Available online at www.sciencedirect.com

ScienceDirect

journal homepage: www.elsevier.com/locate/mjafi

Case Report

Epithelial cyst of spleen presenting as hypersplenism

Brig Jayant Kumar Banerjee (Retd)^a,
Lt Col Ramanathan Saranga Bharathi^{b,*}, Lt Col S. Venkatesan^c,
Col Giriraj Singh^d

^a Head (Gastrointestinal Surgery), Bharati Vidyapeeth Medical College, Pune 411030, India^b Classified Specialist (Surg & GI Surg), Command Hospital (Southern Command), Pune 411040, India^c Assistant Professor, Dept of Path & Hematology, Armed Forces Medical College, Pune 411040, India^d Senior Advisor (Radiology), Command Hospital (Eastern Command), Kolkata 700027, India

ARTICLE INFO

Article history:

Received 28 September 2015

Accepted 23 October 2016

Available online 30 November 2016

Keywords:

Epithelial cyst of spleen

Primary splenic cyst

Nonparasitic splenic cyst

Hypersplenism

Laparoscopic splenectomy

space on percussion. Laboratory investigations showed low Hemoglobin (Hb) – 9 gm%, platelets – $9 \times 10^4/\text{mm}^3$, and borderline total leukocyte count (TLC) – 4100/cumm. The remaining investigation, including hydatid serology, was unremarkable. Computerized tomography (CT) showed a well-defined lobulated and septated cystic lesion measuring 9 cm × 7 cm × 6 cm, located in the posterior and medial aspect of spleen, peripherally. Medially, the cyst was abutting left diaphragmatic crus and posterior aspect of pancreatic body

Introduction

Epithelial splenic cysts are rare. Their presentation as hypersplenism is rarer. We present such an exceptional case, share its images, and provide an overview of the topic.

Case report

An 18-year-old male patient presented with dragging pain in left upper abdomen. There was no history of trauma. Examination revealed pallor and dullness in Traube's

Fig. 1 – Axial CT image showing splenic cyst.

* Corresponding author.

E-mail address: sarangabharathi@gmail.com (R. Saranga Bharathi).<http://dx.doi.org/10.1016/j.mjafi.2016.10.010>

0377-1237/© 2016 Published by Elsevier B.V. on behalf of Director General, Armed Forces Medical Services.

Fig. 2 – Coronal reformatted CT image showing splenic cyst.

and tail. Inferiorly, it was abutting the left adrenal gland and upper pole of left kidney (Figs. 1–3). There were no features suggestive of portal hypertension. The patient underwent laparoscopic splenectomy following immunization against capsulated organisms and had an uneventful recovery (Fig. 5). His blood counts improved postoperatively as the following: TLC – 6800/cumm and platelets – 4.2 lakhs/cumm, immediately, while, Hb – 11.2 gm% improved after 2 weeks. On histopathological examination, the spleen showed a cyst with fibrocollagenous wall lined by low cuboidal epithelium (Fig. 4). The remaining splenic parenchyma revealed areas of expanded red pulp with dilated sinusoids, fibrosis, and hemosiderin-laden macrophages. The patient is well on a follow-up of 1 year and 3 months.

Discussion

Splenic cysts are rare (0.05% of autopsies) and are either parasitic (75%) or nonparasitic (25%) in nature.¹ Presence or absence of epithelial lining divides the nonparasitic cysts further into primary/true cysts and secondary/pseudocysts, respectively.¹

True cysts are further classified as per the cells lining them into: simple/mesothelial cyst – cuboidal/low columnar cells; epidermoid cyst – squamous cells; dermoid cyst – squamous cells with dermal appendages, including, hair follicle/sebaceous gland, etc.; and angiomatous cysts – endothelium.² Except the last one that is referred to as lymphangioma or hemangioma, depending upon the fluid contained in the cyst, the others constitute epithelial cysts of spleen.²

Fig. 3 – Sagittal reformatted CT image showing splenic cyst.

Fig. 4 – Photomicrograph showing splenic parenchyma with expanded red pulp and fibrocollagenous cyst wall lined by low cuboidal epithelium. (Hematoxylin–Eosin stain. Original magnification ×100.)

Fig. 5 – (A) Laparoscopic picture showing the entry into the lesser sac by division of short gastric vessels. (B) Cranial most short gastric vessels being divided. Splenic hilum is exposed showing the cyst inferior to the hilum. (C) Spleen completely dissected off its attachments. (D) Spleen delivered out of the abdomen. Outer surface of the spleen is seen.

Pseudocysts are formed following trauma, abscess, necrotizing infection, or degeneration.¹ However, opinion is divided on the etiopathogenesis of epithelial cysts. Some believe them to be congenital, formed by embryonic inclusions of epithelial cells from adjacent structures and consequent metaplasia, while others attribute their formation to developmental invagination of splenic capsular mesothelium and subsequent fluid accumulation.^{1–3}

Epithelial cysts are seen in 2nd or 3rd decade of life, as evident from our case.^{1,2} Most are asymptomatic or minimally symptomatic, with dull pain and palpable mass in the left upper abdomen.^{1,2} Minority may present with acute abdomen due to intracystic bleed, rupture, or infection. Very few present with hypersplenism, as our case did.⁴ On attaining a large size, the cyst may produce symptoms owing to compression or pressure on adjacent organs like kidney or stomach, where the presentation may be hydronephrosis or early satiety/vomiting/dysphagia.^{1–4}

Epithelial cysts appear as well-defined, thin-walled anechoic, unilocular lesions. However, they may be, occasionally, multilocular as well. They may contain echogenic debris in cases of intracystic bleed/infection.^{1–4} On CT, they appear as well-defined rounded, low-attenuating lesions. However, as in ultrasonography, the attenuation may increase depending on the complications mentioned above.^{1–4} On magnetic resonance imaging, the lesion appears bright on T2-weighted images. Image-guided aspiration does not provide much clue as to the diagnosis.

Imaging aids the diagnosis; however, histopathology confirms whether it is a true cyst or a pseudocyst.⁵ Tumor markers such as CA19.9 and carcinoembryonic antigen (CEA) may be raised in the serum as well as the fluid in the epithelial

cysts.⁵ Immunohistochemistry (IHC) may aid in discerning the origin of splenic cyst.⁵ CD31/34 and factor VIII positivity indicate endothelial origin. Cytokeratin positivity indicates epithelial/mesothelial origin. Further, CEA and CA19.9 positivity are seen in cysts of epithelial origin while they are negative in cysts of mesothelial origin. However, tumor markers and IHC are done when in doubt or for academic purpose. It is not mandatory for successful treatment or follow-up; hence, it was not done in our case.

Imaging, in addition, helps in formulating the treatment plan based on size, location, and extent of splenic involvement. Options include deroofting, marsupialization, fenestration, and partial/complete splenectomy – all of which can be done either by open surgery or laparoscopy.^{1–4} Splenic preservation is desirable, but we opted for complete splenectomy, owing to hypersplenism, large cyst size, and abutment of cyst with splenic hilum. Laparoscopy was chosen owing to benefits of minimal invasion. Image-guided aspiration with injection of sclerosant is an option for smaller cyst (<5 cm) but not an appropriate choice for such large cysts, especially, near splenic hilum.¹

Summing up, epithelial cyst of spleen is rare, whereas its presentation as hypersplenism is rarer. Imaging facilitates the diagnosis while histopathology clinches it. Modality of management, extent of resection, and surgical approach are often determined by the size and location of the cyst. Laparoscopy proves to be a viable treatment option.

Conflicts of interest

The authors have none to declare.

Acknowledgements

Able assistance of Havaldar Satish Kumar, Havaldar Sanjay Nayak, Havaldar Perumal, and Havaldar Ghosh, pivotal to the success of the surgery, is gratefully acknowledged.

REFERENCES

1. Mathur SK, Sangwan M, Singh R, Dahiya N. An unusual case of epidermoid splenic cyst. *Trop Gastroenterol*. 2014;35:62–64.
2. Manoj MG, Misra P, Kakkar S. Multilocular epithelial cyst of spleen: a rare occurrence. *Ind J Pathol Microbiol*. 2012;55:602–604.
3. Avital S, Kashtan H. A large epithelial splenic cyst. *N Engl J Med*. 2003;349:2173–2174.
4. Ellatif MEA. Giant splenic cyst with hypersplenism: laparoscopic splenectomy. *J Gastroenterol Hepatol Res*. 2013;2:549–551.
5. Palmieri I, Natale E, Crafa F, Cavallaro A, Mingazzini PL. Epithelial splenic cysts. *Anticancer Res*. 2005;25:515–522.