DOI: 10.7860/JCDR/2025/78496.22027

Case Report

Pathology Section

Giant Splenic Cyst with Spectacular Gross Morphology: A Case Report

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ABSTRACT

Splenic cysts are rare lesions with a documented prevalence of 0.07% to 2%. Primary splenic cysts have an epithelial lining. A 32-year-old female presented with a complaint of abdominal pain and a dragging sensation on the left side. On examination, a soft lump was palpable 20 cm below the left costal margin. A Computed Tomography (CT) scan of the abdomen revealed splenomegaly and a large, smooth-walled cyst. The diagnostic work-up for splenic cysts typically involves a combination of clinical assessment, imaging studies, and laboratory tests. The imaging modalities can provide information about the size, location, and nature of the cyst. Ultrasound can confirm the cystic nature of the lesion, while CT and Magnetic Resonance Imaging (MRI) offer more detailed visualisation of the cyst's internal structure, septations, and wall characteristics. Radiological examination may not be sufficient to differentiate between various types of splenic cysts. In endemic regions, it is difficult to distinguish from non-parasitic cysts based on imaging alone and additional diagnostic measures are necessary. Cyst fluid examination, such as fluid amylase, titre of echinococcus and other biomarkers, may aid in differentiating between cysts in the pancreatic tail, parasitic cysts and malignancy. Image-guided biopsy or partial or complete splenectomy, and histopathological examination are necessary for definitive diagnosis. In this case, the cyst on histopathological examination showed features of a primary splenic mesothelial cyst. Primary splenic mesothelial cysts are extremely rare lesions with a documented prevalence of 0.07% to 2%. There is limited available literature, which merits the documentation of this case report. The various classifications of splenic cysts, clinico-radiological and morphological findings of mesothelial cysts are discussed in this case report.

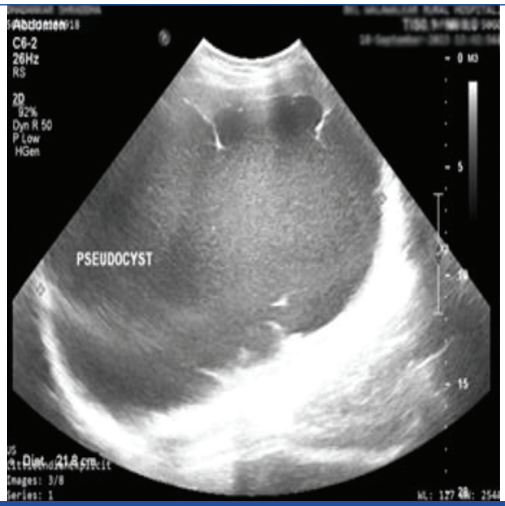
Keywords: Mesothelial splenic cyst, Primary non-parasitic splenic cysts, True splenic cyst

CASE REPORT

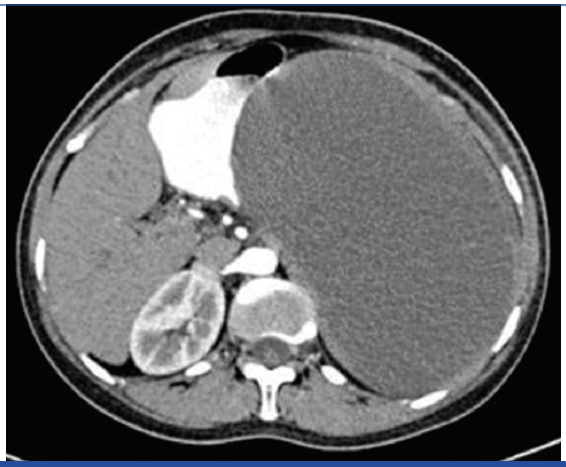
A 34-year-old female presented with a complaint of abdominal pain and a dragging sensation in the left hypochondrium for the last six months. For the last two months, she developed nausea and anorexia and lost 4 kg of weight. There was no history of vomiting. She was non-diabetic, non-hypertensive and did not have any major illness in the past. An abdominal examination revealed tender splenomegaly extending up to the umbilicus. Besides microcytic hypochromic anaemia, other investigations were normal. On ultrasound, a 21×10×17 cm complex cystic lesion was discovered in the left paraumbilical region, which appeared to have arisen from the spleen [Table/Fig-1a]. The cyst showed dense echoes and a few thin septations within. There was no evidence of solid components and calcification. The impression of a complex splenic cyst was given with a remote possibility of a pseudocyst of the pancreas.

A CT scan showed a grossly enlarged spleen measuring 21.8 cm along its axial length with a large smooth-walled cystic lesion of approximate size 16.9×9.5×19.7 cm [Table/Fig-1b]. The cyst showed an imperceptible wall with non-enhancing thin septation at its superior pole. No calcification, fat, or acute haemorrhage was noted. Internal homogeneous fluid density was 24 to 28 Hounsfield Units (HU). The enlarged spleen was displacing the stomach, left kidney, and pancreas toward the right side. A large smooth extrinsic impression was seen over the stomach with a maintained intervening fat plane. The findings were reported as favouring splenic pseudocyst or epithelial cyst over hydatid cyst. As the cyst was giant and symptomatic, involving the entire splenic parenchyma, total splenectomy was performed. The splenectomy specimen measured 19×13×5.5 cm, externally smooth. The spleen was replaced by an unilocular cyst with compressed splenic parenchyma of 10×4 cm at the periphery. The inner surface showed thin white fibrous tree root-like trabeculations [Table/Fig-2].

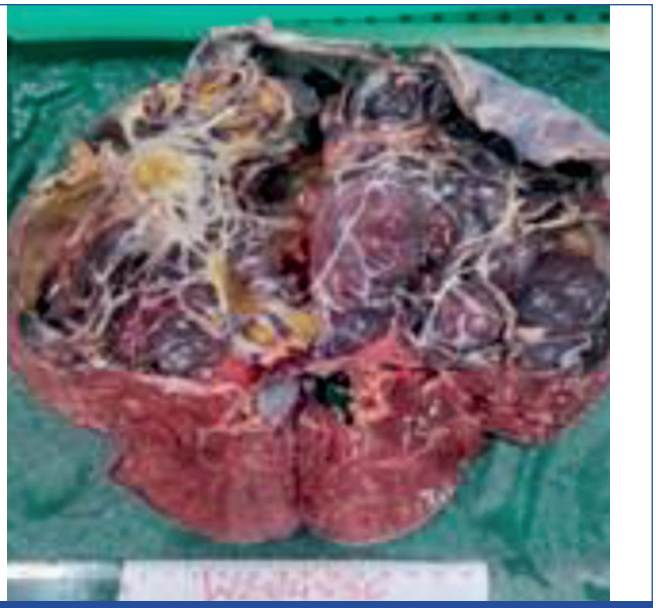
Multiple sections from the cyst wall showed mesothelial-type cuboidal lining epithelium [Table/Fig-3a] with squamous metaplasia



[Table/Fig-1a]: Ultrasonography (USG) abdomen: large complex splenic cyst.

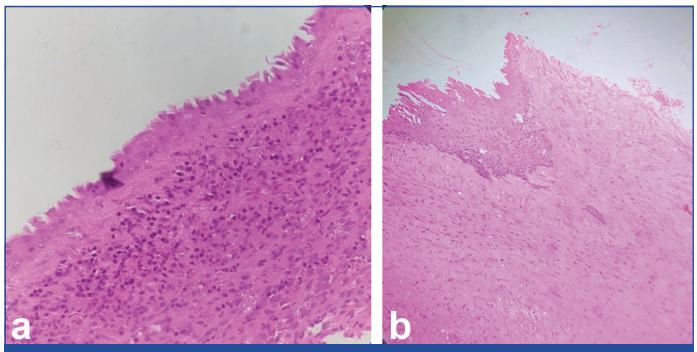


[Table/Fig-1b]: CT scan: Splenic cyst compressing the stomach.



[Table/Fig-2]: Gross-tree root-like trabeculation of splenic mesothelial cyst.

[Table/Fig-3b]. The wall and surrounding splenic parenchyma showed congestion and haemorrhage. A final diagnosis of primary splenic mesothelial cyst was provided after excluding haematological and epithelial malignancy. On follow-up, it was noted that she gained weight and her symptoms were relieved.



[Table/Fig-3]: a) Splenic cyst wall lined by cuboidal mesothelial cells {Haematoxylin and Eosin (H&E), 400x}; b) Mesothelial lining with focal squamous metaplasia (H&E, 400x).

DISCUSSION

Splenic cysts are rarely encountered lesions. The cysts with the presence of epithelial lining are primary (true) cysts. The cysts with low cuboid mesothelial lining on microscopy are labelled as splenic mesothelial cysts. These splenic mesothelial cysts, on gross examination, have characteristic tree-root trabeculations. The splenic mesothelial cysts are rare and usually found in children and young adolescents [1]. We encountered this spectacular splenic pathology in a 32-year-old adult female.

There are three classifications of splenic cysts. Martin JW and Martin RH classified splenic cysts as primary (epithelial/true) cysts and secondary (false/pseudo) cysts based on the presence or absence of epithelial lining [2,3]. A variety of lesions like angiomas, granulomatous lesions, abscesses, infarcts, metastatic tumour deposits and nodules of lymphoma may present in the spleen as cysts. All these cystic lesions are secondary and do not show any epithelial lining.

These secondary cysts constitute 80% of total splenic cysts. Primary (true) splenic cysts have an epithelial lining. According to the cyst wall lining, the cysts are further classified as epidermoid (stratified squamous lining without adnexal structures), dermoid (squamous lining with adnexal structures), and mesothelial (cuboidal to low columnar lining) cysts.

According to recent classification by Morgenstern L, splenic cysts are classified based on the pathogenesis into congenital (primary epithelial cysts), neoplastic (haemangioma and lymphangioma), infectious (mostly hydatid cysts), traumatic, and degenerative cysts. Congenital (primary epithelial cysts) are occasionally reported in the literature with documented prevalence from 0.07% to 2% [4,5]. Primary epithelial cysts have an epithelial lining and, as per the cyst wall lining, they are further classified as epidermoid (stratified squamous lining without adnexal structures), dermoid (squamous lining with adnexal structures) and mesothelial (cuboidal to low columnar lining). The various hypotheses put forward for the development of primary splenic epithelial cysts are as follows [6]:

1. Mesothelial invagination theory-surface mesothelial in vagination with subsequent cyst formation;
2. Embryonic inclusion of epithelial cells from adjacent structures and their metaplasia leads to the formation of cysts with various types of epithelial lining, like squamous, columnar and mesothelial;
3. Lymph space theory suggests that cysts can originate from the spleen's normal lymphatic spaces.

Primary cysts are encountered routinely in children and young adolescents, but few cases have been reported in young adults and in elderly individuals [7,8]. Individuals with mesothelial splenic cysts are usually asymptomatic and are noted incidentally. Symptoms like abdominal pain, increasing abdominal girth, referred left shoulder pain, nausea and vomiting may occur. These symptoms arise due to pressure effects on adjacent structures by a splenic cyst more than 5-8 cm. This patient had complaints of abdominal pain, nausea, and vomiting as the cyst was encroaching upon the stomach.

On ultrasound examination, mesothelial cysts reveal an echoic lesion with acoustic enhancement. On CT and MRI, these cysts show fluid-filled masses with low T1 and high T2 signals depending upon the nature of the fluid. Radiological investigations confirm the cystic nature of the lesion but cannot classify it further [9]. The final categorisation is done by histopathological examination only. Radiological findings in this case revealed a huge smooth-walled homogeneous fluid density cystic lesion of 16.9×9.5×19.7 cm with non-enhancing thin septation. The cyst was compressing the stomach and was the reason for nausea and weight loss in this case.

Elevated serum levels of CA19-9 and Carcinoembryonic Antigen (CA)125 have been reported in a few cases [10]. These markers were not estimated in the current case. If the lining epithelium of the cyst is attenuated, the diagnosis can be made by demonstrating these markers by immunohistochemistry or by increased serum levels. They can also be used to know the post-treatment recurrence. Depending on the symptoms, size and location of the cysts; age; and condition of the patient, the management of splenic cyst varies. Larger or symptomatic cysts and cysts with rupture need surgical intervention. Various surgical techniques opted are radiologically guided aspiration of cyst fluid, marsupialisation, fenestration, laparoscopic decapsulation, partial or total splenectomy by laparoscopy or laparotomy. The Puncture, Aspiration, Instillation, and Reaspiration (PAIR) technique is used where surgery is risky, like in antenatal cases and includes the puncture of the cyst, aspiration of cyst contents, injections to sterilise the cysts and respirations. In cases of total splenectomy, proper vaccination should be carried out to avoid overwhelming postsplenectomy infections. In this case, as the cyst was 16.9×9.5×19.7 cm and symptomatic, the patient underwent total splenectomy by open laparotomy. The primary splenic cysts are usually single but can be multiple. On cut-section, cysts are unilocular or multilocular, thin or thick-walled with a white to grey smooth glistening cyst wall and shows prominent fibrous tree root trabeculations. Intra-cystic fluid consistency varies from thin transparent serous to viscous with or without blood. Surrounding splenic parenchyma shows focal areas of haemorrhage and infarct. Gross findings in our case showed a huge splenic cyst replacing the

entire cyst and with characteristic tree-like whitish trabeculations.

Various differential diagnoses of splenic cysts considered were:

1. Traumatic or inflammatory cysts;
2. Haemangiomas, lymphangiomas, cystadenomas;
3. Cystic teratomas;
4. Cystic degeneration of carcinoma/sarcomas, especially angiosarcoma;
5. Primary non-parasitic cysts.

Histopathological examination helped to differentiate between these. Primary epithelial cysts are lined either by low cuboidal to columnar mesothelial cells or transitional or squamous epithelium. Cysts with squamous lining are classified as epidermoid cysts, while those with mesothelial lining are classified as mesothelial cysts. However, mesothelial epithelium frequently shows focal squamous metaplasia. Immunohistochemistry differentiates between these. Epidermoid cysts show positivity for CK5/6, while mesothelial cysts, besides CK5/6, are also positive for calretin and WT1. Traumatic cysts do not show any epithelial lining. In endemic areas, 50-80% of splenic cysts are hydatid cysts caused by Echinococcus granulosus, which has characteristic gross and microscopic findings. Hydatid cyst on microscopy shows an outer acellular laminated membrane and an inner transparent nucleated germinal membrane with attached protoscolices. Splenic haemangiomas show variable-sized blood vessels lined by bland endothelium. Lymphangiomas on microscopy also show cystic spaces lined by endothelial cells filled with proteinaceous material, along with a variable number of lymphoid cells. Haemangiomas are positive for CD31, CD34, ERG and negative for D2-40, while lymphangiomas are positive for D2-40. Sarcomas with cystic degeneration or angiosarcomas show cytological atypia.

In this case, the splenic cyst showed characteristic tree root trabeculations on gross examination. Multiple sections from the cyst wall revealed a cyst wall lined by cuboidal mesothelial cells with focal squamous metaplasia. Hence, the final diagnosis of splenic mesothelial cyst was provided. The literature review mentioned genomic assessment in very few cases of multiple mesothelial cysts and epidermoid cysts. Three germline variants noted are Hemicentin 1 (HMCN1) and Contactin2 (CNTN2)on1q and DDHD1on14q.

There are a few questions about splenic mesothelial cysts that remains unanswered, like the exact incidence of splenic mesothelial cysts and the exact pathogenesis. Hence, more and more case reports and case studies of cystic splenic lesions and their further uniform classification should be reported.

CONCLUSION(S)

Primary epithelial splenic cysts, either epidermoid or mesothelial, are rare and difficult to diagnose preoperatively. Histopathological examination is the gold standard and mandatory for the confirmation of the diagnosis. Though rare, primary splenic mesothelial cyst should be considered as one of the differential diagnoses while evaluating splenic cystic lesions, especially single cystic lesions.

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PLAGIARISM CHECKING METHODS: [Jain H et al.]
- Plagiarism X-checker: Feb 12, 2025
- Manual Googling: Jun 26, 2025
- iThenticate Software: Jun 30, 2025 (7%)

ETYMOLOGY: Author Origin

EMENDATIONS: 6

AUTHOR DECLARATION:
- Financial or Other Competing Interests: None
- Was informed consent obtained from the subjects involved in the study? Yes
- For any images presented appropriate consent has been obtained from the subjects. Yes

Date of Submission: **Feb 09, 2025**
Date of Peer Review: **Apr 05, 2025**
Date of Acceptance: **Jul 02, 2025**
Date of Publishing: **Nov 01, 2025**