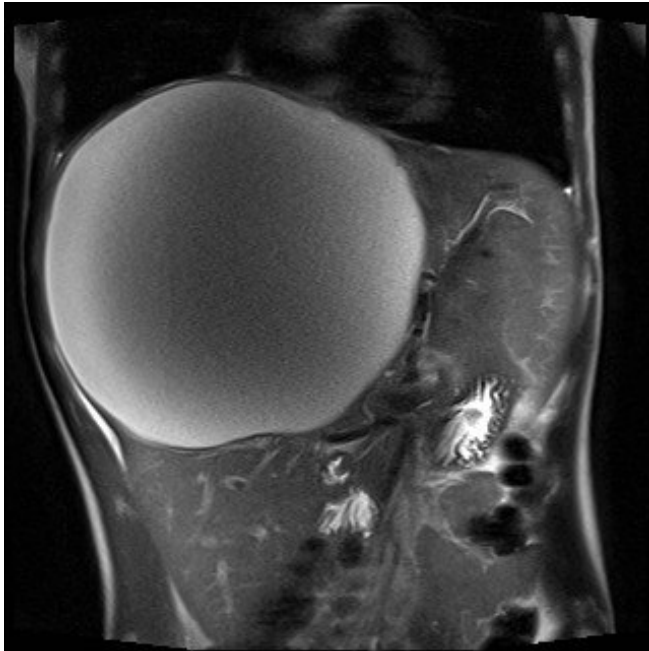


Figure 5: Coronal MRI HASTE sequence shows a large mostly intermediate T2 signal intensity cystic mass in the right lobe of the liver, representing complex fluid and possibly old blood.

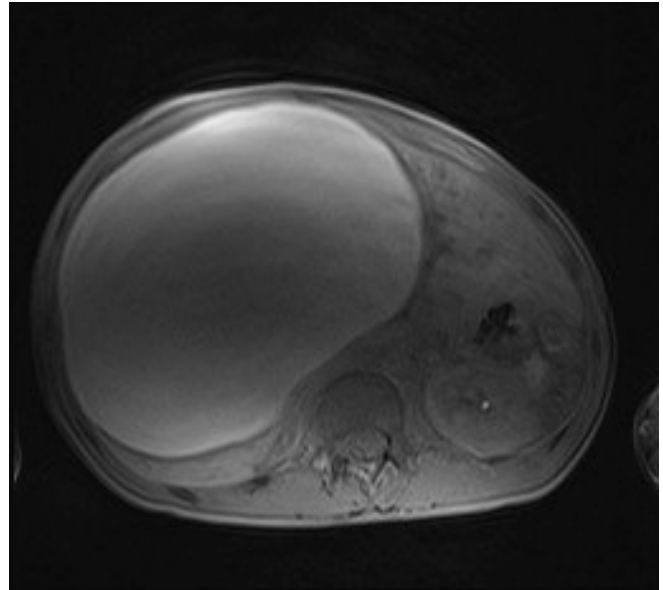


Figure 7: Axial pre contrast T1 f12d sequence with fat saturation of the liver shows mostly an intermediate T1 signal within the cystic liver mass.

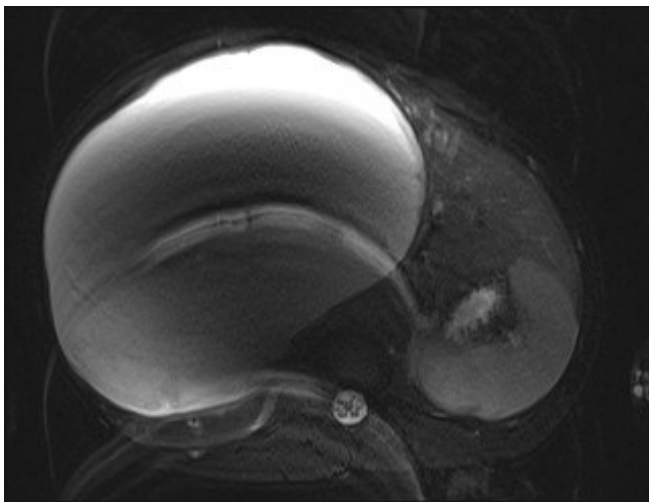


Figure 6: Axial T2 weighted image of the liver showed a brighter T2 signal fluid layering anteriorly within the cystic liver mass.

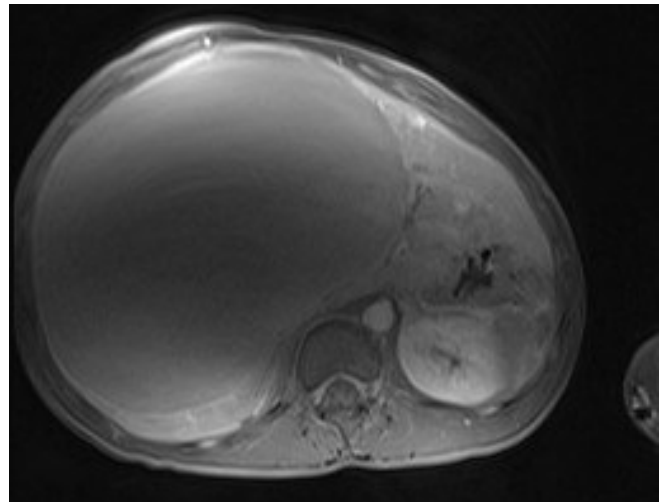


Figure 8: Axial post contrast (8 cc gadolinium) T1 f12d sequence with fat saturation of the liver shows mostly an intermediate T1 signal within the liver mass without contrast enhancement, supporting the possibility of old degraded blood products within the cyst.

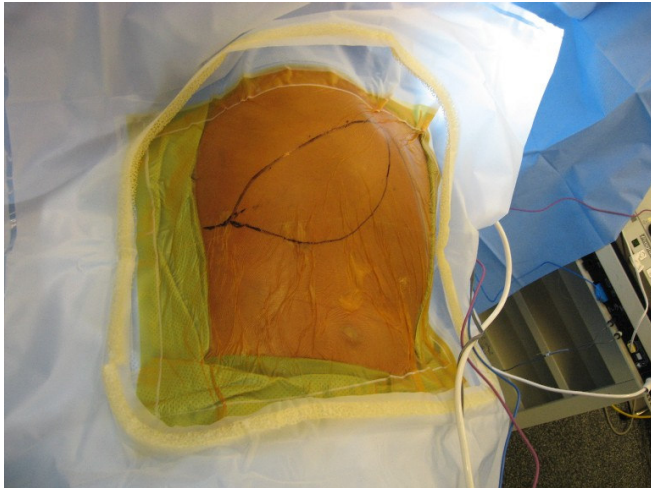


Figure 9: Intraoperative gross specimen showing the roof of the cyst removed during surgery.

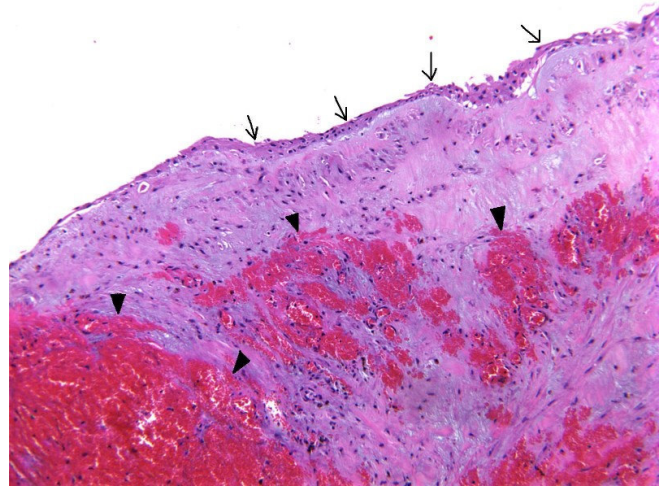


Figure 11: Image with H&E staining at 2x magnification depicting the lining of the cyst which consists of stratified squamous epithelium partly denuded (arrows) with murals of hemorrhage (arrow heads).



Figure 10: Postoperative gross specimen: demarcation of the clinically palpable outline of the liver cyst.

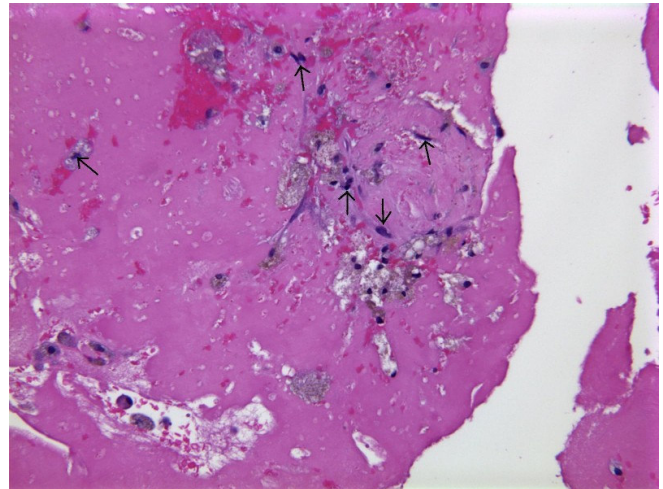


Figure 12: Image with H&E staining at 20x magnification depicting hemosiderin-laden macrophages in the content of the cyst (arrows).

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