

An Uncommon Cystic Mass of the Spleen in a Child

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Abstract

Splenic cysts are extremely rare, particularly in pediatric population. They may be parasitic or non-parasitic in origin. The parasitic splenic cysts, usually caused by *Echinococcus granulosus*, account for 60% of all primary splenic cysts. The primary epithelial cysts or epidermoid cysts account for 10% of all splenic non-parasitic cysts. We report a case of epidermoid cyst of spleen in a 13-years-old boy admitted with an abdominal pain, which on imaging found to be a cystic mass arising from spleen. On laparotomy the spleen was found occupying a major part of the abdomen. Open partial splenectomy was performed. The post-operative clinical course was satisfactory and uneventful. Histopathological examination revealed it to be a primary epithelial cyst of spleen. This case report emphasises on the rarity of the case at this age.

Keywords: Primary epithelial cyst; Spleen; Non parasitic cyst; Partial splenectomy

1. Introduction

Cysts of the spleen in childhood are rare compared to the incidence of such masses in other parenchymatous organs [1]. They may require total or partial splenectomy for either diagnosis or treatment.

Epidermoid splenic cysts are unusual and often an incidental finding in surgical practice [2]. Although their prevalence is increasing due to the widespread use of abdominal imaging and non-operative treatment for splenic trauma. Splenomegaly is the commonest of findings. Upper left quadrant discomfort, pain or tenderness may also be present. Histological examination following surgical removal seems to be the most accurate technique to establish diagnosis [3].

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2. Clinical Case

Boy, 13 years of age, reporting intermittent abdominal pain in the left upper quadrant since the last two months; normal bowel movements and no other symptoms. No personal history of interest. On physical examination a firm, hemotender mass was palpable in left hypochondrium. Laboratory exam found infection signs. Abdominal ultrasound pointed out splenomegaly of 11 cm with a non-specific focal lesion in the upper margin of 5.6 cm × 6.5 cm, not very vascularized. Abdominal scan imaging showed a splenic multicyst focal lesions of the upper pole, bulging out the surface of the spleen which may be related to a lymphangioma cyst. An open partial splenectomy was performed using an above umbilical approach (because of the large size of the cyst). Fortunately, no post-operative incident occurred. Furthermore, antibiotic prophylaxis was administered for 15 days after intervention with pneumococcal vaccination. Regarding Anatomical pathology spleen parenchyma was largely replaced by many cystic cavities filled with a gelatinous material. The lesion was characterized by a squamous epithelium made of bland cells. No atypia or mitotic figures were observed.

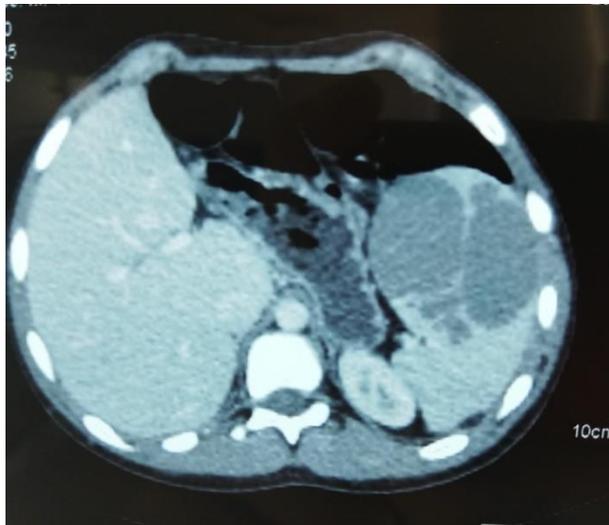


FIG. 1. Axial scan imaging of the child abdomen, showing splenomegaly with a polylobulated well-defined cystic mass.



FIG. 2. Intraoperative image of spleen with epidermoid cyst.



FIG. 3. Image of removed splenic cyst.



FIG. 4. Section of the spleen showed many cystic cavities filled with a gelatinous material. No solid areas were observed.

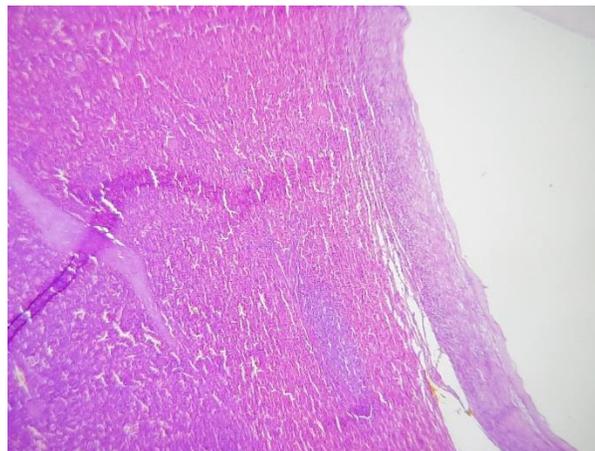


FIG. 5. Microphotography showing that the squamous epithelium is made of bland cells. No atypia or mitotic figures were observed. HE, 400X.

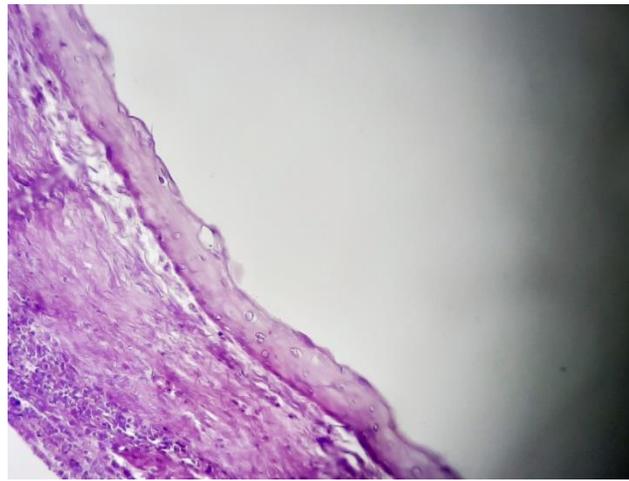


FIG. 6. Microphotography showing that the cystic cavity was layered by a squamous epithelium. HE, 100X.

3. Discussion

Cysts in spleen are unusual in adults and even rarer in children [4]. They have been classified by Martin and Fowler [5]. Splenic cysts may be of parasitic or non-parasitic origin. The epithelial cysts are the most common type of non-parasitic splenic cyst. Their global incidence is 0.007 with a female predominance. The pathogenesis of primary splenic cysts is not clear [4]. Primary cysts are also called true, congenital, epidermoid or epithelial cysts. As per the existing literature, since the first case was reported in 1929 by Andral, the classification of these lesions has evolved into the present system [2]. Most true splenic cysts are epithelial in origin and have embryonic inclusion of epithelial cells from adjacent structures [6].

Small epidermoid cysts in childhood are usually asymptomatic. A painless abdominal mass is the presenting feature in 30%-45% of the cases [6]. Currently asymptomatic small cysts (less than 5 cm in diameter) are treated conservatively with observation and imaging follow-up [7].

Splenomegaly is the main symptom in children. It becomes evident if the cyst size is greater than 5 cm, as seen in the exploration of our patient. Abdominal ultrasound is useful as an initial diagnostic exam in splenic cysts and allows a rapid diagnosis, CT scans show topography, size, probable nature and anatomical features [8].

Histology and immunohistochemical identification helps differentiate epidermoid cyst from other cystic lesions particularly lymphangioma cyst as observed in our case. Epidermoid cysts have stratified squamous epithelium with a fibrocollagenous cyst wall [9]. The differential diagnosis is usually broad, and making an accurate diagnosis pre-operatively is a challenging task for clinicians [10].

The fragile spleen, hidden in its left upper quadrant recess, always presents an interesting and difficult challenge to the surgeon, while the presence of an epidermoidcyst makes it an even harder challenge. Options for intervention include cystectomy, partial or total splenectomy, and more recently percutaneous aspiration with or without sclerotherapy [7].

Current literature recommends splenectomy or cystectomy with very rare use of aspiration with and without sclerotherapy.

The choice of treatment depends on cyst size; traditionally, large and symptomatic lesions have been treated by total splenectomy via median or left subcostal laparotomy, such what was performed on our patient. However, since the first laparoscopic splenectomy of a splenic cystic performed by Know et al. in 2001, 16 several authors proposed the laparoscopic approach as the technique of choice.

Many surgeons recommend total splenectomy as standard treatment of splenic cyst [11]. Although, Since the 1980s, partial splenectomy with resection of the cyst has been proposed as the first option, however it is not defined how much splenic tissue needs to be preserved to maintain function (perhaps 25% of the spleen) [12]. Furthermore, the objective of resection of the splenic cyst should be local exeresis of the cyst, preserving as much splenic parenchymal mass as possible or even the entire spleen in order to prevent the complications of asplenia, which was the attitude in our case. Partial open splenectomy has been proven safe and effective [13]. Surgical resection is performed with the risk of infection (e.g. Salmonella) or spontaneous bleeding or rupture or trauma, which can be life-threatening [9]. Sometimes the resection of the cyst alone may be difficult and total splenectomy must to be performed because of the large size of the cyst and the scarce actual splenic tissue found during the surgical procedure as reported by Beatriz Martínez and al. [12]. Also if there is anatomical variations in the splenic hilar vasculature. When the main splenic artery is long and divides near the hilum, partial splenectomy is more and more difficult [13].

The rarity of this diagnosis limits comparison to assess the efficacy of treatment modalities. Based on the current literature and the data from the study of Jenine Hassoun et al, an open or laparoscopic partial splenectomy may be the best balance of spleen sparing and recurrence risk reduction [7].

4. Conclusion

Splenic epidermoid cysts are exceedingly rare. Despite its rarity, we must keep it in mind in front of all splenic lesions with cyst features and try to achieve the most conservative treatment possible, which is not always feasible, because the real efficacy of prophylactic measures post splenectomy is not fully observed.

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