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Case Report

Large multiloculated splenic mesothelial cyst: A rare case report

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ABSTRACT

The mesothelial splenic cyst is a rare clinical entity, comprising <10% of all the primary splenic cysts. They usually present as a well-defined unilocular cystic lesion. We present a large multiloculated mesothelial splenic cyst in middle-aged women, which was radiologically indistinguishable from hydatid cyst. We discuss the importance of the clinical, radiological, and histopathological correlation in the diagnosis of this rare non-parasitic cyst of the spleen.

KEY WORDS: Computed tomography, epithelial cyst, hydatid cyst, mesothelial cyst, splenic cyst

INTRODUCTION

Splenic cysts are rare with reported incidence of 0.07% in a large autopsy series and 0.5% among all the splenectomies done [1,2]. The splenic cysts are broadly classified into parasitic cysts (75%), mostly caused by *Echinococcus granulosus* and non-parasitic cysts (25%). The non-parasitic splenic cysts are further categorized into primary or true (20%) and secondary or false (80%) cysts based on the presence or lack of inner cellular epithelial lining [1]. Based on the cellular lining, the primary splenic cysts are further sub-classified into three types: (a) Epidermoid cysts (stratified non-keratinizing squamous), (b) Mesothelial cysts (low cuboidal to low columnar), and (c) Dermoid cysts (squamous lining with dermal structures) [3]. Splenic mesothelial and dermoid cysts are very rare with the epidermoid cysts alone accounting for 90% of all primary splenic cysts [4].

We report this rare case of splenic mesothelial cyst in an adult female, detailing its clinico-radiological and pathologic features and discuss the dilemmas encountered during its diagnostic evaluation.

CASE REPORT

A 44-year-old previously healthy female presented with chief complaint of left loin dragging pain, on and off since 1 month. There was no history of fever, weight loss or trauma. Physical examination revealed moderate splenomegaly with a firm consistency. Ultrasonography of abdomen revealed a large, well-defined, multiloculated cystic lesion in spleen and a provisional diagnosis of splenic hydatid cyst was proffered.

Computed tomogram (CT) of the abdomen revealed splenomegaly with a well-defined, non-enhancing, thin

walled, multiseptated cystic lesion measuring ~ 10.2 cm × 9.7 cm × 10 cm involving upper and mid pole of spleen with no calcifications/solid components/hemorrhage within [Figure 1a and b]. A thin rim of normal splenic parenchyma was seen surrounding the lesion. The differentials included splenic hydatid cyst and congenital epidermoid cyst. Routine hematologic and biochemical examinations were normal. Casoni's test was negative.

Explorative laparotomy with splenectomy was performed, and a large splenic cyst was identified surrounded by compressed splenic parenchyma. Post-operative period was uneventful. The patient remains asymptomatic at 6 months follow-up.

The surgical specimen of the spleen measured 13 cm × 10 cm × 9 cm and weighed 493 g. The cyst measured 10 cm × 9 cm × 8.5 cm was multilocular and contained serous fluid. The inner surface showed yellow fatty and membranous areas [Figure 2]. Microscopically, the cyst wall was lined by cuboidal mesothelial epithelium with areas of stratification [Figure 3a and b]. The underlying hyalinized fibrotic stroma showed foci of calcification, giant cell reaction, and cholesterol crystals. Cyst wall was surrounded by compressed splenic parenchyma which showed unstimulated white pulp and congested red pulp. Multiple sections from different areas did not reveal any areas of squamous epithelium, thereby ruling out an epidermoid cyst. On immunohistochemistry, the cyst wall showed strong positivity for calretinin throughout the lining epithelium [Figure 4] confirming the mesothelial origin of the cyst. The lining epithelium also showed cytokeratin positivity. A diagnosis of true mesothelial cyst of the spleen was made.

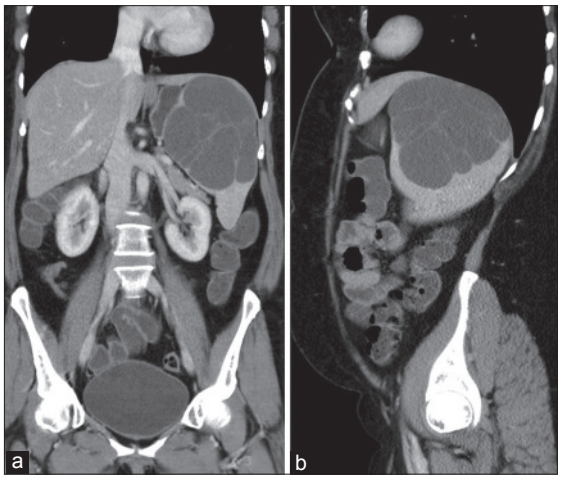

Figure 1: Computed tomography scan abdomen (a) coronal and (b) sagittal views show a large thin walled multiloculated hypodense lesion showing enhancing thin internal septations in the upper-mid zone of the spleen

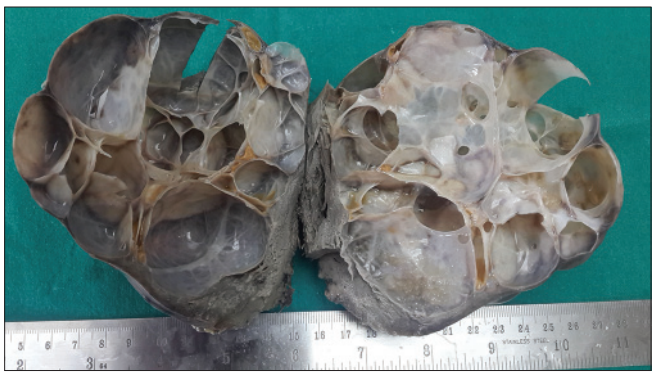

Figure 2: Cut section of spleen shows a multiloculated cyst with surrounding compressed splenic parenchyma

DISCUSSION

Primary cystic disease of the spleen shows female preponderance and occurs predominantly in children and young adults. The clinical presentation varies from asymptomatic (30-60%) to left hypochondrium pain, abdominal fullness/discomfort, nausea, vomiting and complications following its rupture, infection and intracapsular hemorrhage [1,5].

Even though the epithelial splenic cysts are considered to be congenital cysts, its origin remains debatable. Various theories have been proposed regarding its etiopathogenesis and include: (a) Developmental displacement and subsequent metaplasia of epithelial tissue from surrounding structures during embryogenesis, (b) Invagination or entrapment of peritoneal mesothelium within the spleen during development, and (c) Metaplasia of heterotopic endodermal inclusions in the splenic parenchyma [1,3].

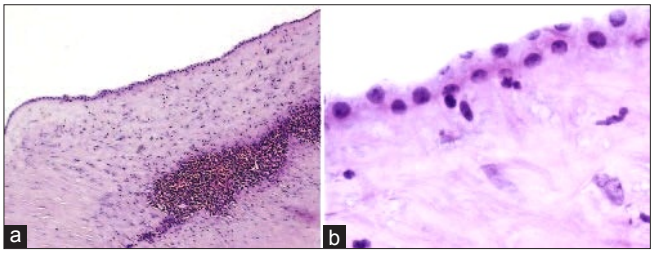

Figure 3: Microscopy shows cyst wall lined by cuboidal mesothelium with focal areas of stratification (a) (H and E, ×40), (b) (H and E, ×400)

Figure 4: Cyst epithelial lining showing positivity for calretinin (immunohistochemistry, ×400)

The various enumerated causes of splenic cyst are hydatid cyst, post-traumatic hematoma/pseudocyst, pancreatic pseudocyst, epidermoid cyst, abscess, mesothelial cyst, dermoid cyst, infarction, and tumors like large B-cell lymphoma, metastasis, hemangioma, and lymphangioma [1,6].

The pre-operative diagnosis of primary splenic cyst remains elusive due to its rarity of occurrence and non-specific imaging features. Ultrasonography is usually the first line of radiological investigation, and the true splenic cysts have been described as well-defined, thin-walled unilocular anechoic mass with occasional internal echoes due to cholesterol crystals or hemorrhage. Occasionally, internal septations and wall calcifications have also been described. On CT, the true splenic cysts appear as a rounded, hypodense water attenuation lesion showing non-enhancing thin imperceptible walls. The internal septations may show contrast enhancement. Magnetic resonance imaging (MRI) confirms the cystic nature of the mass as they are hypointense on T1-weighted and intensely hyperintense on T2-weighted sequences; however, they may appear hyperintense on T1-weighted images, consequent to internal hemorrhage or high proteinaceous content [6,7].

The primary differential of true splenic cyst is a hydatid cyst. They both share common imaging features of unilocular/multilocular cystic mass, thin non-enhancing walls with internal echoes and wall calcifications. However, a well-defined hypointense rim of maximum thickness 4-5 mm on both T1 and T2 weighted, more pronounced on T2-weighted MRI has been reported to be an important characteristic of hydatid cyst, differentiating it from other cystic non-parasitic lesions [8]. Abscess and metastasis can be differentiated on the basis of their rim enhancement. Hemangiomas can be differentiated on the basis of its feature of delayed contrast enhancement and it can also show central punctate calcifications. False cysts may be differentiated on the basis of the antecedent history of trauma or pancreatitis. Differentiating lymphangioma from true cyst is practically impossible on imaging [8].

Grossly, the splenic mesothelial cysts are usually unilocular with a glistening inner surface and often show trabeculations. Cyst fluid may demonstrate cholesterol crystals and macrophages on microscopy. On histology, the wall may be lined by flattened to cuboidal simple or stratified epithelium [9]. A confounding picture of a combination of epithelial cells is occasionally encountered, but its occurrence can be explained by the pluripotent nature of the mesothelial cells [3]. Multiple sections from different areas are recommended, as the lining epithelium may be occasionally missed, leading to an erroneous diagnosis of the secondary cyst.

Immunohistochemistry may help in distinguishing mesothelial from epidermoid cyst as they are positive for calretinin and cytokeratin and negative for CEA and CA 19-9, whereas the epidermoid cysts are positive for CEA and CA 19-9 and negative for calretinin [1].

Surgery is the gold standard treatment for symptomatic and large (>5 cm) splenic cysts and the surgical procedure is tailored depending on the patient's age and the size, nature and location of the cyst. The various surgical procedures described are open complete splenectomy, partial splenectomy, cystectomy, marsupialization and cyst decapsulation [1,3]. Laparoscopic management of splenic cyst offers the advantage of minimally invasive surgery, faster recovery, shorter hospital stay and reduced morbidity. Anterior surface splenic cysts are more amenable to laparoscopic fenestration, than the posterior surface cysts as greater splenic mobility is needed in the latter. Open partial or complete splenectomy is advocated for centrally located splenic cysts [5].

CONCLUSION

In conclusion, we document a rare case of histologically proven large multiloculated mesothelial cyst of the spleen which was radiologically indistinguishable from a hydatid cyst. Ultrasound and CT findings can provide important clues to the diagnosis, but confirmation is only possible on histopathology. Thorough tissue sampling with mandatory multiple sections is advocated for documentation and profiling of the cellular lining to avoid potential pitfall of labeling a true splenic cyst as a false cyst.

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