Huge Non-parasitic Mesothelial Splenic Cyst in a Child: A Case Report and Literature Review

Imed Krichen1, Kais Maazoun1, Murad Kitar1, Naglaa M Kamal2, Ubaidullah Khan1, Mostafa YL Khalifi3, Rasha A1, Haifa Assiri3 and Kawthar Abdulrhim Bokari4

1Department of Surgery, Alhada Armed Forces Hospital, Taif, Saudi Arabia. 2Kasr Alainy Faculty of Medicine, Cairo University, Cairo, Egypt. 3Alhada Armed Forces Hospital, Taif, Saudi Arabia. 4Faculty of Medicine, Taif University, Taif, Saudi Arabia.

ABSTRACT

BACKGROUND: Splenic cysts are one of the relatively rare conditions in pediatric surgery practice. Primary non-parasitic splenic cysts are even more scarce.

CASE PRESENTATION: A 13-years-old female patient presented with chronic left hypochondrial pain of 2 months duration. Abdominal ultrasonography and computed tomography revealed huge 18 cm × 14 cm × 10 cm splenic cyst. Deroofing of the cyst was done which was complicated by secondary infection. Subsequently, the patient was re-operated on and partial splenectomy done with good outcome at 6 months follow up.

CONCLUSION: Partial splenectomy is the best management strategy for huge non-parasitic splenic cysts in children. There is also less recurrence rate of splenic cysts with preservation of splenic functions.

KEYWORDS: Spleen, cyst, children, surgical, huge, mesothelial

Introduction

Splenic cysts are generally rare and are usually discovered accidentally during imaging studies.1-4 The first splenic cyst described in literature was discovered at autopsy by Andral in 1829.3

Splenic cysts can be classified according to Martin's classification into; Martin splenic cyst class 1 that mean a true (primary) cyst with existing lining epithelium,4,5 and Martin splenic cyst class 2 that indicates absence of lining epithelium and is always considered to be secondary to direct splenic/or abdominal wall trauma, splenic infarction or abscess.6-8

In 2007, Mirilas and coworkers,9 critically reviewed literature for a valid classification of splenic cysts. They classified nonparasitic splenic cysts into primary and secondary cysts (Table 1). However, according to the International Society for the Study of Vascular Anomalies (ISSVA), splenic hemangioma, and lymphangioma are considered congenital malformations and not neoplastic lesions.10,11

For a long period of time; surgical management with removal of the spleen was the treatment of choice for splenic cysts and the first splenectomy for a cyst was done on 1867 by Splenotomy.12

Nowadays, the conservative school is the most agreed for management of small and asymptomatic cysts. Total splenectomy conducts to infectious complications, that is why conservative management is preferred. Deroofing and partial splenectomy have their particular indications while total splenectomy is rarely needed.

We here in report the first case of primary huge mesothelial cyst in a child from Saudi Arabia and discuss her management pitfalls.

Case Description

A 13 years old Saudi female presented to the pediatric emergency department with chronic left upper quadrant pain for 2 months. The patient was in her usual status of health until 2 months prior to presentation. The pain was localized in the left upper quadrant with no radiations, throbbing in nature, intermittent in course, aggravated by movement and relieved by rest. Inspiratory movements also aggravated the pain.

There was no associated history of fever, yellowish discoloration of the skin & mucus membranes, weight loss, change in appetite, headache, blurred vision, chest pain, hemoptysis, vomiting, bowel habits changes, urinary symptoms, or any other symptom suggestive of any body system affection. There was no history of abdominal trauma.

At the meantime, there were neither history of contact with animals nor travel to endemic areas of infections or infestations. She had none significant family and surgical histories.

On examination, the patient looked clinically well and vitally stable with average anthropometric measurements. Her abdomen was soft, but with a left upper quadrant mass detected by deep
palpation. The mass was immobile, hard, 18 cm × 14 cm × 10 cm in size, dull on percussion with no auscultatory findings. It was tender on palpation with pain score of 8 over 10.

Accordingly, the patient was admitted for workup. Basic laboratory testing including: complete blood picture, liver and kidney functions tests, C-reactive-protein, erythrocyte sedimentation rate and urine, and stool analyses were normal.

Her abdominal ultrasonography (US) showed huge splenic cyst which was hyper echoic with posterior enhancement. The study was completed by a computed tomography (CT) of the abdomen and pelvis which revealed large septate intra-splenic cyst with fluid collection measuring 18 cm × 14 cm × 10 cm, located at the upper pole of the spleen away from the hilum (Figure 1). Hydatid disease serology was performed to exclude parasitic splenic cyst (PSC) which came out to be negative.

The diagnosis of non-parasitic splenic cyst (NPSC) was retained and the patient underwent laparoscopic unroofing dome cystectomy.

On the third day post operation; the patient developed high spikes of fever and follow up US showed recollection with turbid fluid inside with an estimated size of 10.1 cm × 8.3 cm × 7.6 cm suggesting recollection with secondary infection. CT abdomen and pelvis confirmed the US findings (Figure 2).

Table 1. Classification of non-parasitic splenic cysts.7

| CLASSIFICATION | CRITERIA |
|----------------|----------|
| Primary        |          |
| Congenital     | Cyst lining: mesothelial, transitional, and/or stratified squamous. |
|                | Gross appearance of the cyst (interior): glistening white color, trabeculation |
| Neoplastic     |          |
| Angiomas       | Cystic lining: endothelial |
| Hemangiomas    | Blood content in cyst |
| Lymphangiomas  |          |
| Dermoid cysts  | Cyst lining: ectopic, mature ectodermal tissues |
| Secondary      |          |
| Traumatic      | Positive trauma history |
|                | Gross appearance of the cyst (interior): “shaggy, hemorrhagic,” normal splenic architecture |
|                | Cyst wall: thick, collagenous |
| Necrotic       | Infarct: pain in the left upper quadrant |
|                | History or active bacterial endocarditis |
|                | Nonspecific acute splenitis: for example, Typhoid fever Infectious mononucleosis Blood dissemination of hemolytic streptococcus Generalized lymphadenopathy |

Table reproduced with permission. Permission license attached as Supplemental File.

Figure 1. Preoperative abdominal CT scan picture of the splenic cyst.

Broad spectrum antibiotics were immediately started, and re-exploration laparoscopy was done. On laparoscopy, adhesions were found between the remnants of the cyst, the diaphragm and the great stomach curvature.

The surgeon decided to convert to open laparotomy due to the extensive adhesions to the diaphragm. Adhesions were cut,
vessels were divided, and an upper pole partial splenectomy resecting the whole upper pole of the spleen including the entire cyst was done with hemostasis sutures sutured at the cut edge as shown in Figure 3. This was followed by smooth uneventful postoperative course.

Histopathological assessment of the removed part of the spleen with the cyst revealed primary non-parasitic splenic mesothelial in origin (arrows in Figure 4(A)), there was no squamous metaplasia, there was no evidence of clear septation or trabeculations (Figure 4(A)). Immunostaining with PanKeratin and Calretinin were positive supporting the primary mesothelial nature of the cyst (stained brown in Figure 4(B)). CEA staining was negative.

Other differential diagnosis should be always kept in mind, including lymphangioma as well as hydatid cysts and pseudocysts.

The patient was discharged after completing the antibiotics course and is currently under regular follow up. Her 1, 3, 6-, and 12-months post-surgery follow up was uneventful with normal abdominal US with no recurrence.

Discussion
Splenic cysts are rare entity that is commonly coincidentally discovered in imaging studies specially during investigations for different gastrointestinal complaints, following abdominal trauma, during laparotomies or at autopsies.\(^1\)\(^,\)\(^2\)\ There are conflicting data about their incidence; in one report it was mentioned to be around 0.5\% to 2.0\% of the whole population\(^1\)\(^3\)\ while in another report of 42,327 autopsies, splenic cysts were found in only 0.07\% of the series.\(^1\)\(^4\),\(^1\)\(^5\)\ Up to our knowledge, there are only around 1000 reported cases of primary (1ry) splenic in literature.

As mentioned before; there are 2 main categories of splenic cysts according to presence or absence of a lining epithelium; true/primary splenic cysts (PSC) or secondary splenic cysts (SSC) respectively.\(^1\)\(^6\) PSC can be further subdivided into; parasitic (PPSC) and non-parasitic (NPPSC).\(^1\)\(^7\) In Europe and

---

Figure 2. Postoperative abdominal CT scan showing recurrence of the splenic cyst.

Figure 3. Post recurrence surgical splenic picture with resection of the cyst and upper pole of the spleen.

Figure 4. Histopathological picture of the resected cyst with mesothelial lining. (A) Hematoxylin and eosin. (B) PanKeratin and Calretinin immunostains.
North America, it is common to see the NPPSC, while in Africa and Central America it is commonly to see the parasitic ones.\(^{18}\) The NPPSC can be either congenital or neoplastic. The congenital cysts can be either epidermoid, endodermoid or dermoid cysts.\(^{19}\)

PSC accounts for 20% to 25% of all splenic cysts.\(^{20}\) Among PSC, epidermoid cysts constitute around 25%.\(^{21}\) The PSC were found to be most common in children, adolescents, and young adults.\(^{1}\) On the other hand, females were found to be more affected by splenic cyst in general and congenital type in particular.\(^{22,23}\)

Approximately 75% to 80% of total splenic cysts are SSC which can be either neoplastic (endothelial in origin like: hemangioma, lymphangioma) or even pseudocysts without any lining epithelium, which is can be a result of direct splenic/abdominal blunt trauma or splenic infarction, infection or abscess.\(^{24}\)

Like in our patient, splenic cysts usually symptomize when there is increase in size more than 8 cm due to compression of nearby organs like stomach and kidney. Our patient was symptomatic due to the huge cyst (18 cm × 14 cm × 10 cm) she had. She presented with abdominal pain which agreed with other reports which stated that abdominal pain is the most important complaint in symptomatic cases.\(^{27}\) Hypertension can occur in some patients due to renal artery compression.\(^{29,30}\)

Most patients present at pubertal age as the cyst growth was found to be affected by hormonal changes during puberty, especially around the age of 11 to 12 years old.\(^{31}\) Which coincides with the presentation of our patient.

When it comes to diagnosis, ultrasound as initial step followed by computed tomography are the most used modalities,\(^{32}\) and histopathology is the definitive diagnostic modality.\(^{25,33}\) We used the same strategy in our patient.

The management of NPSC remains controversial. Surgical resection is indicated for symptomatic or complicated forms, but no guidelines are available for asymptomatic NPSC. A recent report released in 2017 by Delporte and his colleague suggested a practical logical management algorithm for NPPSC Table 2 and Figure 5. Different surgical techniques are used for symptomatic cases including unroofing techniques used in our patient. When it comes to diagnosis, ultrasound as initial step followed by computed tomography are the most used modalities,\(^{32}\) and histopathology is the definitive diagnostic modality.\(^{25,33}\) We used the same strategy in our patient.

The post traumatic pseudocysts usually develop due to hematoma that failed to be collected under the splenic capsule or inside the parenchyma and they can be a result of direct splenic/abdominal blunt trauma or splenic infarction, infection or abscess.\(^{24}\)

The post traumatic pseudocysts usually develop due to hematoma that failed to be collected under the splenic capsule or inside the parenchyma and they can be a result of direct splenic/abdominal blunt trauma or splenic infarction, infection or abscess.\(^{24}\)

The management of NPSC remains controversial. Surgical resection is indicated for symptomatic or complicated forms, but no guidelines are available for asymptomatic NPSC. A recent report released in 2017 by Delporte and his colleague suggested a practical logical management algorithm for NPPSC Table 2 and Figure 5. Different surgical techniques are used for symptomatic cases including unroofing techniques used in our patient. When it comes to diagnosis, ultrasound as initial step followed by computed tomography are the most used modalities,\(^{32}\) and histopathology is the definitive diagnostic modality.\(^{25,33}\) We used the same strategy in our patient.

When it comes to diagnosis, ultrasound as initial step followed by computed tomography are the most used modalities,\(^{32}\) and histopathology is the definitive diagnostic modality.\(^{25,33}\) We used the same strategy in our patient.

| STUDY                        | NUMBER OF PATIENTS | MEAN AGE (YEARS) | TOTAL SPLENECTOMY | PARTIAL SPLENECTOMY | CYSTECTOMY | UNROOFING | RECURRENCE WITH TOTAL SPLENECTOMY | RECURRENCE WITH PARTIAL SPLENECTOMY | RECURRENCE WITH CYSTECTOMY | RECURRENCE WITH UNROOFING |
|-----------------------------|--------------------|-----------------|-------------------|---------------------|------------|-----------|--------------------------------|---------------------------------|--------------------------|--------------------------|
| Sinha and Agrawal\(^{29}\)   | 2                  | 11.5            | 0                 | 2                   | 0          | 0         | 0                              | 0                               | 0                        | 0                        |
| Fisher et al.\(^{30}\)      | 8                  | 12.5            | 0                 | 1                   | 0          | 7         | 0                              | 0                               | 0                        | 7                        |
| MacKenzie et al.\(^{31}\)   | 3                  | 11.5            | 0                 | 0                   | 0          | 3         | -                              | -                               | -                        | -                        |
| Czauderna et al.\(^{32}\)   | 50                 | 11.9            | 6                 | 26                  | 9          | 9         | 0                              | 1                               | 0                        | 7                        |
| Kimber et al.\(^{33}\)      | 6                  | 13.3            | 1                 | 5                   | 0          | 0         | 0                              | 0                               | 0                        | 0                        |
| Till and Schaarschmidt\(^{34}\) | 8             | 11.1            | 0                 | 0                   | 8          | 0         | 0                              | 0                               | 0                        | 0                        |
| Brown et al.\(^{35}\)       | 7                  | 10.8            | 1                 | 6                   | 0          | 0         | 0                              | 0                               | 0                        | 0                        |
| Jain et al.\(^{36}\)        | 1                  | 8               | 0                 | 1                   | 0          | 0         | 0                              | 0                               | 0                        | 0                        |
| Khan et al.\(^{37}\)        | 3                  | 11.6            | 0                 | 1                   | 0          | 0         | 0                              | 0                               | 0                        | 0                        |
| Musy et al.\(^{38}\)        | 13                 | 11.2            | 1                 | 0                   | 3          | 0         | 0                              | 0                               | 0                        | 0                        |
| Schier et al.\(^{39}\)      | 14                 | 6.5             | 0                 | 0                   | 14         | 0         | 0                              | 0                               | 0                        | 9                        |
| Total                       | 106                | 10.9            | 9                 | 44                  | 20         | 33        | 0                              | 1 (2.3%)                        | 1 (0.5%)                 | 24 (72.7%)               |

Table reproduced with permission. Permission license attached as Supplemental File.
The patient’s clinical course was complicated by infection which necessitated a second surgery with partial splenectomy. Most recent literature reported high recurrence rate and postoperative infection and/or bleeding.33,45

We admit that with this huge size of the mass, the choice of the deroofing was not appropriate and that partial splenectomy should have been the initial treatment of choice. Partial splenectomy preserves good tissue of the spleen with low recurrence rate.46

**Conclusion**

Partial splenectomy might be an excellent line of management of large splenic cysts to avoid recurrence and postoperative complications.

**Authors’ Contributions**

IK, KM, MK, UK, MK, RA: Diagnosed the patient, performed the surgical procedure, set the idea of the study and designed the study. HA, NK: collected data, reviewed literature, drafted the manuscript, critically analyzed the data. All authors reviewed and approved the manuscript for final publication.

**Availability of Data and Materials**

All data and materials related to the study are included in the current manuscript.

**Ethical Approval and Consent to Participate**

The study was approved by the research and ethical committee of the participating hospitals. All parents of enrolled children signed written informed consents for participation of their children in the current study.

**Consent for Publication**

All parents of enrolled children signed written informed consents for publication the current study.

**ORCID iD**

Naglaa M Kamal [https://orcid.org/0000-0002-8535-3838](https://orcid.org/0000-0002-8535-3838)
REFERENCES

1. Hansen MB, Moller AC. Splenic cysts. Surg Laparosc Endosc Percutan Tech. 2004;14:316-322.
2. Thippavong S, Duigenan S, Schindera ST, Gee MS, Philips S. Nonneoplastic, benign, and malignant splenic diseases: cross-sectional imaging findings and rare disease entities. AJR Am J Roentgenol. 2014;203:315-322.
3. Andral G. Splenic Cyst. Presid’ Anatomic Pathologique. Gabon; 1829.
4. Martin JW. Congenital splenic cysts. Am J Surg. 1958;96:302-308.
5. Garg M, Kataria SP, Sethi D, Mathur SK. Epidermoid cyst of spleen mimicking splenic lymphangiomata. Adv Biomed Res. 2013;2:49.
6. Ingle SB, Hinge CR, Jatal SN. An interesting case of primary epithelial cyst of spleen. J Clin Ultrasound. 1978;187:231-235.
7. Schwarts SI. The spleen. In: Schwartz SI, Harold E, eds. Maingot’s Abdominal Operations. 9th ed. Appleton and Lange; 1990:1039-304.
8. Mirilas P, Mentessidou A, Skandalakis JE. Splenic cysts: are there so many types? J Am Coll Surg. 2007;204:459-465.
9. Di Serafino M, Verde F, Ferro F, et al. Ultrasonography of the pediatric spleen: a pictorial essay. J Ultrasound. 2007;4:340-377.
10. Williams RJ, Glazer G. Splenic cysts: changes in diagnosis, treatment and aetiology. J Am Coll Surg. 1997;178:688-694.
11. Reddi VR, Reddi MK, Srinivas B, Sekhar CC, Ramesh O. Mesothelial splenic cysts: a report of three cases. Am J Surg Pathol. 1998;22:704-708.
12. Swartz SI. The spleen. In: Schwarts SI, Harold E, eds. Maingot’s Abdominal Operations. 9th ed. Appleton and Lange; 1990:1039-304.
13. Igaki K, Jimi A, Watanabe J, Kusaba A, Kojiro M. Epidermoid cyst of the spleen with CA19-9 or carcinoembryonic antigen productions: report of three cases. Am J Surg Pathol. 1998;22:704-708.
14. Macheras A, Misiakos EP, Liakakos T, Mpistarakis D, Fotiadis C, Karatzas G. Partial decapsulation of a splenic epithelial cyst: a case report. Adv Biomed Res. 2013;2:49.
15. Schwarts SI. The spleen. In: Schwartz SI, Harold E, eds. Maingot’s Abdominal Operations. 9th ed. Appleton and Lange; 1990:1039-304.
16. Nakae Y, Hayakawa T, Kondo T, et al. Epidermoid cyst occurring in a pancreatic accessory spleen. J Clin Gastroenterol. 1993;17:362-364.
17. Giovannoni A, Gioioli C, Goteri G. Tumours of the spleen. Cancer Imaging. 2005;5:73-77.
18. Klompmaker IJ, Haagsma EB, Slooff MJ. Splenic vein obstruction due to a solitary echinococcal splenic cyst, resulting in gastric fundus varices: an unusual cause of variceal bleeding. HPB Surg. 1995;2:2175-2180.
19. Klompmaker IJ, Haagsma EB, Slooff MJ. Splenic vein obstruction due to a solitary echinococcal splenic cyst, resulting in gastric fundus varices: an unusual cause of variceal bleeding. HPB Surg. 1995;2:2175-2180.
20. Hilmes MA, Strouse PJ. The pediatric spleen. Semin Ultrasound CT MR. 2007;28:3-11.
21. Karia N, Lakhoor K. Complicated congenital splenic cyst: saved by a splenectomy. Afr J Paediatr Surg. 2011;8:98-100.
22. Avital S, Kashtan H. A large epithelial splenic cyst. Am J Surg. 1978;187:231-235.
23. Fisher JC, Gurung B, Cowles RA. Recurrence after laparoscopic excision of nonparasitic splenic cysts. J Pediatr Surg. 2008;43:1644-1648.
24. Kurfi EA, Roustan F, Tsimoyiannakis EC. Surgical management of nonparasitic splenic cysts. JSLS. 2009;13:207-212.
25. Koutezou C, Haritopoulos KN, El Tayar AR, Hakim NS. Post-traumatic cyst of the spleen: a case report and review of the literature. Int Surg. 2002;87:152-156.
26. Tsakayannis DE, Mitchell K, Kozakewich HP, Shamberger RC. Splenic preservation in the management of splenic epidermoid cysts in children. J Pediatr Surg. 1995;30:1468-1470.
27. Qureshi MA, Hafner CD. Clinical manifestations of splenic cysts: study of 75 cases. Am J Surg. 1965;110:605-608.
28. Delorge F, Chaussey Y, Borrego P, et al. Management of nonparasitic splenic cysts in children. J Pediatr Surg. 1997;32:1465-1470.
29. Klompmaker IJ, Haagsma EB, Slooff MJ. Splenic vein obstruction due to a solitary echinococcal splenic cyst, resulting in gastric fundus varices: an unusual cause of variceal bleeding. HPB Surg. 1995;2:2175-2180.
30. Klompmaker IJ, Haagsma EB, Slooff MJ. Splenic vein obstruction due to a solitary echinococcal splenic cyst, resulting in gastric fundus varices: an unusual cause of variceal bleeding. HPB Surg. 1995;2:2175-2180.
31. Elit L, Aylward B. Splenic cyst carcinoma presenting in pregnancy. Am J Hematol. 1989;32:57-60.
32. Robertson F, Leander P, Ekberg O. Radiology of the spleen. Eur Radiol. 2003;13:207-212.
33. Schier F, Waag KL, Ure B. Laparoscopic unroofing of splenic cysts results in a high rate of recurrences. J Pediatr Surg. 2007;42:1860-1863.
34. Heidenreich A, Canero A, di Pasquale A. Laparoscopic approach for treatment of a primary splenic cyst. Surg Laparosc Endosc Percutan Tech. 2004;14:316-322.
35. Wu HM, Kortbeek JB. Management of splenic pseudocysts following trauma: a retrospective case series. Am J Surg. 2006;191:631-634.
36. Touloukian RJ, Mahajria A, Ghousoub R, Reyes M. Partial decapsulation of splenic epithelial cysts: studies on etiology and outcome. J Pediatr Surg. 1992;27:272-274.
37. MaKenzie R, Youngson G, Mahomed A. Laparoscopic decapsulation of congenital splenic cysts: a step forward in splenic preservation. J Pediatr Surg. 2004;39:88-90.
38. Czauderna P, Vajda P, Schausschmidt K, et al. Nonparasitic splenic cysts in children: a multicentric study. Eur J Surg Laparosc. 2006;16:415-419.
39. Kimber C, Piroo A, Drake D, et al. Hemisplenectomy for giant splenic cysts in children. Pediatr Surg Int. 1998;14:116-118.
40. Till H, Schausschmidt K. Partial laparoscopic decapsulation of congenital splenic cysts. A medium-term evaluation proves the efficiency in children. Surg Endosc. 2004;18:626-628.
41. Brown MF, Ross AJ, Bishop HC, Schnaufer L, Ziegler MM, Holcomb GW. Partial splenectomy: the preferred alternative for the treatment of splenic cysts. J Pediatr Surg. 1989;24:694-696.
42. Jain P, Parellkar S, Shah H, Sanghi B. Laparoscopic partial splenectomy for congenital splenic cysts. J Laparoendosc Adv Surg Tech A. 2008;18:899-902.
43. Khan AH, Renuossanu AL, Quinet A, Blanchard H, Grignon A, Noyerey M. Partial splenectomy for benign cystic lesions of the spleen. J Pediatr Surg. 1986;21:749-752.
44. Musy PA, Roche B, Belli D, Bugmann P, Nussle D, Le Coutre C. Splenic cysts in pediatric patients: a report on 8 cases and review of the literature. Eur J Pediatr Surg. 1992;2:137-140.
45. Sinha CK, Agrawal M. Nonparasitic splenic cysts in children: current status. Surgonlin. 2011;9:49-51.
46. Fisher JC, Gurung B, Cowles RA. Recurrence after laparoscopic excision of nonparasitic splenic cysts. J Pediatr Surg. 2008;43:1644-1648.