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Case Report

A massive splenic cyst in a 13 year old girl: Rare entity

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Abstract

Splenic cysts are rarely encountered and mostly seen in paediatric age group. It can be either a true or pseudocyst in nature and epithelial lining is found in true cysts. We report a case of 13-year- old girl who presented with an abdominal swelling not associated with pain since 5 months. Ultrasonography of the abdomen showed a large cystic lesion in relation to pancreatic tail. CECT abdomen revealed an unilocular cyst arising from the spleen. Open splenectomy was performed. Histopathological examination revealed it to be a primary epithelial/epidermoid cyst of the spleen. The post-operative course was uneventful and she was asymptomatic at the 3 months follow-up.

Keywords: Epithelial cyst; Paediatric age; Splenic cyst.

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1. Introduction

An incidence rate of 0.07% of splenic epithelial cyst is found according to available literature.[1] Cysts can be of either parasitic or non- parasitic origin. True splenic cysts include parasitic cases and nonparasitic cysts. Parasitic cysts maybe due to Echinococcus granulosus. Nonparasitic cases, can be congenital, epithelial, vascular, and neoplastic.[2] Splenic cysts are difficult to diagnose clinically and definitive diagnosis of splenic epithelial cyst is possible only after splenectomy on histopathological examination.

2. Case Report

A 13 year old girl presented with mass per abdomen since 5 months. Not associated with pain, fever, anorexia or weight loss. No significant past history was elicited.

On clinical examination abdomen showed a mass in left hypochondrium region. The mass was soft and non-tender. Clinically, an initial diagnosis of massive splenomegaly was considered.

Serum CA19.9 levels >500U/ml noted. Serum amylase, lipase and CEA levels were normal.

Abdominopelvic ultrasonography showed a large cystic space occupying lesion in relation to pancreatic tail. CT contrast scan showed a large cystic lesion in epigastric to left hypochondrium measuring 12.5 x 11.2cm. Thin peripheral calcification noted [**Figure 1**]. Possibility of isolated Hydatid cyst to be ruled was mentioned.

The patient was taken for surgical exploration in view of the large mass. Intra-operatively a massive spleen which is cystic in nature was found and cyst fluid was aspirated. Analysis revealed hemorrhagic sample with neutrophils and lymphocytes. No evidence of malignant cells or parasites noted. Open total splenectomy was performed.

We received an enlarged spleen measuring 16x10x1.6cm in size and cystic in nature [**Figure 2**a]. Cut section showed an uniloculated cyst and exuded hemorrhagic fluid [**Figure 2**b]. Microscopy revealed a cyst lined by stratified squamous epithelium and wall shows foci of hemosiderin laden macrophages [**Figure 3**a,c]. Surrounding splenic parenchyma shows congestion and dilated blood vessels [**Figure 3**b]. No parasites were seen. No atypical cells or evidence of malignancy observed.

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Since the cyst was greater than 5cm in diameter, open splenectomy was performed. Conservative approach is preferred for smaller cysts to prevent post splenectomy complications like sepsis leading to increased mortality.

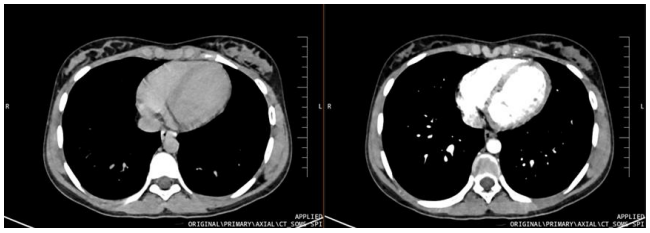

Figure 1: CT scan shows large splenic cystic lesion with thin peripheral calcification noted.

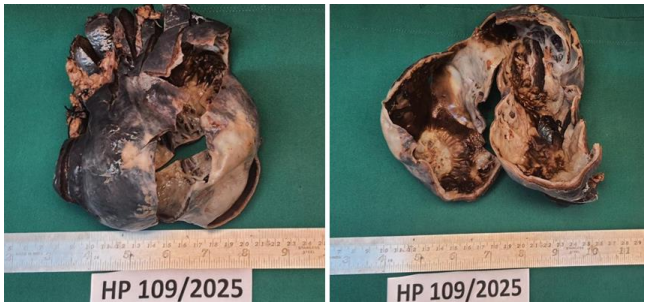

Figure 2: a: Excised cystic mass with an irregular external surface; **b:** Cut-surface of the mass shows uniloculated cyst with areas of hemorrhage.

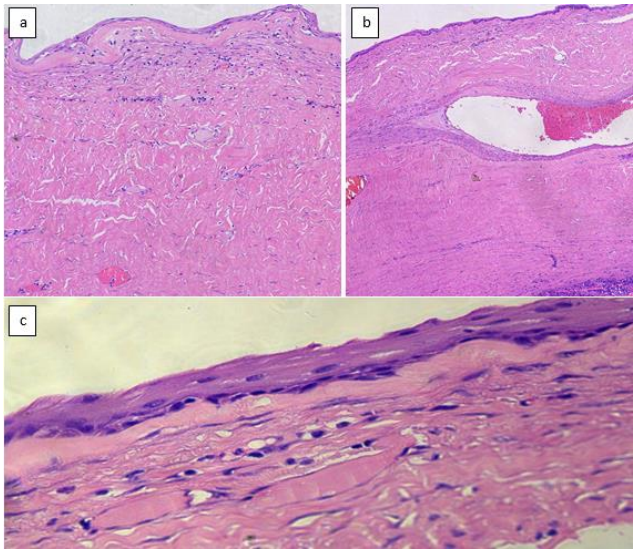

Figure 3: a: Splenic cyst with epithelial lining and fibrocollagenous wall; **b:** Residual parenchyma seen in wall; **c:** Stratified squamous lining.

3. Discussion

Splenic cysts are very rare. Splenic cysts are classified as primary or secondary cysts based on epithelial lining.[3,4] Embryonic inclusion of epithelial cells are seen in epithelial lined cysts from adjacent tissues.[5]

Epithelial lined cysts are usually seen in the second and third decades of life. Even pediatric age group can be affected.[6] Some cysts are asymptomatic and can present with abdominal distension. Splenic cysts can present with pain in left hypochondrium, mass effect and abdominal distension. The initial symptoms can be nausea, vomiting and dysphagia.[7]

On microscopy, primary splenic cysts have either squamous, flattened, cuboidal or low columnar epithelial lining. Stratification can be present and no atypia seen. Epidermoid cysts have stratified squamous epithelium with a fibrocollagenous cyst wall. The cystic fluid may contain cholesterol crystals, protein particles or breakdown products of hemorrhage. The peritoneal mesothelium infolding due to splenic capsule rupture or from entrapped mesothelial cells in the splenic sulci cause the epithelial lining of congenital cysts.[8]

On physical examination an abdominal mass is usually elicited. Routine laboratory investigations are normal. Tumor markers, carcinoembryonic antigen (CEA) and CA19-9, may be elevated.[9]

The differential diagnoses for splenic cysts include infection, parasitic echinococcal infestation, congenital cyst, pseudocyst, infarction, splenic abscess, lymphangioma, metastatic disease, and hemangioma.

Excision is recommended for a symptomatic splenic cyst and for cysts larger than 5 cm. Marsupialization or fenestration as treatments are indicated for superficial cysts, but they have a high recurrence rate.10 In our case since the cyst was greater than 5 cm, total splenectomy was our indicated treatment of choice.

Splenic epithelial cyst is a rare entity and usually diagnosed histopathologically. Radiological differentiation is difficult. Total splenectomy is preferred choice of treatment and preventative intervention against future rupture, hemorrhage, infection, or recurrence.

4. Source of Funding

None.

5. Conflict of Interest

None.

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