

## Splenic Cysts in Adolescents: A Case Report and Review of the Differential Diagnosis and Contemporary Management

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### Abstract

Splenic cysts are uncommon in adolescents. The differential diagnosis includes parasitic, neoplastic and traumatic etiologies each with unique management principles. When surgery is indicated, the importance of preserving viable splenic tissue in children to eliminate the risk of overwhelming post splenectomy infection (OPSI) is critical. We present the case of a 17-year-old male with a splenic cyst. The case allows for a discussion of the etiologies, concerns and contemporary management of splenic cysts in children.

**Keywords:** Laparoscopic Partial Splenectomy; Congenital Splenic Cyst; Epidermoid Cyst; Hydatid Cyst; OPSI

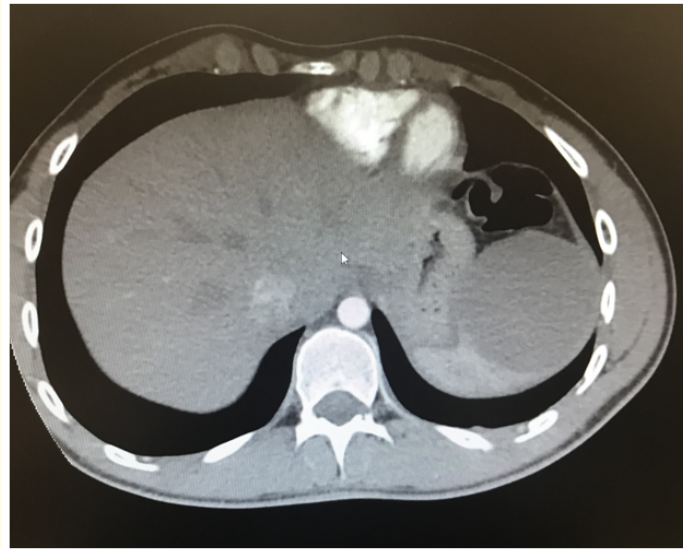
### Introduction

Splenic cysts include a diverse group of pathologic conditions that have few evidence-based management strategies [1]. In children, these cysts often become symptomatic and require removal for pathologic analysis [2]. There has been a focus on partial splenectomy in an attempt to preserve splenic parenchyma and function to reduce the risk of OPSI which can occur in as many as 5% of children post operatively [3-5]. Laparoscopic partial splenectomy (LPS) in these circumstances is ideal. We present a case of a 17-year-old with a complex splenic cyst of unknown etiology managed successfully with LPS.

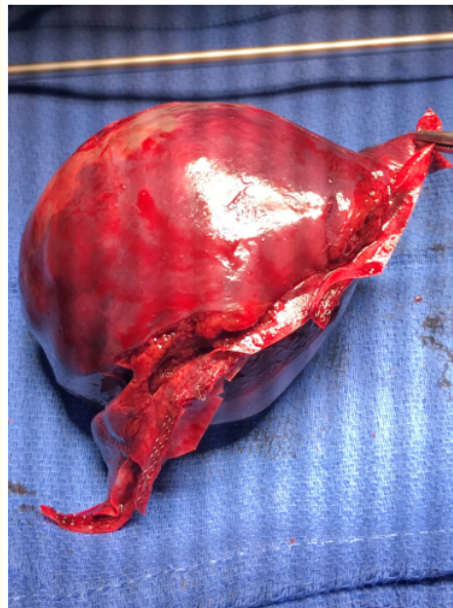
### Case Report

A 17-year-old active and athletic teenager presented with mild left upper quadrant pain and left shoulder pain after exertion. He had a history of previous rigorous skiing the week prior to symptoms. He had an otherwise normal ROS and physical exam that was significant for mild left upper quadrant tenderness. His electrolytes, liver function tests and complete blood count (CBC) were normal except for eosinophilia. An ultrasound of the abdomen revealed a complex cystic lesion in the upper pole of the spleen. A CT of the abdomen revealed a 7.2 x 6.1 x 7.1 cm irregular cystic lesion in the spleen (Figure 1). He had a family history of living in Madagascar as a young child with an uncle and cousin who had undergone hydatid cyst surgery. He underwent a work up that included normal cold agglutinin and hydatid serum titers. Because his exam was normal, a period of observation was recommended during which he underwent the appropriate vaccination for potential splenectomy. The working diagnosis was traumatic splenic cyst. A repeat US 5 weeks later revealed a slightly larger cyst measuring 7.3 x 6.8 x 8.3 cm with irregular walls and multiple internal echoes that was also increased from the prior US. Surgical

removal was indicated. To preserve splenic tissue and mitigate the risks of OPSI, he underwent LPS. The final pathology proved to be an epidermoid cyst. His post-operative course was unremarkable and he has a normal exam on 1 year follow up.



**Figure 1:** Representative CT image of the patient's complex splenic cyst.



**Figure 2:** Intact subtotal splenectomy with complex cyst and margin of normal tissue.

### Discussion

Splenic cysts in children are uncommon. The differential diagnosis includes parasitic and non-parasitic cysts. Non-parasitic cysts can be further subdivided as primary and secondary cysts which is based upon the single characteristic of the presence or absence of an epithelial cell lining. Primary or type I cysts possess this epithelial lining whereas secondary or type II cysts (pseudocysts) do not. Congenital, vascular, and neoplastic splenic cysts make up the non-parasitic class of primary cysts, whereas secondary type II cysts usually are the result of splenic trauma. These type II cysts can often be diagnosed preoperatively by medical history and imaging studies [1,7]. These cysts are usually managed successfully non operatively. However, determining whether a cyst is parasitic or a non-parasitic primary type I cyst can be challenging [5].

Up to 60% of primary splenic cysts are parasitic from an infection by the zoonosis of the larval form of the tapeworm *Echinococcus granulosus* (EG). EG is one of the four species of *Echinococcus* that cause a pathology in humans known as cystic echinococcus (CE), or hydatid cyst [8]. Parasitic cysts are the most common cause of splenic cysts worldwide and are the predominant type of splenic cyst found in patients in Africa and South America. Non-parasitic cysts have a higher prevalence in North America and Europe. Although CE primarily affects the liver and lungs, splenic hydatid cysts can be present in up to 2% of abdominal and 0.5 - 8% of total CE cases. EG is responsible for 50 - 80% of parasitic cysts of the spleen in south and central Europe, South America, and Australia. These hydatid cysts can present as a single, solitary complex lesion in 38% of patients, with the remainder (61%) of splenic hydatid cyst cases presenting as multiple lesions with peripheral daughter cysts [9].

Non-parasitic primary type I cysts are often found in patients less than 20 years of age suggesting a congenital origin. There are 3 types of congenital cysts: epidermoid, dermoid, and endodermoid. Epidermoid cysts result from embryonic inclusion of epithelial cells from adjacent structures, followed by cystic dilation or may be the result of invagination of capsular surface mesothelium. Dermoid cysts are considered to be cystic teratomas, containing structures derived from all three germ layers, and endodermoid cysts represent cystic vascular lesions comprised of multiple ectatic vessels [10].

Ultrasound is often the first diagnostic test. It can reveal many important features of a splenic cyst, including whether it is uni- or multilocular, location in and/or on the spleen, relationship to surrounding structures, septations, irregularities of the cystic wall, internal debris (cholesterol crystals or breakdown products following hemorrhage), hemorrhage, and calcifications [11]. Computed tomography (CT) or magnetic resonance imaging (MRI) can further elucidate the nature of the cyst, such as composition of fluid, dimensions of length, and exact anatomic location [12]. However, despite these modalities, it can be a challenge to distinguish between parasitic and non-parasitic cysts. The classic ultrasound findings of a splenic hydatid cyst are infoldings of the inner cyst wall, separation of the hydatid membrane from the wall of the cyst, and hydatid sand. Infoldings of the inner cyst wall, representing daughter cysts, can appear as characteristic multiloculated internal septations with a thickened wall. These findings can also be present in non-parasitic cysts and indeed were present in our patient. The hydatid sand that is described as an intricate sonographic image, consists primarily of the distinct hook-shaped structures on the head of the juvenile parasite in its larval form and is diagnostic [13]. Although, hydatid sand can be used to distinguish parasitic from non-parasitic cysts, many non-parasitic cysts can have irregular floating debris mimicking this ultrasound findings which also existed in our patient. Serological testing, specifically immunoelectrophoresis, ELISA, and indirect hemagglutination, is then done to confirm *Echinococcus* infection. A combined approach of imaging and serological testing does improve the diagnostic accuracy of a splenic hydatid cyst in majority of cases, however, 10% of the cases remain equivocal prior to surgery and pathologic analysis [14-16]. Although the goal of an accurate pre-operative classification of the splenic cyst is optimal, it is not uncommon for the diagnosis to remain equivocal, especially in children.

Our patient was a diagnostic dilemma. He was very active and his acute presentation was more consistent with a complex traumatic cyst. Therefore, the decision for a trial of nonoperative management was indicated. In addition, spontaneous regression of congenital

splenic cysts has been reported [17]. We utilized this 5-week time period to have him undergo vaccination and we also performed the serologic testing for EG which was normal. The repeat US was concerning because of the increase in septations and interval growth over a short period of time.

The importance of maintaining viable splenic tissue is of paramount importance. LPS may be technically challenging with a higher risk than laparoscopic total splenectomy. However, the benefit of preserving spleen and its immunologic function in children may outweigh this increase risk. Patients who undergo splenectomy or have congenital asplenia are vulnerable to sepsis and OPSI. OPSI occurs at an estimated incidence of 0.23 - 0.42% per year, with a lifetime risk of 5%. The risk is much higher in children. In fact, if splenectomy is required it should be delayed until after the age of 6 years [18]. Episodes of OPSI are emergencies with a mortality of 38 to 69%. *Streptococcus pneumoniae* is the most common organism accounting for 50 - 90% cases. In children, surgical conservation of the spleen with LPS is optimal. In preparation for any operation in a child where total splenectomy is possible vaccination against *S. Pneumonia*, *H. Influenza* type B, and *N. Meningitidis* is indicated [19]. This should be done as far in advance of the procedure as possible to allow for an appropriate immunologic response.

### Conclusion

In conclusion, children with complex splenic cysts often require surgery for diagnosis and treatment. Surgical options should be based on the unique experiences of the team. However, a contemporary management strategy should include attempts at preserving functional splenic tissue to mitigate the risk of OPSI.

### Disclosure Statement

No competing financial interests for the author.

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