

CASE REPORTS

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Symptomatic giant solitary bile cyst with intracystic bleeding in a 13-year-old girl: a case report and literature review

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Abstract

Background: Simple hepatic cyst is a rare disease in childhood. They occur in the general population with a prevalence of up to 5%. The incidence and size of cysts increase dramatically in adults older than 50 years. We performed a systematic literature review of all cases of simple hepatic cysts in the pediatric population undergoing surgical treatment. We found 52 cases reported in literature with a mean age of 2.54 years, 15% were pedicled cysts and only one case reported intracystic bleeding, detected only in the anatomopathological examination. We report a case of a 13-year-old girl who was presented with a symptomatic giant solitary bile cyst, the second biggest simple hepatic cyst reported in pediatric population, the biggest pedicled cyst and the only case of intracystic bleeding detected in pre-operative image examination in this group. This case brings important considerations about this complication and its pre-operative diagnosis.

Case presentation: We report a case of a 13-year-old girl with a giant solitary biliary cyst in the left hepatic lobe, symptomatic and with intracystic bleeding noted on magnetic resonance imaging. Diagnosis was difficult due to nonspecific symptoms and the non-typical images of simple hepatic cyst due to intracystic bleeding. The patient underwent a laparotomy, showing a large pedicled cyst, linked to segments II and III without adherence to other organs. Complete excision of the lesion was performed because the risk of torsion. The postoperative period passed without complications.

Conclusions: Although intracystic bleeding is the most common complication in adults, this is not reported in the pediatric population according to our review. Knowing how to recognize intracystic bleeding is important, because this complication predisposes the cyst to rupture and changes the appearance of the lesion on imaging tests, which can be more easily confused with cystadenoma or a cystadenocarcinoma. The magnetic resonance imaging is essential in the intracystic bleeding investigation. Comparisons between ultrasound and computed tomography findings associated with anemia history can prove the probability of this complication.

Keywords: Simple hepatic cyst, Congenital liver cyst, Nonparasitic hepatic cysts, Solitary bile cyst, Case report, Literature review

Background

Simple hepatic cyst is a congenital biliary malformation, its origin is derived in aberrant bile ducts that have lost communication with the biliary tree and continue to secrete intraluminal fluid [1, 2]. Numerous terms

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have been used to designate this cystic lesion in the literature, including biliary cyst, nonparasitic hepatic cysts, benign hepatic cyst, congenital liver cyst, unilocular cyst of the liver, and solitary liver cyst [1, 3]. They occur in the general population with a prevalence of up to 5% and are increasingly diagnosed due to the increased use of imaging methods [3–5]. Female-to-male ratio is 1.5:1 for all simple cysts shown at imaging or necropsy, while for symptomatic or complicated simple cysts is 9:1 [2]. Localization in the right hepatic lobe is twice as frequent as the left hepatic lobe, especially in segment 5 [6]. The incidence and size of cysts increase dramatically in adults older than 50 years [1, 2, 4]. Congenital solitary nonparasitic cysts of the liver are extremely rare in the pediatric population [6]. Surgical treatment is indicated for symptoms, complications, cysts with progressive increase in size and to rule out malignancy [1]. Complete excision has the lowest recurrence rates and good long-term results [7].

We performed a systematic literature review of all cases of simple hepatic cyst in the pediatric population (0–16 years) undergoing surgical treatment (Table 1). We found 52 cases with a mean age of 2.54 years, 51% were girls and 60% were symptomatic, with abdominal distension being the most common symptom. In the 29 cases reporting liver function tests, only 2 (7%) were altered tests. As for the location, 44% were exclusively in the right lobe and 29% were exclusively in the left lobe, the remaining cases occupied both hepatic lobes. All 52 cases were treated surgically, and in 56% complete excision of the cyst was performed, 96% of cases had an uneventful postoperative course with a mean follow-up time of 21 months. Fifteen percent of the cases were pedicled cysts, 9.6% had cystobiliary communication, and only 1 case reported intracystic bleeding, detected only in the anatomopathological examination.

We report a case of a 13-year-old girl who was presented with a symptomatic giant solitary bile cyst with intracystic bleeding. This case is the second biggest simple hepatic cyst reported in pediatric population, the biggest pedicled cyst, and the only one with intracystic bleeding detected in pre-operative image examination in this group. Intracystic bleeding is the most common complication of simple hepatic cyst in the general population [8], but rare in the pediatric population according to our review. This case brings important considerations about this complication and its pre-operative diagnosis.

Case presentation

A 13-year-old girl, melanodermic, presented with the history of sporadic abdominal pain a few years ago, worsened in the last three months, increased abdominal volume and feeling of heaviness in the abdomen. The pain

was initially mild, oppressive, located mainly in the right upper quadrant of the abdomen, without reports of anorexia, nausea, vomiting, or weight loss. At the physical exam, the patient was in good general condition, stained, hydrated, and anicteric. Cardiovascular and respiratory system examination showed no alteration. Absence of adenomegaly. Abdomen bulging in epigastric and right flank regions, presence of palpable mass, hardened and painful to deep palpation, with rounded margins and firm consistency.

Admission ultrasound (US) showed liver with normal size, regular contours, homogeneous sonic echotexture, and absence of biliary tract dilatation. Presence of expansive formation, predominantly solid with cystic areas, occupying almost the entire upper abdomen, measuring about 23 cm × 16 cm. Kidneys and spleen without alterations. Laboratory tests showed mild anemia, hemoglobin of 11.8 g/dL (normal 12–17 g/dL) and hematocrit of 36.7% (normal 36–50%). Electrolytes and liver function tests were normal. Alpha-fetoprotein and human chorionic gonadotropin were also measured and were within normal limits.

Abdominal computed tomography (CT) revealed a large abdominal expansile formation, in close relation to the lateral portion of the left liver lobe (Fig. 1A). It extends to the level of L4, dislocating the stomach, transverse colon, and small bowel loops, causing a bulging of the anterior wall of the abdomen (Fig. 1B). It had well-defined limits, encapsulated, regular contours, heterogeneous content of cystic predominance, and no evidence of calcifications or fats inside. The estimated size and volume were 25 cm × 22 cm × 12.5 cm and 3595 cm³, respectively. At magnetic resonance imaging (MRI) of the abdomen, the lesion presented predominantly high signal intensity on T2-weighted sequences, with heterogeneous hypersignal content on T1-weighted sequences, with no evidence of contrast enhancement, suggesting cystic lesion with hematic content due to intracystic bleeding. Absence of dilation of the bile ducts intra and extra-hepatic and laterally displaced gallbladder.

Among the diagnostic hypotheses, the possibilities of hepatic lesion with exophytic growth (mesenchymal hamartoma, with cystic predominance) and congenital cystic lesion with heterogeneous content due to probable bleeding were considered. Although the differentiation between a hepatic cyst with a blood clot and a hepatic cystic tumor is often difficult, in this case, as MRI well reflect the morphological features of a simple hepatic cyst with intracystic bleeding, they may provide the most important information for the diagnosis. In general, simple hepatic cysts are hypointense on T1 and hyperintense on T2; however, in the presence of intracystic bleeding, they are hyperintense on both T1 and T2 [9]. In addition,

Table 1 Simple hepatic cyst in pediatric population (0–16 years)

Author, Year of Publication	Age	Gender	Cyst Size (cm)	Hepatic Lobe	Pedicated Cyst	Symptoms	Liver Function	Cystobiliary Communication	Intracystic Bleeding	Surgical treatment	Outcome	Follow up
Allan, 2019	2 months	Female	5	Right and Left	No	Asymptomatic	-	No	No	Partial cyst excision and marsupialization	Uneventful	18 months
	1 year	Female	5	Right and Left	No	Asymptomatic	Normal	Yes	No	Partial cyst excision and cyst-jejunos-tomy	Uneventful	42 months
	2 days	Female	12	Left	Yes	Asymptomatic	-	No	No	Complete excision	Uneventful	36 months
	6 months	Female	10	Left	No	Asymptomatic	-	No	No	Partial cyst excision and marsupialization	Uneventful	36 months
	19 days	Female	10	Right and Left	No	Asymptomatic	-	No	No	Partial cyst excision	Uneventful	24 months
Kozlov, 2019	2 weeks	Male	5x6	Right	No	Malnutrition and palpable mass in abdomen	Normal	No	No	Complete excision	Uneventful	1 year
	10 days	Female	4x5	Left	No	Asymptomatic	Normal	No	No	Complete excision	Uneventful	1 year
	3 weeks	Female	5.5	Right	No	Asymptomatic	Normal	No	No	Complete excision	Uneventful	6 months
Kishore, 2018	15 months	Male	15x10x11	Right and Left	No	Decreased feeding and intermittent vomiting	Altered	Yes	No	Partial cyst excision and Roux-en-Y hepaticojejunostomy	Uneventful	1 year
Yadav, 2018	20 months	Male	5.9x4.7x5	Right	No	Asymptomatic	Normal	Yes	No	Complete excision and Roux-en-Y hepaticojejunostomy	Uneventful	1 year
Bhosale, 2016	3 days	Male	15x12x12	Right and Left	No	Abdominal distension and respiratory distress	Normal	No	No	Enucleation and marsupialization	Patient died	36 hours

Table 1 (continued)

Author, Year of Publication	Age	Gender	Cyst Size (cm)	Hepatic Lobe	Pedicated Cyst	Symptoms	Liver Function	Cystobiliary Communication	Intracystic Bleeding	Surgical treatment	Outcome	Follow up
Linden, 2016 [7]	7 weeks	Male	4.7x5.7	Right	No	Feeding intolerance and a palpable abdominal mass	-	No	No	Complete excision	Uneventful	1 year
	6 years	Male	4.5x5x5	Left	No	Abdominal pain	-	No	No	Complete excision	Uneventful	1 year
	5 days	Male	-	Left	Yes	Asymptomatic	-	No	No	Complete excision	Uneventful	1 year
	14 years	Male	25x11	Left	No	Asymptomatic	-	No	No	Complete excision	Uneventful	1 year
Banerjee, 2013	4 years	Female	18x14x19	Right	No	Abdominal distension, intermittent abdominal pain and vomiting	Normal	No	No	Drained and complete excision	Uneventful	1 year
Nordin, 2013	6 months	Male	4.5x2.3x4.0	Right	No	Asymptomatic	Normal	No	No	Aspirated and unroofed laparoscopically	Uneventful	2.5 years
Oh, 2012	8 days	Female	10x8x3	Right	No	Asymptomatic	-	No	No	Drained and partial cyst excision	Uneventful	6 months
Fabrizzi, 2009	11 years	Male	5.2x6.2x9.4	Right and Left	No	Abdominal pain	Normal	No	No	Aspiration and sclerotherapy	Uneventful	1 year
Aoyagi, 2007	13 years	Female	21x15x15	Left	No	Abdominal distension and abdominal pain	Normal	No	No	Left hepatic lobectomy	Uneventful	2 years
Rogers, 2007	22 days	-	12.5x8.5	Left	No	Feeding intolerance and abdominal distension	-	No	No	Left lateral sectionectomy	Uneventful	7 months
	13 months	-	5x4	Right and Left	No	Asymptomatic	-	No	No	Partial cyst excision	Uneventful	2 years
	15 years	Female	4x4	Left	No	Asymptomatic	-	No	No	Complete excision	Uneventful	10 months

Table 1 (continued)

Author, Year of Publication	Age	Gender	Cyst Size (cm)	Hepatic Lobe	Pediced Cyst	Symptoms	Liver Function	Cystobiliary Communication	Intracystic Bleeding	Surgical treatment	Outcome	Follow up
Soyer, 2007	21 days	Male	11x10.5	Right	No	Abdominal distension, jaundice and respiratory problems	Normal	Yes	No	Partial cyst excision and unroofing	Uneventful	10 days
Rygl, 2006	2 days	Male	8x7x5	Right and Left	No	Hepatomegaly	Normal	No	No	Partial cyst excision	Uneventful	15 days
	2 days	Female	10x9x5	Right and Left	No	Hepatomegaly	Normal	No	No	Complete excision	Uneventful	16 days
	15 days	Female	8x8x6	Right	No	Hepatomegaly	Normal	No	No	Complete excision	Uneventful	15 days
	5 months	Male	8x7x6.5	Right	No	Hepatomegaly	Normal	No	No	Complete excision	Uneventful	10 days
	8 years	Female	10x9x6	Right	No	Abdominal pain	Normal	No	Yes	Complete excision	Uneventful	13 days
	17 years	Female	8x6x7	Right and Left	No	Abdominal pain	Altered	No	No	Complete excision	Uneventful	10 years
Saxena, 2006	12 days	Female	2.1x5	Right	Yes	Asymptomatic	-	No	No	Complete excision	Uneventful	2 days
Otani, 2005	11 months	Male	13x13x9	Right	No	abdominal distension and loss of appetite	Normal	No	No	Partial cyst excision and unroofing	Uneventful	1 year
Berg, 2002	14 days	Female	7.5x4.4x4.6	Right and Left	No	Flatulence, constipation and feeding intolerance	Normal	No	No	Partial cyst excision and marsupialization	Complicated	1 year
Charles, 2001	8 years	Female	30x25x18	Right	No	Abdominal distension and nonspecific abdominal pain	Normal	No	No	Complete excision	Uneventful	7 days
Hackmon-Ram, 2000	1 week	Male	20x15	Right	No	Asymptomatic	-	No	No	Complete excision	Uneventful	-
Macken, 2000	4 months	Female	6.6x6.5x3.5	Right	No	Feeding intolerance	-	No	No	Complete excision	Uneventful	5 months
Raboei, 2000	4 months	Male	6.1x7.9	Right	No	Asymptomatic	-	No	No	Aspiration and sclerotherapy	Uneventful	6 years

Table 1 (continued)

Author, Year of Publication	Age	Gender	Cyst Size (cm)	Hepatic Lobe	Pedicated Cyst	Symptoms	Liver Function	Cystobiliary Communication	Intracystic Bleeding	Surgical treatment	Outcome	Follow up
Shankar, 2000	1 day	Female	20x20	Right	No	Abdominal distension, respiratory distress and vomiting	-	No	No	Partial cyst excision and marsupialization	Uneventful	-
Pul, 1995	22 months	Female	20x20x15	Right and Left	No	Abdominal distension	Normal	No	No	Aspiration, partial cyst excision and marsupialization	Uneventful	7 years
Quillin, 1992	1 week	Female	5	Left	No	Asymptomatic	-	No	No	Complete excision	-	-
Brown, 1990	12 weeks	-	2x1.8x1.5	Right and Left	No	Asymptomatic	-	Yes	No	Complete excision	Uneventful	5 months
Merine, 1990	Newborn	Female	14x8	Right	Yes	Abdominal distension and respiratory distress	-	No	No	Complete excision	Uneventful	1 year
Athey, 1986	2 months	Male	13.5x12.6x3.8	Right	Yes	Abdominal distension, respiratory distress and vomiting	-	No	No	Complete excision	Uneventful	4 days
Chu, 1986	5 days	Male	7.5x2.5	Left	Yes	Respiratory distress	-	No	No	Complete excision	Uneventful	-
Chung, 1986	1 day	Male	11x6x10	Left	No	Asymptomatic	-	No	No	Left hepatic lobectomy	Uneventful	9 days
Byrne, 1982	13 months	Male	9x10x18	Left	Yes	Abdominal distension	Normal	No	No	Drained and complete excision	Uneventful	7 years
Porter, 1982	5 months	Male	19x15x6	Left	Yes	Abdominal distension and hemihypertrophy	Normal	No	No	Complete excision	-	-
Rösch, 1978	3 weeks	Male	7x8	Right and Left	No	Asymptomatic	Normal	No	No	Partial cyst excision and marsupialization	Uneventful	18 months
	3 years	Female	14x17	Right	No	Abdominal distension	Normal	No	No	Complete excision	Uneventful	10 months

Table 1 (continued)

Author, Year of Publication	Age	Gender	Cyst Size (cm)	Hepatic Lobe	Pedicled Cyst	Symptoms	Liver Function	Cystobiliary Communication	Intracystic Bleeding	Surgical treatment	Outcome	Follow up
Desser, 1956	2 years	Male	12x20	Right	No	Abdominal distension, peri-orbital edema, enuresis and jaundice	Normal	No	No	Complete excision	Uneventful	1 year
	11 years	Male	-	Left	No	Abdominal distension	Normal	No	No	Marsupialization	Uneventful	8 years
	7 years	Female	25	Right	No	Intermittent abdominal pain, abdominal distension and melena	Normal	No	No	Drained	Uneventful	10 months



Fig. 1 Computerized tomography of the abdomen. **A** Contrast CT, late venous phase, showing a large cystic formation, in close relation to the lateral portion of the left liver lobe, displacing the stomach, transverse colon, and small bowel loops. **B** It extends to the level of L4, with well defined limits, encapsulated, regular contours and no evidence of calcifications or fats inside

disagreement between US (solid predominant lesion) and CT (cystic predominant lesion) findings is also important to establish the diagnosis of a simple hepatic cyst associated with intracystic hemorrhage, because US usually visualizes the intracystic blood clot such as papillary, nodular, or irregular septal images, and CT cannot clearly visualize the intracystic hemorrhage [10].

With the probable diagnosis of symptomatic and complicated simple hepatic cyst, surgical treatment was indicated. The patient underwent a supra and infra umbilical median laparotomy, showing a large cystic lesion, linked to segments II and III of the liver by a narrow bridge of hepatic tissue, without adherence to other organs (Fig. 2A, B). Complete excision of the lesion, which weighed 3890 g, was performed.

The postoperative period passed without complications, the patient discharged on the 5th day for follow-up

in the outpatient clinic. The anatomopathological study revealed a cystic lesion covered by fibrotic wall, with entrapment of biliary ductal structures and hepatocyte parenchyma, containing macrophagic-like cells inside, a large amount of cytoplasm, hematic, and necrotic material. Immunohistochemistry confirmed cystic lesion with hemorrhagic areas covered by mucosecretory columnar epithelium simple without atypia. In the lesion wall, there was entrapment of hepatocytes and bile ducts, besides positivity for cytokeratin in the lining cells, suggesting the diagnosis of solitary biliary cyst.

Conclusions

Simple hepatic cyst is a rare condition in childhood. The absence of symptoms, laboratory alterations or the presence of nonspecific abdominal manifestations make this diagnosis difficult. Imaging examination are essential and histopathological analysis is decisive for diagnostic confirmation. Although intracystic bleeding is the most common complication in adults, this is not repeated in the pediatric population according to our review. Knowing how to recognize intracystic bleeding is important, because in addition to being a factor that predisposes the cyst to rupture, it also changes the appearance of the lesion on imaging tests, from a homogeneous lesion to a heterogeneous one, which can be more easily confused with malignant lesions (cystadenoma or a cystadenocarcinoma). The MRI is essential in the investigation, because, in the presence of intracystic bleeding, the images are hyperintense on T1 and T2. Comparisons between US (solid predominant lesion) and CT (cystic predominant lesion) findings associated with anemia history can improve the probability of this complication.

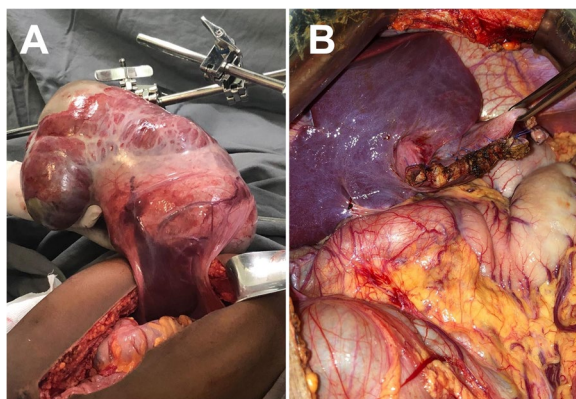


Fig. 2 Intraoperative images. **A** A pedicled cyst linked to the liver by a narrow bridge of hepatic tissue. **B** Final aspect of the left lobe of the liver (segments II and III) after complete excision of the lesion

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Authors' contributions

All authors assisted in study design, data collection, and manuscript preparation. All authors have given final approval to the manuscript.

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Declarations**Ethics approval and consent to participate**

The study was approved by the Institutional Review Board at our institution (Ethics Committee for Research with Human Beings HU-UFJF) and informed consent was obtained from the patient. The Ethical Appreciation Presentation Certificate is 47990021.8.0000.5133.

Consent for publication

Written informed consent was obtained from the patient for publications.

Competing interests

The authors declare that they have no competing interests.

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