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A 16-Year-Old Saudi Boy with a Symptomatic Large Splenic Epidermoid Cyst

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Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
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Patient: Male, 16-year-old
Final Diagnosis: Splenic epidermoid cyst
Symptoms: Abdominal asymmetry • abdominal distention • decreased oral intake
Medication: —
Clinical Procedure: Open splenectomy
Specialty: Surgery

Objective: Rare coexistence of disease or pathology

Background: Splenic epidermoid cysts rarely occur and the majority of cases are discovered incidentally. However, large cysts can present with symptoms due to mass effect. Splenectomy is the treatment indicated for most splenic cysts and definitive diagnosis can only be made after histopathological examination.

Case Report: A 16-year-old boy was referred to the general surgery clinic from a local hospital with a history of abdominal asymmetry, distention, decreased oral intake, and early satiety. Abdominal computed tomography (CT) was performed and revealed a large splenic mass measuring 18.4×14×17.4 cm with multiple mural calcifications at the rim. The mass was displacing the stomach, jejunal loops, pancreas, left kidney, and transverse colon. Preoperative biopsy was not possible because a parasitic cyst was suspected; therefore, splenectomy was performed through midline laparotomy. The patient recovered well, with no complications except for transient reactive thrombocytosis. Histopathology was reported as benign epithelial cysts, with the most likely diagnosis being a splenic epidermoid cyst. Subsequently, the patient was followed up at the surgery clinic. During the last outpatient visit, the patient exhibited good recovery with no problems.

Conclusions: Large splenic epidermoid cysts are rare, particularly in the pediatric population. Nonspecific clinical and radiological findings are hurdles to an accurate diagnosis. Preoperative diagnosis is crucial to determine the type of intervention. However, a final diagnosis can only be made after histopathological examination.

Keywords: Pediatrics • Spleen • Splenic Neoplasms

Full-text PDF: https://www.amjcaserep.com/abstract/index/idArt/934503

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Background

Splenic cysts are rare and are classified as primary or secondary based on the presence of an epithelial lining [1]. Primary cysts are lined by epithelium and account for 10% of all benign nonparasitic splenic cysts [2], whereas secondary cysts are lined by fibrous tissue [1].

Epidermoid cysts of the spleen are considered to be primary congenital splenic cysts. Splenic epidermoid cysts are commonly present in children and young females. It is mostly diagnosed incidentally during imaging and has a good prognosis [1,3]. Some large cysts are associated with symptoms mainly due to the effect of the mass. The symptoms may include abdominal pain, nausea, and a palpable mass in the left upper quadrant. Furthermore, a small percentage of patients may present with complications such as bleeding or rupture [3].

It is difficult to diagnose epidermoid cysts through imaging alone, as differentiating them from parasitic cysts poses a challenge in many cases. Therefore, histological examination is necessary to confirm the diagnosis [1,4]. The management of splenic cysts depends mainly on the presence of symptoms and on the size of the cyst. Surgery is indicated in cysts >5 cm or in cysts that cause symptoms [2,5,6].

In this case report, a giant epidermoid cyst of the spleen measuring 18.4×14×17.4 cm in a symptomatic pediatric patient is presented. Written informed consent was obtained from the patient's legal guardian.

Case Report

A 16-year-old boy was referred to the general surgery clinic at King Saud University Medical City due to abdominal asymmetry and distention for more than a year that has been noticed by his parents. The patient also reported early satiety and decreased oral intake. CT of the abdomen performed in a local hospital revealed a large splenic cyst. The rest of his medical and surgical histories were unremarkable. The patient did not have any history of fever, night sweating, or weight loss. There was no history of travel to parasite-endemic areas and the patient denied any history of trauma. During physical examination, the patient was vitally stable, with a soft abdomen. A large, soft, and nontender left upper quadrant mass that is crossing the midline was observed on palpation. Routine laboratory tests, including complete blood count, liver function test, and renal profile, were performed and were unremarkable.

CT of the abdomen and pelvis was performed in the hospital with i.v. contrast. The CT result was reviewed and reported by a senior radiologist. The findings included splenomegaly of 21 cm in the craniocaudal dimension displacing the stomach, jejunal loops, and pancreas toward the right side and displacing the kidney and transverse colon inferiorly. Moreover, a large unilocular cystic splenic lesion measuring 18.4×14×17.4 cm with no obvious internal septations or enhancement was observed (**Figures 1, 2**). Multiple foci of mural calcifications were also observed on the rim (**Figure 3**).

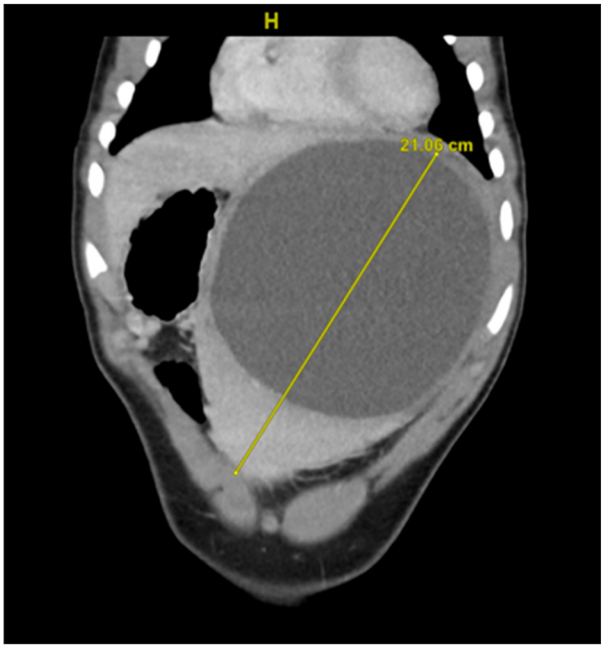

Figure 1. Coronal view of the abdominal computed tomography scan, revealing a large splenic cyst displacing the stomach, with a craniocaudal dimension of 21.06 cm.

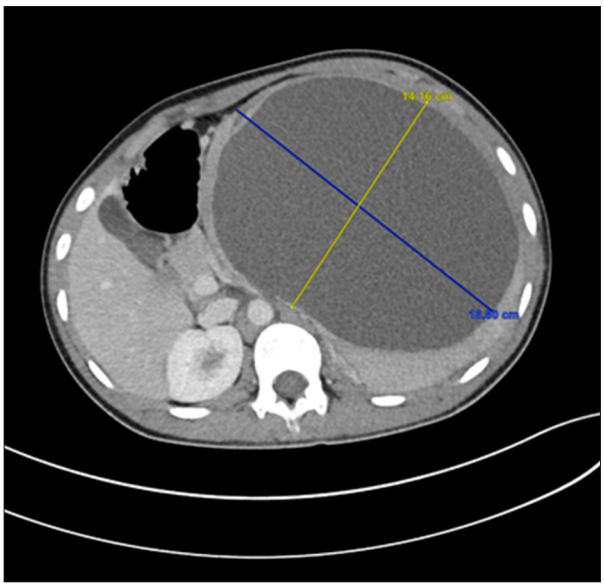

Figure 2. Axial view of the abdominal computed tomography scan, revealing an 18×14 cm splenic cyst displacing the stomach and major vessels.

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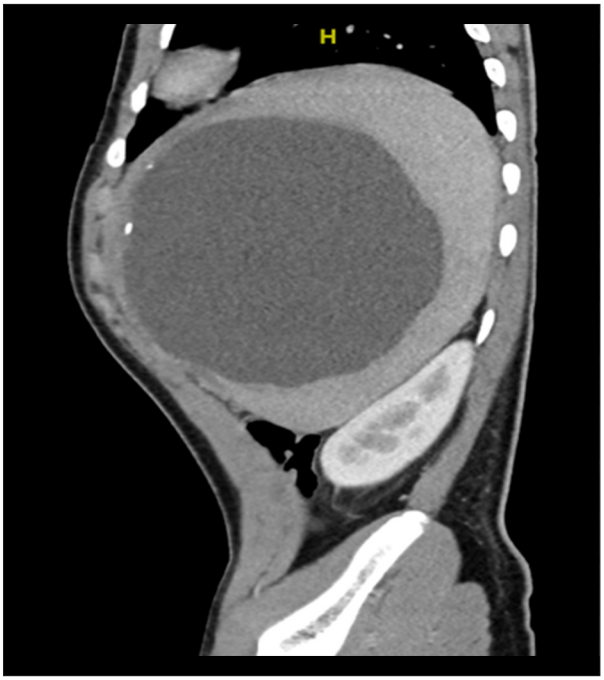

Figure 3. Sagittal view of the abdominal computed tomography scan, revealing a large splenic cyst with multiple foci of mural calcifications.

A plan made was to proceed with splenectomy, as the patient had a large symptomatic splenic cyst. Due to the presence of calcification, hydatid cysts were one of the differential diagnoses. Enzyme-linked immunosorbent assay (ELISA) test for *Echinococcus granulosus* was planned, but unfortunately it was not available on our hospital at that time. The open approach was the best option in this case because the cyst was very large and was located in the small abdominal cavity of a pediatric patient. In addition, the suspicion of hydatid cysts makes cyst rupture and anaphylactic shock possible risks, leading to the exclusion of the minimally invasive surgery option.

The patient was prepared for surgery; meningococcal and pneumococcal vaccines were administered 2 weeks prior to admission. Written informed consent was obtained from the patient's legal guardian. The anesthesia team was informed about the possibility of the cyst being a hydatid splenic cyst, and full preparation was executed by the team accordingly. Normal saline (3%) was prepared in the operative room for cyst content spillage. Full midline laparotomy and exploration were performed, and an enlarged spleen with a large cyst was revealed. The spleen was crossing midline medially and below the umbilicus level inferiorly, with engorged and multiple abdominal vessel collaterals. Owing to the effect of the mass, it was difficult to explore and control the splenic hilum. Hence, lateral mobilization was achieved to facilitate hilum exposure. The splenocolic, splenorenal, and splenophrenic ligaments were removed. The short gastric vessels were controlled with multiple silk ties. The spleen was delivered through a midline incision. The splenic hilum was dissected and controlled using a stapler device. The specimen was removed from the abdomen (**Figure 4**). The weight and dimensions were obtained and documented as 3378 g and 35×13×20 cm, respectively (**Figure 5**). The cyst was then opened, revealing a dark hematoma-like fluid. Fluid samples and specimens were sent for examination.

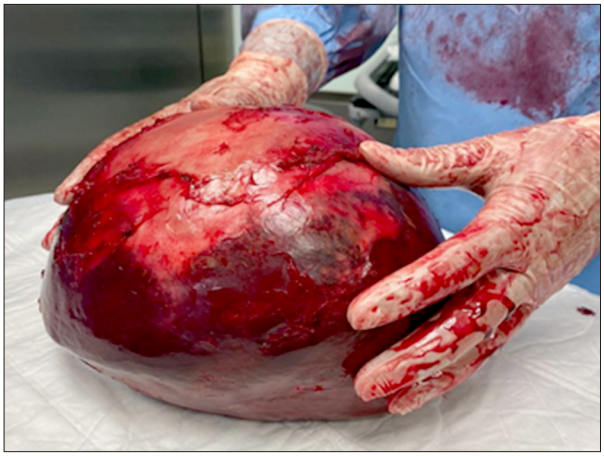

Figure 4. Specimen gross picture. The spleen is presented in relation to the surgeon's hand.

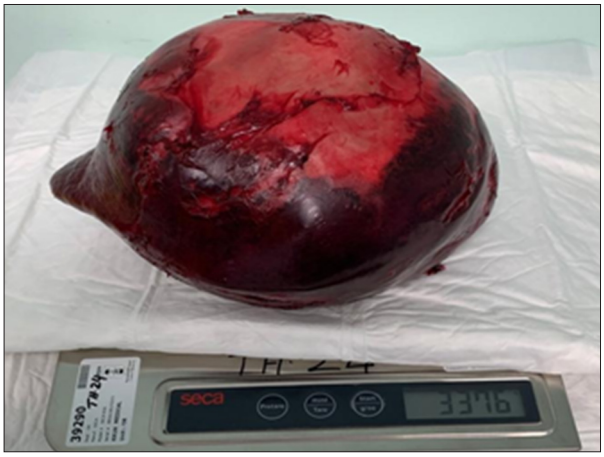

Figure 5. Immediate specimen weight of 3376 grams.

The patient was shifted to the ward postoperatively. He was immediately started on a diet, which was advanced as tolerated, and he tolerated the regular diet very well. There was also a mild transient increase in platelet counts (1 746 000), which was initially managed by starting the patient on oral acetylsalicylic acid 81 mg daily. The Hematology Department was consulted, and their impression was postsplenectomy transient thrombocytosis. As such, they advised aspirin discontinuation. Subsequently, the platelet counts gradually decreased. The patient recovered well and was discharged on day 5 postoperatively.

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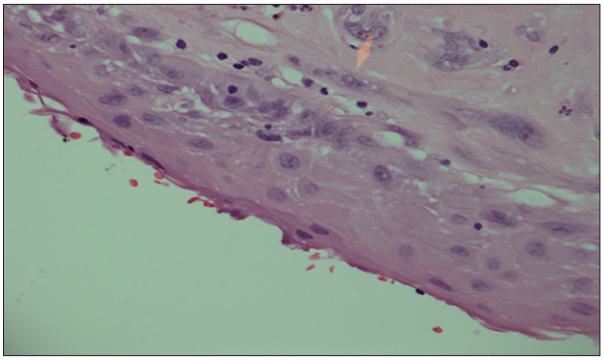

Figure 6. High-power microscopic view of the cyst lining, showing that the splenic cyst was lined by squamous epithelium. Hematoxylin and eosin staining, ×400.

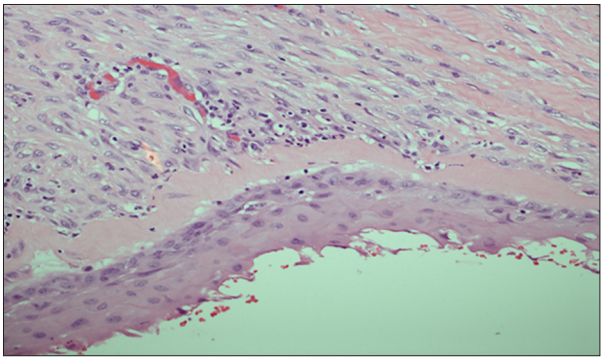

Figure 7. Middle power microscopic view of the cyst showing a lining of squamous epithelium with underlying partly hyalinized fibrous connective tissue and inflammation. Hematoxylin and eosin staining, ×200.

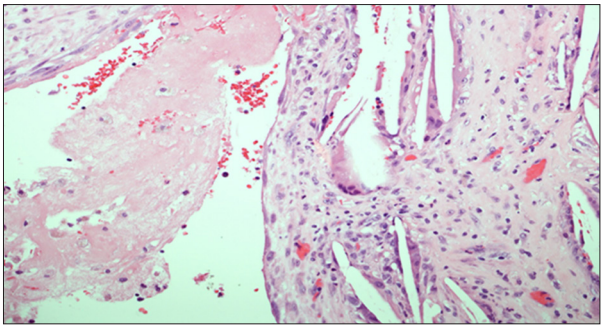

Figure 8. Part of the cyst was lined by fibrinous material with cholesterol clefts and giant cells, which are indicative of old hemorrhage. Hematoxylin and eosin staining, ×200.

Splenic cyst content cytology showed blood with foamy macrophages, consistent with the contents of a hemorrhagic cyst. Histopathology reported it as a benign epithelial cyst lined by squamous epithelium, consistent with an epidermoid cyst (**Figures 6-8**).

The patient was observed in the clinic shortly after discharge. He demonstrated good recovery and had no problems. The surgical site was examined and no signs of infection were observed during removal of the surgical clips. A repeat complete blood count was performed, which showed a normal platelet count. The patient and his family were reassured by the pathology report. The patient was observed in the clinic 23 days postoperatively. There were no problems, and he was discharged from the surgery clinic with a medical report for future reference.

Discussion

Primary splenic cysts are divided into parasitic and nonparasitic cysts. Nonparasitic cysts are usually congenital, such as epidermoid cysts. By contrast, parasitic cysts are usually caused by the parasite *Echinococcus granulosus*, known as a hydatid cyst [7]. These cysts are difficult to differentiate based on clinical and radiological findings alone [8,9] due to similarities among various cystic lesions in terms of their clinical presentations and the findings from laboratory and imaging studies. Their clinical manifestations are usually nonspecific and are shared among a wide variety of intra-abdominal masses. Therefore, achieving an accurate preoperative diagnosis of a primary cyst in the spleen is often difficult. A study conducted by Chen et al showed that symptoms such as left upper abdominal mass (57.7%) and left upper abdominal pain (39.0%) predominated their sample size of 115 splenic epidermoid cyst cases [6]. Similarly, our patient presented with a large splenic cyst, with symptoms mainly related to the mass size effect.

Serological tests can provide clues about the diagnosis, especially in epidermoid cysts. In many cases, elevated CA-19-9 and CA-125 levels dropped precipitately after splenic resection [10]. In addition, serological testing for echinococcus – ELISA and passive hemagglutination – with the absence of a hydatid cyst on ultrasound and CT can provide hints for the exclusion of echinococcus as the cause of the splenic cyst [1].

Features such as intramural calcifications and septations favor the diagnosis of parasitic cysts [9]. However, epidermoid cysts usually appear as well-circumscribed unilocular cystic lesions with variable presentation of wall calcifications [9]. Therefore, ultrasound and CT scans are invaluable tools for further characterization of the cyst in question. The i.v. contrast CT findings in our patient demonstrated a unilocular splenic cystic lesion with intramural calcifications. This posed yet another obstacle, as the presence of calcifications raised the suspicion of a hydatid cyst as opposed to a simple epidermoid cyst. Therefore, the preoperative scope of our investigation became limited due to our inability to safely biopsy or aspirate the cystic content without risking the possibility of spillage and anaphylaxis [11].

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Surgery is indicated in cysts >5 cm or in the presence of symptoms. However, surgical techniques mainly depend on the cyst size and type [1]. Open splenectomy is the criterion standard for treating large symptomatic cysts. Recently, preservation of parenchyma has been highly indicated to prevent lethal infections, especially in the pediatric population. Moreover, partial open splenectomy has been proven safe and effective, and anatomic variations of the splenic hilar vasculature should be considered in cases of partial splenectomy [3].

In a study conducted by Szczepanik et al, 11 patients with benign splenic cysts were evaluated, and their average cyst diameter was 9.1 cm; moreover, 10 of these patients underwent open partial splenectomy. All of them had uneventful postoperative complications, with no cyst recurrence or infections documented on follow-up [12]. This indicates that open partial splenectomy is safe and ensures complete cyst removal and furthermore decreases the recurrence rate with preservation of splenic function.

Laparoscopic partial splenectomy is also feasible for benign splenic cysts according to a study conducted by Chen et al, wherein 16 patients were preoperatively diagnosed with different types of splenic cysts. None of the patients were converted to open surgery, and none required blood transfusions or had postoperative complications [13]. This was further supported by a study conducted by Wang et al, who performed laparoscopic partial splenectomies on 11 patients with focal benign splenic cysts. Their patients were likewise discharged uneventfully with preserved splenic function [14]. By contrast, our patient had a cyst >5 cm, which had been suspected to be a hydatid cyst. This excluded the option for partial splenectomy and minimally invasive surgery in our case.

Percutaneous aspiration is a treatment option documented in the scientific community; however, in many cases, this approach leads to recurrence [3]. Sclerotherapy is another reasonable option for treatment of epidermoid cysts; but the process of choosing a sclerosing agent remains unclear due to limited data, and recurrence and residual cysts have also been reported numerous times in the literature [15]. Furthermore, due to the invasive nature of sclerotherapy, it is crucial to exclude the possibility of a parasitic cyst to prevent intraperitoneal dissemination and anaphylaxis [15].

In our case, a young male patient presented with mass effect symptoms secondary to a large splenic cyst. The patient underwent an open-approach splenectomy with a full midline laparotomy. The operation proceeded smoothly, with no intraoperative or long-term complications. The diagnosis of epidermoid splenic cysts was confirmed by histopathological examination.

Conclusions

The decision to use surgical intervention for splenic cyst is dependent on multiple factors. The diagnosis is crucial to select the approach and type of surgery. Open splenectomy is the treatment of choice and criterion standard approach, particularly in large cysts. Nevertheless, the minimally invasive approach has also gained popularity and should be utilized in selected cases.

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