**Case Report**

**A rare gigantic primary epithelial cyst of spleen in an adolescent boy**

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**ABSTRACT**

Cysts arising from spleen are a rare clinical entity, with their discovery usually being incidental. We present a case of the largest reported primary epithelial cyst of spleen. In present study a 16-year-old child presented with abdominal distension for 1 month associated with abdominal pain for 2 weeks. On examination, a mass was felt in the epigastrium, left hypochondrium and left flank, extending across from the right midclavicular line to the left midaxillary line, and extended inferiorly till 6cm caudal to umbilicus. CT abdomen revealed a unilocular cyst of 17x20x24cm arising from the spleen with a volume of 3700ml. Splenectomy was performed; intra-operatively there was a huge splenic cyst measuring 30x30x30cm. Histopathological examination was diagnostic of Primary congenital epithelial cyst of the spleen. Till date the largest congenital splenic cyst documented in literature is 20 cm × 13 cm × 21 cm, as reported by Valentina et al, in 2014. Present specimen bests this previous case considerably.

**Keywords:** Largest congenital epithelial cyst, Splenectomy, Splenic cyst

**INTRODUCTION**

Splenic cysts are generally classified as primary and secondary(pseudocyst).1-3 Primary cysts have an epithelial lining and can be nonparasitic or parasitic (echinococcal). Worldwide, Echinococcus infection is the most common cause of a splenic cyst. Non-parasitic cysts include simple cysts, epidermoid cysts, and dermoid cysts and neoplastic.4 There have been reported in literature cases of primary splenic cystadenocarcinomas and of splenic lymphoma presenting as a splenic cyst.5,6 Isolated splenic metastasis is diagnosed in only 5.2% of case on autopic studies.7,8 They are mostly secondary to melanoma and cancer of the breast, lung, ovary, colon, stomach and pancreas.9,10 Although they usually appear as solid lesions, haemorrhagic phenomena, cystic or necrotic degenerations can occur, conferring to the metastasis a cystic feature.11-14 Moreover a cystic adenocarcinoma of the pancreatic tail, extended within the splenic parenchyma should be excluded.2,14 Various classifications have been proposed for splenic cyst based on whether they are lined with mesothelial, transitional, or epidermoid linings and also whether they are neoplastic, or secondary/pseudocyst (Figure 1).15

![Figure 1: Classification of splenic cysts.](image-url)
Primary epithelial congenital cysts are lined by flattened or cuboidal cells originating from in folding of peritoneal mesothelioma during splenic development. These lesions are usually small and asymptomatic and do not require excision. When these cysts are large and symptomatic, they can be removed by laparoscopic or open total or partial splenectomy.  

CASE REPORT

A 16-year-old male presented with abdominal distension for 1 month associated with abdominal pain for 2 weeks. Examination revealed a mass in the epigastrium, left hypochondrium and left flank which moved well with respiration. The swelling extended across from the right midclavicular line to the left midaxillary line, and extended inferiorly till 6cm caudal to umbilicus. Abdominal sonogram shows massive splenomegaly (16 cm size) with a large hypoechoic collection with internal echoes in the peri- pancreatic region extending upto spleen measuring 17x16x17.5cm, with volume of 2533ml (Figure 2).

Computed tomography of abdomen revealed a large, thin walled, unilocular cyst measuring 17.11x20.04x24.5 (APxMLxCC) dimension; a) 20.04x24.5cm in MLxCC (mediolateral & craniocaudal) dimension; b) 17.11cm in AP (Antero-posterior) dimension. No fluid-fluid level or debris were noted within the cyst. No daughter cysts were noted within the cyst (Figure 3 A, B).

A differential diagnosis of primary splenic cyst vs echinococcal cyst was considered the patient was prepared for surgery. Serology for E. granulosus was negative. After pre-treatment with meningococcal, pneumococcal and haemophilus influenza vaccines, the patient underwent surgery. A huge splenic cyst measuring 30x30x26cm was present with thinned out walls. Minimal residual splenic tissue was present at the periphery of the cyst. Multiple collateral vessels were present. Stomach and pancreas were displaced to the right side of midline. (Figure 4 A-D).

Figure 2: Abdominal sonogram shows massive splenomegaly (16 cm size) with a large hypoechoic collection with internal echoes in the peri- pancreatic region extending upto spleen measuring 17x16x17.5cm with volume of 2533ml.

Figure 3: Computed tomography of abdomen revealed a large, thin walled, unilocular cyst measuring; a) 20.04x24.5cm in MLxCC (mediolateral & craniocaudal) dimension; b) 17.11cm in AP (Antero-posterior) dimension.

Figure 4 (A-D): Huge splenic cyst measuring 30X30X26CM with thinned out walls. minimal residual splenic tissue was present at the periphery of the cyst.
Splenectomy was performed and the final specimen weighed 5.2 kg. Histopathology of the specimen shows splenic tissue with portions of cyst wall lined by low cuboidal to columnar epithelium showing focal stratification. The cyst wall showed hyalinized connective tissue (Figure 5.1-2).

![Figure 5: a) A-Inner aspect of cyst, B-Epithelial lining of cyst which is low cuboidal to columnar epithelium, C-Fibroconnective tissue of cyst wall, D-Normal splenic tissue, E-splenic capsule; b) In high power.](image)

All above mentioned features were diagnostic of Primary Congenital Epithelial cyst of Spleen.

**DISCUSSION**

Splenic cysts are relatively rare entities, with an incidence of 0.07% as reported in a review of 42327 autopsies. The differential diagnosis for these lesions includes parasite infections (which is the commonest), results of previous trauma or infarction, congenital forms, primary splenic neoplasm or cystic metastasis etc. Non-parasitic cysts of the spleen are relatively uncommon. These include dermoid, epidermoid and epithelial cysts. Primary true cysts of the spleen account for approximately 10% of all nonparasitic splenic cysts. These cysts, however, are benign and apparently do not have malignant potential beyond that of the surrounding native tissue.

True splenic cysts are often asymptomatic and discovered incidentally. Patients may complain of abdominal fullness, early satiety, pleuritic chest pain, shortness of breath, and/or left shoulder or back pain. They may also experience renal symptoms from compression of the left kidney. On physical examination, an abdominal mass may be palpable. Rarely, splenic cysts present with acute symptoms related to rupture, haemorrhage, or infection. Diagnosis is best made by CT. The treatment of nonparasitic cysts depends on whether or not they produce symptoms. Asymptomatic nonparasitic cysts may be observed with close follow-up by ultrasound to exclude significant expansion. Patients should be advised of the risk of cyst rupture with even minor abdominal trauma if they elect for nonoperative management for large cysts. Small symptomatic nonparasitic cysts may be excised with splenic preservation, and large symptomatic nonparasitic cysts may be unreftive or splenectomy may be performed. These may be removed through either laparoscopy or laparotomy.

Till date the largest congenital splenic cyst documented in literature was 20 cm × 13 cm × 21 cm, as reported by Valentina Pastore, et al, from the University of Foggia, Italy, in the year 2014. Our specimen bests this previous case considerably.

**CONCLUSION**

Splenic cyst is an uncommon clinical entity; and specifically non-parasitic splenic cyst is rare. Large symptomatic cysts necessitate intervention and histopathological confirmation of the same.

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