AN EPITHELIAL CYST OF Spleen- A RARE CLINICAL ENTITY: A CASE REPORT

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INTRODUCTION
Primary splenic cysts are very rare clinical entity and most of the times an incidental clinical finding. Primary cysts have a well defined epithelium which is usually squamous epithelium. According to the literature first case of such primary cyst was reported in 1829 by Andral. These cysts can be classified as: Primary cyst and Secondary cyst. Primary cysts are classified as parasitic and non-parasitic. Non parasitic cysts are classified as congenital and neoplastic. Parasitic cysts are usually formed by infection with Echinococcus species (E. Granulosus or E. Multilocularis). Secondary cysts are usually post traumatic cysts and are also known as pseudo cyst as there is no lining epithelium. Most of these cysts are asymptomatic and diagnosed incidentally during imaging. Symptoms usually depend on size of the cyst. The larger the cyst more the symptoms probably because of stretch on the capsule or compression on surrounding structures. The most common symptom is left sided abdominal pain. The diagnostic modalities available is Ultrasound and CT scan. Treatment of the cyst depends on the size of cyst. According to the consensus cyst of >5cm should be operated.

CASE PRESENTATION
Clinical feature:
A 45year old female, married, housewife with no comorbidities, no significant previous surgical history, presented with c/o pain in left upper abdomen since last 3 days which was sudden in onset, dull aching, non-radiating, associated with nausea and two episodes of vomiting. On examination patient had left hypochondrial tenderness. On palpation spleen was enlarged and was palpable 6 cm below costal margin.

Investigation:
Blood profile:
- RBC 4.67 M/μL
- TLC 5100/μL
- Platelet ↓ed 1,12,000/μL
- Neutrophil ↑ed 81.50%
- Lymphocyte ↓ed 11.80%
- Eosinophil ↓ed 0.8%

Radiological Imaging:
Patient underwent Ultrasound imaging which showed large multiloculated cystic lesion in left hypochondrial region with splenic size of 15x9 cm. Patient underwent further investigation CT scan which showed moderate splenomegaly with large cystic lesion with mild wall enhancement and multiple thick septa measuring 10.2x13.3x15.2 cm (TR x AP x CC). (Figure.1) (Figure.2)

Histopathology:
Gross: splenectomy specimen 14*9*5 cm weighing 411 gms with external fibrinous area and cut surface shows multiple septations. (Figure.4)

Microscopic: cyst lined by cuboidal epithelium with thick capsule and fibrous septa. Cyst macrophage and mixed inflammatory infiltrate. (Figure.5)

Impression: Epithelial Cyst of Spleen

KEYWORDS
Epithelial cyst, Splenic cyst, Laparoscopic Splenectomy
Splenic cysts are rare in nature. Fowler and Martin [1][2] classified splenic cyst into Primary and Secondary according to the presence or absence of epithelial lining. Primary cysts are further classified into Parastatic and non-parastastic cysts. Non-parastatic cysts are again divided into congenital and neoplastic. Primary cysts are also known true cyst and secondary cysts are also known as pseudo (false) cysts. Parastatic cysts are almost always due to Echinococcus and are found twice as frequently as non-parastatic cyst. Primary splenic cyst constitutes 10% of all non-parasitic cysts. Parasitic cysts develop because of infection with echinococcus species infection. [3]

**Histologically**, this type of congenital cyst contains squamous epithelium but may also contain other type of epithelium like cuboidal as shown in our case. Theory of cyst development:

1. Mesothelial invagination theory – It is postulated that during development, because of invasion of mesothelial lining along with the capsule these cysts develop. As these linings are pluripotent in nature, they have propensity to undergo metaplasia and secret fluid leading to form the cysts.[4]
2. Lymph space theory- As per this theory the splenic cysts may arise from the normal lymph spaces present in the spleen.[5]
3. Endodermal inclusion theory- This theory proposes that epithelial splenic cysts develop by true metaplasia of a heterotopic endodermal inclusion within the spleen. As the mesothelium is pluripotent in nature, metaplasia in this lining may be the main causative factor in formation of these cyst with different types of epithelial linings like, squamous, columnar etc. [6][7]

**Incidence:**
Splenic epithelial cyst most commonly occur in the second or third decade of the life. [8] These cysts are also common in females and below 40 years. But it may present later in life in fourth or fifth decade (as in our case where the age of the patient is 47 year) and also may occur in children or infants.

**Symptomatology:**
Most of the times these cysts are asymptomatic in nature and identified on radiological investigation done for other complaints. But sometimes it may present with vague symptoms like mild left sided upper abdominal pain with physical findings suggestive of splenomegaly. Initial symptoms include vague abdominal pain, early satiety, nausea and vomiting.

**Investigation:**
This type of cyst can be diagnosed using radiological imaging like ultrasound and CT scan. Splenic cyst on ultrasound are usually hypoechoic or sometimes anechoic well defined intrasplenic lesion. [5] On contrast-enhanced CT, congenital cysts can be unilocular or multilocular and can be found anywhere within the spleen. They appear as well-defined, thin-walled, spherical lesions with homogeneous content. [9]

Few blood investigations should also be done such as complete blood count to see splenic function, and tumor markers like carcinoembryonic antigen (CEA) and CA-19-9 which are sometimes elevated in patients with splenic cyst. [10]

**Treatment:**
Treatment of the splenic cysts depends on the size of the cyst. Most of the times cyst size of 4-5 cm is managed conservatively but larger than this should be managed surgically because of increased risk of complication like spontaneous rupture.[11][12] At present, options available are percutaneous aspiration or percutaneous drainage, partial splenectomy, deroofing, total cystectomy, marsupialisation, total splenectomy.[13][14] Splenectomy can be performed both open and laparoscopically. But laparoscopy being minimally invasive is a better technique because of lest post-operative pain, shorter hospital stay, and improved cosmetic results. In our case also we did laparoscopic splenectomy.[15] Now a days because of risk of overwhelming post splenectomy infection, vaccination against common organisms like Pneumococcal, Meningococcal and Haemophilus Influenzae b. In case of planned splenectomy vaccination is given 2 week prior to operation and in case of emergency splenectomy it is given 2 weeks post splenectomy.[16][17] In our case as the cyst was huge in size decision was taken to do total splenectomy laparoscopically and was successfully performed with uneventful recovery till present follow up.

**REFERENCES**