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Benign Epithelial Cyst of Spleen: A Case Report with Review of Literature

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Abstract: Splenic epithelial cysts are rare. We present such a case in a 14 year old boy who underwent splenectomy. The cyst had multiple septations and satellite lesions prompting a provisional diagnosis of a parasitic cyst on radiology. The diagnosis of epithelial cyst was made on histopathological examination.
Keywords: Splenic epithelial cyst, IHC, non parasitic cyst.

INTRODUCTION

Splenic cysts are rare with an incidence of 0.07 % [1]. Parasitic cysts caused by Echinococcus granulosus are common [2]. Epithelial cysts comprise a mere 10% of non parasitic cysts. Therefore we present this case ofan epithelial cyst of spleen, its diagnostic aspects and role of IHC along with review of literature.

CASE REPORT

A 15 year old boy presented to our hospital with complaints of left upper abdominal pain since 1 year and intermittent fever for past 3 weeks. The onset of pain was insidious, dull aching type on and off for past 1 year radiating to left shoulder. This pain was aggravated by running, coughing, sneezing and relieved by rest. There was no nausea, vomiting, altered bowel habits or renal symptoms. There was a prior history of trauma.On examination patient was under nourished with pallor, clubbing and koilonychia. His pulse rate was 102/ minute, BP 100/70 mmHg and temperature was 100^0 F at the time of admission. Per abdomen there was massive splenomegaly, liver was not palpableand there was no evidence of ascitis. On complete haemogram Hb was 8.6gm%, WBC count was 12,500/cu mm with neutrophilic leucocytosis, PCV 25 Vol%, MCV 70 fl, MCH 25.2 pg, MCHC 32.2%. Platelet count was 2.8 lakh/ cumm.

USG abdomen revealed a large hypoechoic lesion in the spleen measuring 15×12 cm with areas of necrotic material within the mass suggestive of splenic cyst. CECT abdomen showed a large cystic lesion 11.4×11.7 cm with multiple satellite cysts, compressing greater curvature of stomach and inferiorly pushing the kidney. Differential diagnoses of splenic abscess or hydatid cyst were offered. Massive splenomegaly with multiple adhesions to stomach, colon, diaphragm and a large cyst occupying the superior pole of the spleen was seen peroperatively. Complete splenectomy was done.

Gross examination showed a large cyst measuring 12×14cmwithseptations and satellite cysts filled with gelatinous to haemorrhagic fluid. On histopathological examination a cyst wall with dense inflammatory infiltrate lined by flattened to cuboidal epithelium was seen. Cyst lumen showed eosinophilic to haemorrhagic fluid interspersed with cyst macrophages. Differential diagnosis of epithelial cyst or endothelial cyst was considered. On immunohistochemistry CD34 was negative, Pan cytokeratin was positive. A final diagnosis of benign epithelial splenic cyst was given.

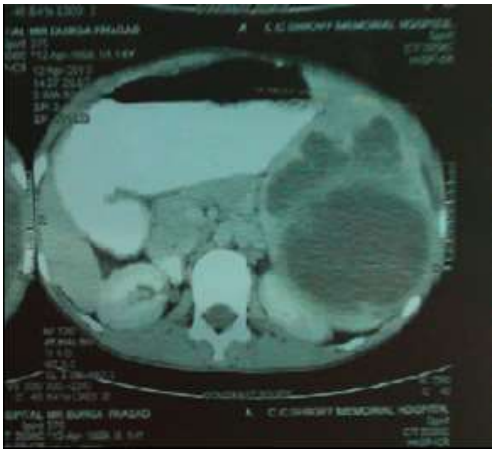

Fig. 1: Spleen with large cyst

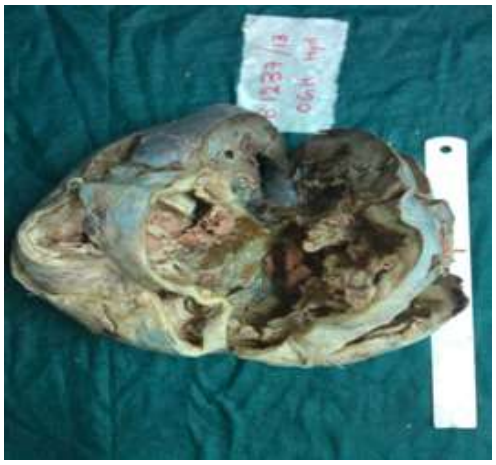

Fig. 2: CT of abdomen showing large splenic cyst with satellite cysts compressing left kidney

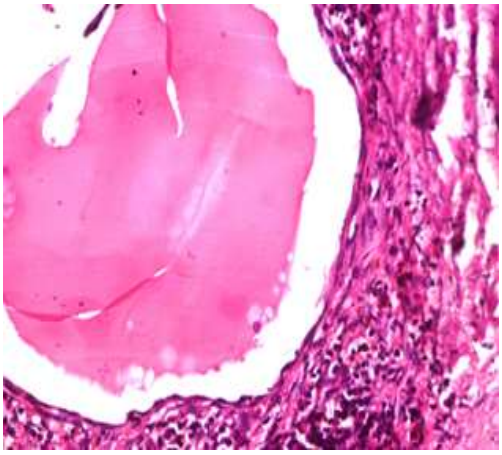

Fig. 3: Cyst wall lined by flattened epithelium

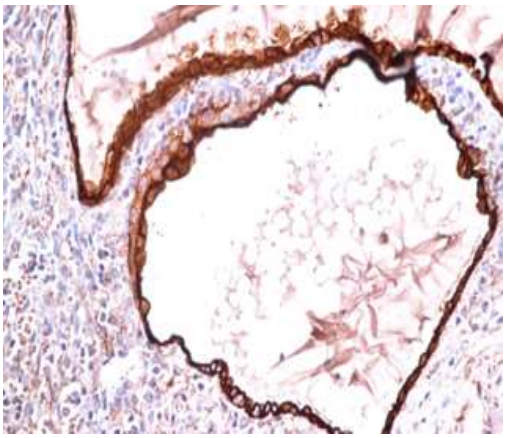

Fig. 4: IHC: Positive for Pan cytokeratin

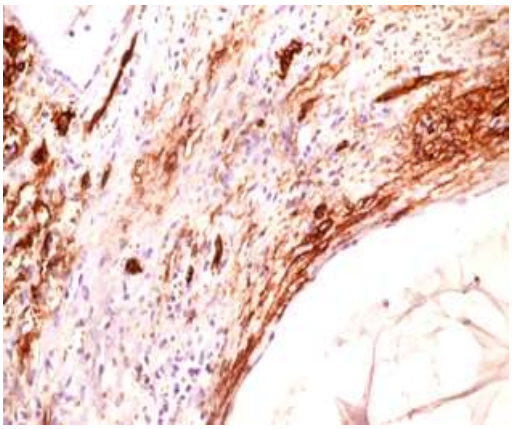

Fig. 5: IHC: Lining epithelium negative for CD 34

DISCUSSION

Splenic cysts with an incidence of 0.07% were proposed by Robbin's FG *et al*. [1] in 1953 by reviewing about 42,327 autopsies. Most of the splenic cysts are asymptomatic and may be incidental findings during abdominal ultrasound examination. Diagnosis of splenic cysts may be on rise due to increased use of imaging techniques. Ultrasonography and CT scan usually gives information regarding the cyst, its location and to a certain extent the nature of the cyst fluid. Histopathological examination confirms the diagnosis of true or false cyst.

Splenic cysts according to Fowler and later modified by Martin were classified into primary and secondary cysts. Primary cysts were further sub classified into parasitic and non parasitic cysts based on the etiology [2, 3]. Splenic cysts can also be classified into true cysts or false cysts (pseudocysts) based on the presence or absence of lining epithelium respectively. Parasitic splenic cysts are frequently caused by Echinococcus granulosus. False or pseudocysts which comprise 75% of non parasitic cysts are caused by trauma, inflammation or degenerative changes.

True cysts are rare of which the epithelial cysts comprise only 10%. A typical true cyst lining is positive for keratin (epithelium) and negative for factor VIII (endothelium). Only cysts with an epithelial lining are designated as true cysts. These true cysts can be further designated based on the type of lining epithelium as epidermoid (stratified non keratinizing squamous), dermoid (squamous lining with appendages), mesothelial (cuboidal or low columnar epithelium). Mesothelial cysts are positive for cytokeratin and calretenin. Lymphangiomas on the other hand have an endothelial lining and are positive for endothelial markers.

There are several theories to propose the pathogenesis of epithelial splenic cysts. They could arise due to displacement of epithelial tissue during development or can arise from the invagination of

peritoneal mesothelium fromthe capsule or cells trapped in the splenic sulci [4, 5]. The epithelial tissue can occur as embryonal inclusions of adjacent structures like gonads, dorsal mesogastrium or wolffian duct [6-8].

Epithelial splenic cysts are usually common in second to third decades of life. Two thirds of patients are females [9, 10] but there are studies where higher frequencies in males have been reported [11]. Symptoms of splenic cyst depend on its size and location and begin only after attaining a significant size. They may cause pain, heaviness, dragging sensation or present as mass per abdomen. Compression on adjacent structures might cause nausea, vomiting and irritation of left diagphragm. Patient may also present with complications like inflammation, intracystic hemorrhage or peritonitis. Inflammation is commonly due to bacteria of Salmonella group [12]. Spontaneous rupture with trivial injury can result in fatal intraperitoneal haemorrhage [13].

Treatement of splenic cyst include aspiration, decapsulation, partial splenectomy or total splenectomy. Brown *et al*. [14] reported that a total splenectomy was required in 1 out of 7 patients where cyst was large especially involving the hilum.

However a conservative surgery like partial splenectomy, cystectomy, marsupialisation or splenic decapsulation is now an option keeping in view the possibility of life threatening infections after splenectomy .These procedures however risk a chance of adhesions thereby further resulting in splenectomy.

CONCLUSION

A case of benign epithelial cyst is presented. Preoperative diagnosis of parasitic cyst was considered which proved to be epithelial cyst on histopathology and IHC. Though the sex ratiofavors females our patient is a male. History of trauma should always be enquired to rule out secondary causes. Patient is under regular follow up and is doing well.

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