Case Report

Unusual Cause of Splenomegaly: Splenic Cyst

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ABSTRACT

Cysts in the spleen are rare, however the true incidence is not documented. There are very few cases reported in the literature. The diagnosis of splenic cyst is incidental, since majority of patients remain asymptomatic. If symptomatic, they present with a palpable mass in the left upper quadrant. The diagnosis of splenic cysts have increased due to the advancement of computed tomography(CT) and the various management options. We discuss two cases of splenic cysts as they are rare and the diagnosis and management has always been a challenge to a pathologist and the treating surgeon.

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Introduction

Epithelial cysts of the spleen are the most commonly seen primary splenic cysts in clinical practice. They constitute 10-15% of benign non parasitic cysts[1]. A palpable mass in the left upper quadrant of the abdomen is the most common presentation[2]. We report two cases, a 15 year old female who presented with mass per abdomen. Computed Tomography(CT) scan revealed the possibility of a splenic cyst. Histopathology confirmed the diagnosis of primary epithelial splenic cyst. A 35 year old female presented with recurrent pain in the abdomen. Diagnosis of haemorrhagic splenic cyst was made on CT scan. Histopathology confirmed the diagnosis of a pseudo cyst of the spleen.

Case Series

CASE 1: A 15 year old female presented with a mass in the upper abdomen with pain and epigastric discomfort. On examination, a large globular mass was palpable in the epigastrium measuring 20cm x 18cm extending to the left hypochondrium. Haematological (Hb-11.8g%, TLC – 9500c/cmm, Platelets – 2,85,000c/cmm) and biochemical(Serum Creatinine–0.56mg/dl, Urea – 2.7mmol/L, Liver function test, and electrolytes) parameters were normal. Ultrasonography(USG) and CT scan of the abdomen revealed a large well encapsulated fluid filled splenic cyst measuring 20x15cmin the left upper quadrant. The cyst was thick walled and was not delineated from the spleen and pancreas.The diagnosis of splenic cyst was made. Laparoscopic Marsupialization was performed. The membranous splenic cyst mass was sent for histopathology. Microscopy showed a cyst wall withfibro-collagenous tissue lined by low cuboidal epithelium and centrally located benign nuclei [Figure 1A]. Numerous gamna-gandy bodies were present [Figure 1B]. The diagnosis of primary epithelial cyst of the spleen and chronic venous congestion was made. Immunohistochemistry profile was not done. The patient was discharged without any complications.

CASE 2: A 35 year old female, presented with recurrent pain in the left upper abdomen. On examination, a globular mass was palpable in the hypogastrium. Haematological (Hb–12.4g%, TLC-7500 c/cumm, Platelets – 4,10,000c/cumm and biochemical(Serum creatinine-0.74mg/dl, Urea-2.96mmol/L, Liver function test and Electrolytes) parameters were normal. CT scan of the abdomen revealed a large fluid attenuated cystic lesion arising from the spleen measuring 16.3cm x 10.8cm x 17.5cm. It showed multiple thin septations and peripheral punctate calcification [Figure 2A]. Diagnosis of haemorrhagic splenic cyst was made and splenectomy was performed. The spleen was sent for histopathology. Grossly, the spleen was enlarged measuring 18cm x 6cm x 4cm.Outer surface appeared congested and cystic in consistency. Cut surface exuded haemorrhagic serous fluid. Solid and cystic areas were noted. Some areas showed honey comb appearance [Figure 2B].Surrounding splenic tissue appeared normal and no calcification was seen. Microscopically, cyst wall was lined by fibrous tissue with areas of calcification. Some areas showed necrotic tissue with haemorrhage [Figure 2C]. The diagnosis of pseudo cyst of the spleen was made. The patient was discharged without any complications.

Discussion

Splenic cysts are not frequently encountered in everyday surgical practise[1]. The cysts can be due to benign disease like infections, malignancy and metastasis[2].Clinically,most of the cysts are asymptomatic and diagnosed incidentally[2,3]. They present as a palpable mass in the left upper quadrant of the abdomen[4]. When they are oversized(5cm),they can manifest with symptoms which cause atypical pain and heaviness in the left hypochondrium. The other non-specific symptoms are nausea, vomiting, diarrhoea and flatulence. If the spleen is oversized they cause pressure

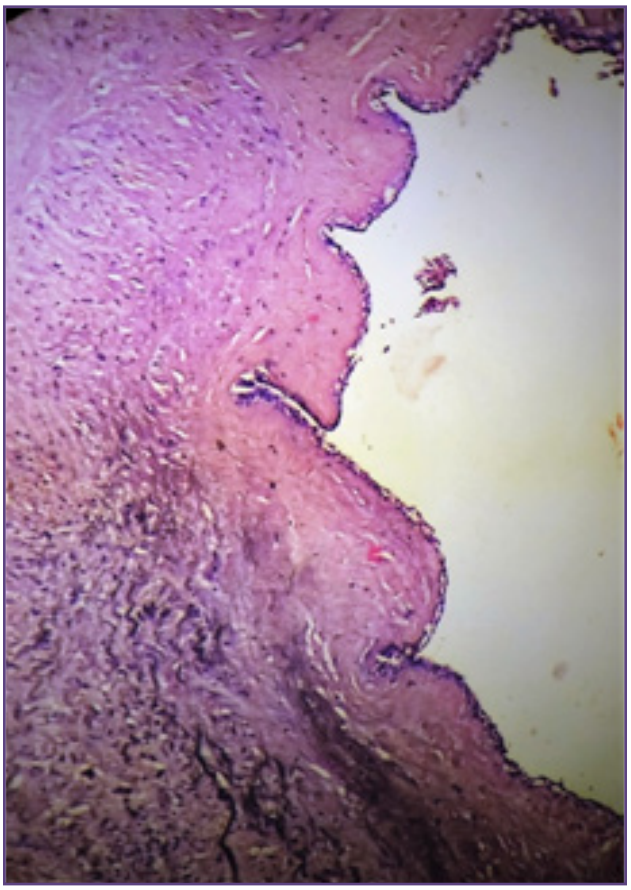

Fig. 1A:H & E stained section. High power view showing features of primary epithelial cyst, lined by low cuboidal epithelium.

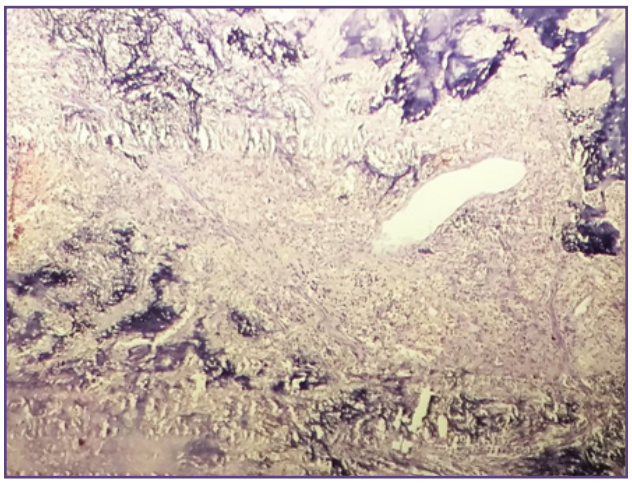

Fig. 1B:H &E stained section. High power view showing presence of Gamna gandy body indicating chronic venous congestion of spleen.

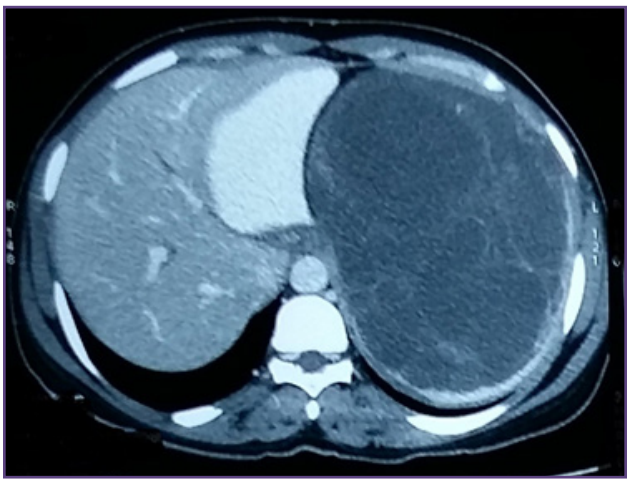

Fig. 2A: CT Scan of the abdomen showing enlarged spleen with cystic changes, multiple septations and peripheral rim of calcification.

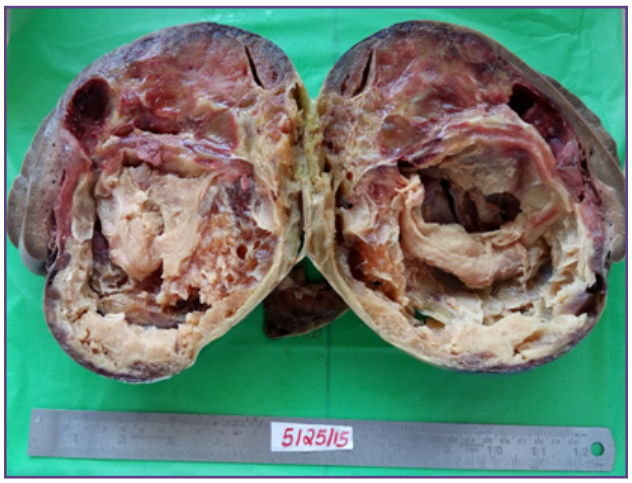

Fig. 2B: Cut surface of the enlarged spleen showing cystic areas with focal areas showing honey comb appearance.

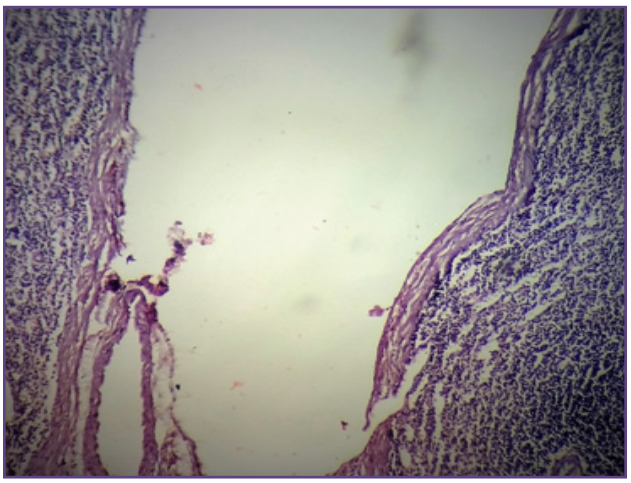

Fig. 2C: H &E stained section.High power view showing features of pseudo cyst, lined by fibro-collagenous tissue and absence of lining epithelium

symptoms on the secondary organs causing chest pain and dyspnoea[2,3]. The cases reported, presented with a palpable mass in the upper quadrant of the abdomen with no secondary symptoms. The findingscorrelate with literature. It is essential to exclude other causes of splenomegaly which include infections like schistosomiasis, infectious mononucleosis, chronic liver disease, vascular disorders, metabolic disorders like Gaucher's disease and Niemann-Pick disease, benign and malignant infiltrations like lymphoma and leukaemia[2].

Splenic cysts can be classified as primary and secondary cysts. Primary cysts are true cysts, most commonly seen in the first decade. They can be sporadic or familial[3]. They are asymptomatic. On USG, they are well demarcated and anechoic cystic mass[2,3]. CT scan shows a low density cystic mass with an ill-defined wall. Magnetic resonance imaging shows a typical cystic lesion with hyper-intense signalling[2]. Microscopically they show the presence of a lining epithelium which can be columnar, cuboidal or stratified epithelium. The first case presented with the similar manifestations as described above. Primary cysts can be classified as parasitic and non-parasitic. Non-Parasitic cysts can further be classified as congenital, neoplastic, degenerative and traumatic[3,4].

Secondary splenic cysts are pseudocysts, lacking an epithelium and replaced by thick fibrous wall which is often calcified[2,3]. They are commonly seen following inflammation, infarction and thrombosis. On USG, peripheral echogenicity with distal shading and calcification of the wall is noted. CT scan shows a well demarcated

homogenous non contrasting cystic mass[2]. The second case presented similar to the findings mentioned above.

CA 19-9 and carcinoembryonic antigen levels are increased in primary epithelial cysts. Serum levels of CA 19-9 have been shown to reduce after cyst removal, offering a screening test to indicate recurrences in case of spleen preserving surgeries. These levels were not estimated in our case since histopathology confirmed the diagnosis of primary epithelial cyst with cuboidal lining epithelium[5].

With the advancement of USG,CT scan and successful management options available, various techniques are adopted for management of splenic cyst[7]. They are aspiration of cysts by injecting tetracycline and alcohol into the cyst wall for destroying the cyst lining[4].Resection of the cyst and a portion of the parenchyma is another treatment option available but highly unsuccessful[6].

Marsupialization is creating an opening into the cyst wall for drainage. It can be done internally into the peritoneal cavity or externally through a purposefully created cysto-cutaneous fistula. The first case report was managed by the above method. But the chances of recurrence is very high. The other formof marsupialization is a near total splenic resection. It is simple, rapidly performed procedure. The chances of cyst recurrence is high because a portion of cyst lining is left intact[8].

Splenectomy is the most commonly performed surgical technique. The first attempt was performed by Jules Pean. Due to the awareness of the immunological functions of the spleen, spleen sparing surgeries are being adopted[3,8].

Conclusion

There are numerous causes of splenomegaly. It is important to consider splenic cyst as a differential diagnosis, since the patient is asymptomatic.Differentiation of splenic cysts should be made preoperatively. Preservation of the spleen is important to preserve the functions of the organ. The two cases discussed above highlight the features which help differentiate true and pseudo cysts and the role of histopathology in the diagnosis and the various management strategies available.

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