

Case Report

A rare gigantic primary epithelial cyst of spleen in an adolescent boy

Kiran George N.*, Gayatri Balachandran, L. N. Mohan

Department of General Surgery, St. John's Medical College, Bangalore, Karnataka, India

Received: 17 March 2017

Accepted: 20 April 2017

***Correspondence:**

Dr. Kiran George N.,

E-mail: kirangrign@gmail.com

Copyright: © the author(s), publisher and licensee Medip Academy. This is an open-access article distributed under the terms of the Creative Commons Attribution Non-Commercial License, which permits unrestricted non-commercial use, distribution, and reproduction in any medium, provided the original work is properly cited.

ABSTRACT

Cysts arising from spleen are a rare clinical entity, with their discovery usually being incidental. We present a case of the largest reported primary epithelial cyst of spleen. In present study a 16-year-old child presented with abdominal distension for 1 month associated with abdominal pain for 2 weeks. On examination, a mass was felt in the epigastrium, left hypochondrium and left flank, extending across from the right midclavicular line to the left midaxillary line, and extended inferiorly till 6cm caudal to umbilicus. CT abdomen revealed a unilocular cyst of 17x20x24cm arising from the spleen with a volume of 3700ml. Splenectomy was performed; intra-operatively there was a huge splenic cyst measuring 30x30x30cm. Histopathological examination was diagnostic of Primary congenital epithelial cyst of the spleen. Till date the largest congenital splenic cyst documented in literature is 20 cm × 13 cm × 21 cm, as reported by Valentina et al, in 2014. Present specimen bests this previous case considerably.

Keywords: Largest congenital epithelial cyst, Splenectomy, Splenic cyst

INTRODUCTION

Splenic cysts are generally classified as primary and secondary(pseudocyst).¹⁻⁵ Primary cysts have an epithelial lining and can be nonparasitic or parasitic (echinococcal). Worldwide, Echinococcus infection is the most common cause of a splenic cyst. Non-parasitic cysts include simple cysts, epidermoid cysts, and dermoid cysts and neoplastic.⁶ There have been reported in literature cases of primary splenic cystadenocarcinomas and of splenic lymphoma presenting as a splenic cyst.^{3,4,7,8} Isolated splenic metastasis is diagnosed in only 5.2% of case on autoptic studies.^{9,10} They are mostly secondary to melanoma and cancer of the breast, lung, ovary, colon, stomach and pancreas.⁹⁻¹² Although they usually appear as solid lesions, haemorrhagic phenomena, cystic or necrotic degenerations can occur, conferring to the metastasis a cystic feature.¹¹⁻¹⁴ Moreover a cystic adenocarcinoma of the pancreatic tail, extended within the splenic parenchyma should be excluded.^{2,14} Various classifications have been proposed for splenic cyst based

on whether they are lined with mesothelial, transitional, or epidermoid linings and also whether they are neoplastic, or secondary/pseudocyst (Figure 1).¹⁵

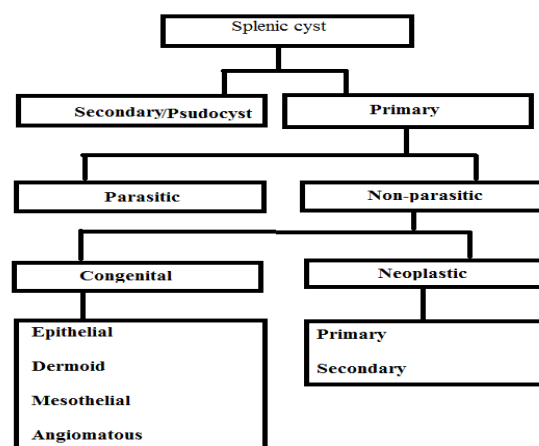


Figure 1: Classification of splenic cysts.

Primary epithelial congenital cysts are lined by flattened or cuboidal cells originating from in folding of peritoneal mesothelioma during splenic development. These lesions are usually small and asymptomatic and do not require excision. When these cysts are large and symptomatic, they can be removed by laparoscopic or open total or partial splenectomy.⁶

CASE REPORT

A 16-year-old male presented with abdominal distension for 1 month associated with abdominal pain for 2 weeks. Examination revealed a mass in the epigastrium, left hypochondrium and left flank which moved well with respiration. The swelling extended across from the right midclavicular line to the left midaxillary line, and extended inferiorly till 6cm caudal to umbilicus. Abdominal sonogram shows massive splenomegaly (16 cm size) with a large hypoechoic collection with internal echoes in the peri- pancreatic region extending upto spleen measuring 17x16x17.5cm, with volume of 2533ml (Figure 2).

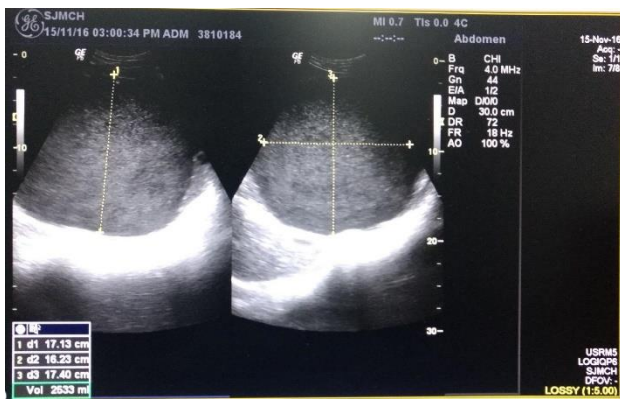


Figure 2: Abdominal sonogram shows massive splenomegaly (16 cm size) with a large hypoechoic collection with internal echoes in the peri- pancreatic region extending upto spleen measuring 17x16x17.5cm with volume of 2533ml.

Computed tomography of abdomen revealed a large, thin walled, unilocular cyst measuring 17.11x20.04x24.5 (APxMLxCC), with approximate volume of 3700ml, arising from the spleen. The cyst has thin imperceptible walls. There were no enhancing septa or mural nodules, nor any calcification or fat globules. No fluid-fluid level or debris were noted within the cyst. No daughter cysts were noted within the cyst (Figure 3 A, B).

A differential diagnosis of primary splenic cyst vs echinococcal cyst was considered the patient was prepared for surgery. Serology for E. granulosus was negative. After pre-treatment with meningococcal, pneumococcal and haemophilus influenza vaccines, the patient underwent surgery. A huge splenic cyst measuring 30x30x26cm was present with thinned out walls. Minimal residual splenic tissue was present at the

periphery of the cyst. Multiple collateral vessels were present. Stomach and pancreas were displaced to the right side of midline. (Figure 4 A-D).

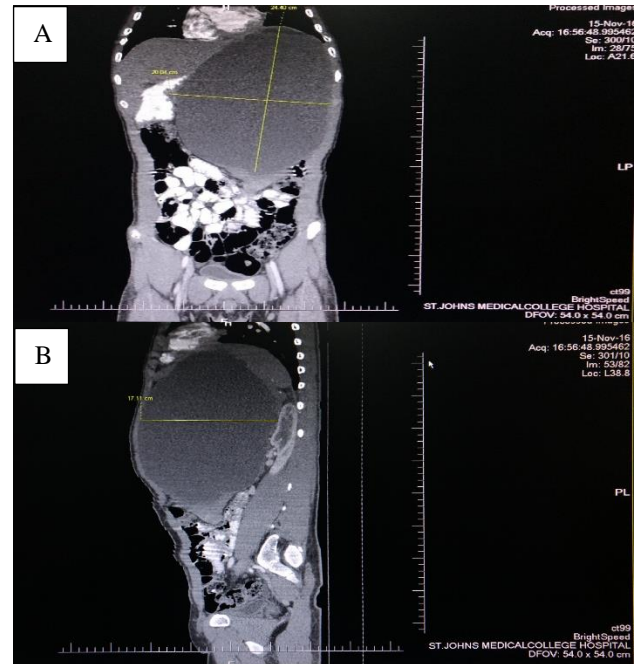


Figure 3: Computed tomography of abdomen revealed a large, thin walled, unilocular cyst measuring; a) 20.04x24.5cm in MLxCC (mediolateral & craniocaudal) dimension; b) 17.11cm in AP (Antero-posterior) dimension.

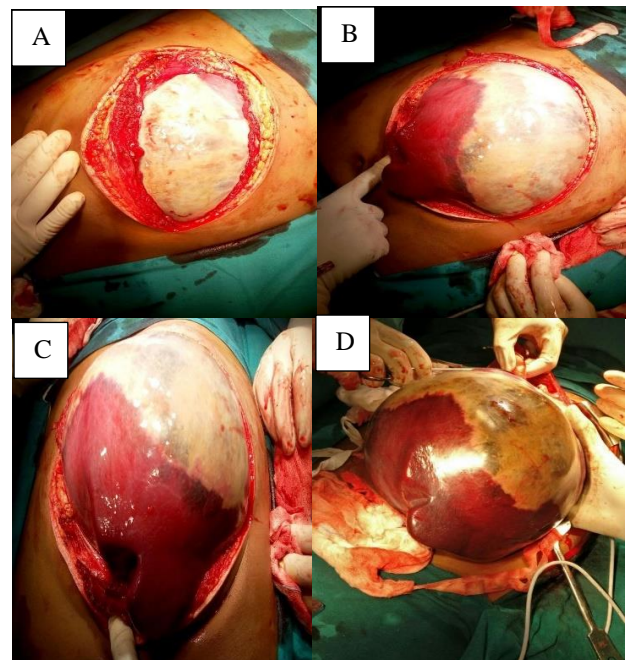


Figure 4 (A-D): Huge splenic cyst measuring 30X30X26CM with thinned out walls. minimal residual splenic tissue was present at the periphery of the cyst.

Splenectomy was performed and the final specimen weighed 5.2 kg. Histopathology of the specimen shows splenic tissue with portions of cyst wall lined by low cuboidal to columnar epithelium showing focal stratification. The cyst wall showed hyalinized connective tissue (Figure 5.1-2).

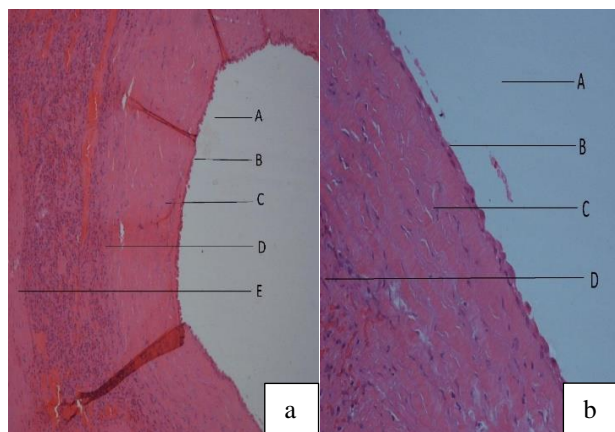


Figure 5: a) A-Inner aspect of cyst, B-Epithelial lining of cyst which is low cuboidal to columnar epithelium, C-Fibroconnective tissue of cyst wall, D-Normal splenic tissue, E-splenic capsule; b) In high power.

All above mentioned features were diagnostic of Primary Congenital Epithelial cyst of Spleen.

DISCUSSION

Splenic cysts are relatively rare entities, with an incidence of 0.07% as reported in a review of 42327 autopsies.¹⁶ The differential diagnosis for these lesions includes parasite infections (which is the commonest), results of previous trauma or infarction, congenital forms, primary splenic neoplasm or cystic metastasis etc. Non-parasitic cysts of the spleen are relatively uncommon. These include dermoid, epidermoid and epithelial cysts. Primary true cysts of the spleen account for approximately 10% of all nonparasitic splenic cysts.³⁻⁸ These cysts, however, are benign and apparently do not have malignant potential beyond that of the surrounding native tissue.

True splenic cysts are often asymptomatic and discovered incidentally. Patients may complain of abdominal fullness, early satiety, pleuritic chest pain, shortness of breath, and/or left shoulder or back pain. They may also experience renal symptoms from compression of the left kidney. On physical examination, an abdominal mass may be palpable. Rarely, splenic cysts present with acute symptoms related to rupture, haemorrhage, or infection. Diagnosis is best made by CT. The treatment of nonparasitic cysts depends on whether or not they produce symptoms.⁹⁻¹² Asymptomatic nonparasitic cysts may be observed with close follow-up by ultrasound to exclude significant expansion. Patients should be advised of the risk of cyst rupture with even minor abdominal

trauma if they elect for nonoperative management for large cysts. Small symptomatic nonparasitic cysts may be excised with splenic preservation, and large symptomatic nonparasitic cysts may be unroofed or splenectomy may be performed. These may be removed through either laparoscopy or laparotomy.¹³⁻¹⁶

Till date the largest congenital splenic cyst documented in literature was 20 cm × 13 cm × 21 cm, as reported by Valentina Pastore, et al, from the University of Foggia, Italy, in the year 2014.¹⁷ Our specimen bests this previous case considerably.

CONCLUSION

Splenic cyst is an uncommon clinical entity; and specifically non-parasitic splenic cyst is rare. Large symptomatic cysts necessitate intervention and histopathological confirmation of the same.

Funding: No funding sources

Conflict of interest: None declared

Ethical approval: Not required

REFERENCES

- Hansen MB, Moller AC. Splenic cysts. Surg Laparosc Endosc Percutan Tech. 2004;14:316-22.
- Brauner E, Person B, Ben-Ishay O, Kluger Y. Huge splenic cyst with high level of CA 19-9: the rule or the exception? Isr Med Assoc J. 2012;14:710-11.
- Elit L, Aylward B. Splenic cyst carcinoma presenting in pregnancy. Am J Hematol. 1989;32:57-60.
- Morinaga S, Ohyama R, Koizumi J. Low-grade mucinous cystadenocarcinoma in the spleen. Am J Surg Pathol. 1992;16:903-8.
- Palmieri I, Natale E, Crafa F, Cavallaro A, Mingazzini PL. Epithelial splenic cysts. Anticancer Res. 2005;25:515-21.
- Zinner MJ, Ashley SW. Maginot's Abdominal operations. McGraw-Hill Publishers. 12th Edi. 2016:1247.
- Nakashima A, Nakashima K, Seto H, Kamei T, Kakishita M, et al. Primary splenic lymphoma presenting as a large cyst. Radiat Med. 1994;12:42-5.
- Takabe K, Al-Refaie W, Chin B, Chu PK, Baird SM, et al. Can large B-cell lymphoma mimic cystic lesions of the spleen? Int J Gastrointest Cancer. 2005;35:83-8.
- Lam KY, Tang V. Metastatic tumors to the spleen: a 25-year clinicopathologic study. Arch Pathol Lab Med. 2000;124:526-30.
- Schon CA, Görg C, Ramaswamy A, Barth PJ. Splenic metastases in a large unselected autopsy series. Pathol Res Pract. 2006;202:351-6.
- Comperat E, Bardier-Dupas A, Camparo P, Capron F, Charlotte F. Splenic metastases: clinicopathologic presentation, differential diagnosis, and

- pathogenesis. *Arch Pathol Lab Med.* 2007;131:965-9.
12. Delaunoy T, Peny MO, Mignon M, Dili A. Splenic metastasis from gastrointestinal neoplasms: a review. *Acta Gastroenterol Belg.* 2012;75:3-4.
 13. Urrutia M, Mergo PJ, Ros LH, Torres GM, Ros PR. Cystic masses of the spleen: radiologic-pathologic correlation. *Radiographics.* 1996;16:107-29.
 14. Kaza RK, Azar S, Al-Hawary MM, Francis IR. Primary and secondary neoplasms of the spleen. *Cancer Imaging.* 2010;10:173-82.
 15. Morgenstern L. Nonparasitic splenic cysts: pathogenesis, classification, and treatment. *J Am Coll Surg.* 2002;194:306-14.
 16. Robbins FG, Yellin AE, Lingua RW, Craig JR, Turrill FL, et al. Splenic epidermoid cysts. *Ann Surg.* 1978;187:231-5.
 17. Pastore V, Bartoli F. A report of a giant epidermoid splenic cyst. *Afr J Paediatr Surg.* 2014;11:67-70.

Cite this article as: George KN, Balachandran G, Mohan LN. A rare gigantic primary epithelial cyst of spleen in an adolescent boy. *Int Surg J* 2017;4:2084-7.