Splenic Epithelial Cyst: A Rare Entity.
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Citation

Abstract
Congenital splenic cysts are also called epidermoid or epithelial cysts. They are uncommon, comprising only about 10% of benign nonparasitic cysts. Most of the cysts are asymptomatic, and they are incidental findings during abdominal ultrasonography. The number of diagnosed splenic cysts seems to rise because of the increased use of abdominal imaging techniques. Laparotomy with splenectomy has been the method of choice for the treatment of primary splenic cysts.

CASE HISTORY
A 28-year-old Hindu female came with complaints of pain in the left upper abdomen for two and a half months. The pain was dull aching in the left hypochondrium, and would radiate to the back. There was no diurnal variation in the severity of pain and the pain was unrelated to food or posture. She had undergone exploratory laparotomy for a cystic lesion in the upper abdomen seven years ago. But details of the procedure or diagnosis were not available. She had low-grade fever off and on, but did not have any fever with chills or rigors. She did not give a history of any significant medical illness in the past. She had been married for three years, but did not have any children.

On examination, she had pallor. She was normotensive and all vital parameters were stable. On abdominal examination, she had mild tenderness in the left hypochondriac region, but she did not have any palpable organomegaly. Her hemoglobin levels were low (7.9g% on admission) and her ESR was raised (115). The rest of the blood investigations were within normal limits.

Ultrasonography of the abdomen was suggestive of a 10.6 x 9.3cm, well-defined, encapsulated, mixed echogenic lesion in the anterior portion of the spleen (subdiaphragmatic). Multiple hypo-anechoic lesions were also seen in this lesion, the largest measuring 4.6cm. This was suggestive of splenic hydatid.

CT scan showed a lesion of 10 x 7.8 x 8cm in the spleen, with a large exophytic component - representing a splenic hydatid. Minimal left pleural effusion was also noted.

Echinococcus IgG antibody was negative.

The patient was taken up for exploratory laparotomy with splenectomy. The spleen was densely adherent to the stomach on its anterior and medial side. There were adhesions with the pancreas and also dense, inseperable adhesions with the diaphragm. There was a multiloculated
abscess with foul smelling pus. Pus culture did not show any growth of organisms.

**Figure 2**
Figure 2: Resected specimen of spleen showing the cyst cavity

The histopathology report of the specimen of spleen showed cysts lined by tubulocolumnar, pseudostratified (mesothelial line) epithelium. Focally, the lining epithelium was flattened and the cyst wall was composed of fibrocollagenous tissue, the cyst was filled with eosinophilic material and foamy and hemosiderin-laden macrophages. The largest cyst was devoid of any epithelium. The diagnosis on histopathology was epithelial (primary) splenic cyst.

The patient’s post-operative course was uneventful. The patient was started on Injection Benzathine Penicillin prophylaxis. She is following up regularly, without any complaints.

**DISCUSSION**
Cystic changes of the spleen are very rare. Based on the presence or absence of cellular lining of the cystic wall, splenic cysts are classified as primary (true) or secondary (pseudo) cysts [1,2]. Splenic cysts other than those of hydatid disease are also very uncommon. Most true splenic cysts are epithelial in origin and have embryonic inclusion of epithelial cells from adjacent structures [3]. Splenic cysts may be of parasitic or non-parasitic origin. Taenia echinococcus infection is the most common cause of parasitic cysts, occurring most frequently in regions endemic to that tapeworm.

Congenital splenic cysts are also called epidermoid or epithelial cysts. They are uncommon, comprising only about 10% of benign non-parasitic cysts. Splenic epithelial cysts occur predominantly in children and young women [4]. Small cysts are usually asymptomatic. The initial symptoms and signs referable to large cysts may include vague abdominal pain and a palpable mass in the left upper quadrant with or without symptoms due to compression of adjacent organs [5].

Histologically, epidermoid cysts have a squamous epithelial lining with intracellular bridges and a thick collagenous wall. The interior cyst wall may be composed of thick trabeculated fibrous bands covered by epithelium. The cystic fluid may contain cholesterol crystals, protein particles, or breakdown products of hemorrhage. The cellular lining of congenital cysts is thought to arise from infolding of peritoneal mesothelium following splenic capsule rupture or from mesothelial cells trapped in splenic sulci. The mesothelium undergoes metaplasia to squamous epithelium secondary to chronic irritation. Another postulation is that congenital cysts arise from normal lymph spaces in the spleen [6].

A comprehensive differential diagnosis for a cystic lesion of the spleen includes parasitic echinococcal disease, congenital cyst, intrasplenic pancreatic pseudocyst, pseudocysts from splenic trauma, infarction, infection, pyogenic splenic abscess, metastatic disease, and cystic lymphangioma/hemangioma (rare). Posttraumatic cysts are actually false cysts that typically have a smooth, fibrous non-cellular lining, can lead to hemorrhage, and may calcify. About half of the patients found to have this most common type of splenic cyst can recall a significant abdominal trauma.

Ultrasoanography is able to show that the cysts are either anechoic or hypoechoic and that they have a smooth thin wall [7], whereas solid tumors are either isoechoic or hypoechoic. In addition, computerized tomography and magnetic resonance imaging may give most of the necessary information, regarding the morphology of the cyst, the
composition of the cystic fluid, the location in the spleen, the position of the cyst and its relationship with the surrounding tissues [8]. Calcifications of both primary and secondary cysts are frequently found, which is useful in differentiating between cysts and other causes of splenomegaly.

Due to the increased risk of complications, splenic cysts with a diameter larger than 4-5cm should be managed surgically [9], because conservative options, such as percutaneous aspiration or sclerosis, do not result in long-term control. There are different types of surgical treatment according to the patient’s age and the size, location and nature of the cyst. The classical approach to splenic cysts has been open complete splenectomy.

Today the optimal treatment options are partial splenectomy, total cystectomy, marsupialization, or cyst decapsulation (unroofing), accessed either by open laparotomy or laparoscopy [9]. Partial splenectomy preserves more than 25% of splenic parenchyma, which is the minimal splenic tissue to preserve immunologic protection without increasing the risk of recurrence [10].

However, any type of conservative procedure is difficult to perform, if the cyst is very large, is located in the splenic hilum, or is covered completely by the splenic parenchyma (intrasplenic cyst), or if there are multiple cysts (polycystic cases): in these cases, a complete splenectomy should be performed either using the open or the laparoscopic approach.

References
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