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# **Giant Epidermoid Cyst of the Spleen: A Case Report**

I. Boucebaine<sup>1\*</sup>, S. Irtan<sup>1</sup>, L. Karima<sup>2</sup>, L. Derradj<sup>3</sup>, Y. Benaissa<sup>3</sup>, L. Ammari<sup>3</sup>, Taibi<sup>3</sup>

<sup>1</sup>Department of Pediatrics, Creteil Intercommunal Hospital, Creteil, France

<sup>2</sup>Department of Pediatrics, Oran University Hospital, Algeria, France

<sup>3</sup>Department of Pediatrics, Bordj Bou Arreridj EPH, Algeria, France

\*Corresponding author: I. Boucebaine, Department of Pediatrics, Creteil Intercommunal Hospital, Creteil, France; Email: imeneboucebaine@gmail.com

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## Abstract

Splenic cysts are a rare entity. They are divided into two groups: Parasitic and non-parasitic splenic cysts. Epidermoid cysts of the spleen belong to the primary non-parasitic group. They usually occur in children. They are most often asymptomatic, but they can present with abdominal discomfort.

**Case presentation:** We report the case of a 12-year-old child with a large splenic cyst. Additional examinations suggested an epidermoid cyst. A total splenectomy was performed *via* laparotomy. The pathological examination confirmed the diagnosis.

**Conclusion:** Recently, conservative surgical treatment in children has become the gold standard; in certain cases, a total splenectomy is mandatory if the cyst is large or if its anatomical position do not allow conservative treatment.

Keywords: Splenic cysts; Epidermoid cyst; Splenectomy

### Introduction

Epidermoid cysts of the spleen are very rare. They are primarily Primary Non-Parasitic Splenic Cysts (PNSCs), representing approximately 2.5% of splenic cysts. They most commonly occur in children and young women and have a good prognosis overall. Splenic cysts are asymptomatic in approximately 30% of cases and are usually discovered incidentally. In exceptional cases, they present with certain complications, such as infection, rupture and bleeding. Currently, the diagnosis of epidermoid cysts has become easier given the advances in diagnostic techniques; however, only a few cases have been published in the literature to date. Traditionally, surgical treatment is recommended for symptomatic cysts larger than 5 cm. We report in this work a case of a giant epidermoid cyst of the spleen [1].

### **Case Presentation**

A 12-year-old child, with no past medical history, presented to the pediatric surgery emergency department with a left hypochondrium mass (Figure 1). Upon questioning the patient, it was noted that he had few symptoms apart from an early gastric fullness sensation during meals associated with left hypochondrium pain and a palpable mass extending from the left hypochondrium to the pelvis. The abdominal ultrasound, performed as a first-line examination, indeed revealed a large mass in the left hypochondrium. We supplemented this with an abdominal CT scan [2].

#### Diagnostic

Abdominal ultrasound reveals a large, well-circumscribed cystic formation measuring  $133 \times 134$  mm with a finely thickened fluid content with fine echogenic spots located in the left hypochondrium. The structure appears to be dependent of the spleen located in its lower pole [3].



Figure 1: A curvature of the left hypochondrium.

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The CT scan confirmed a large well-defined cystic formation (95  $\times$  107  $\times$  117 mm) with regular contours and heterogeneous density.

- Medially, it comes into contact with the splenic vein, which is laminated.
- Inferiorly, it contacts the loops and a branch of the mesenteric artery over a 30 mm length.
- It contacts the aorta over a 40 mm length and the renal vascular pedicle.
- It pushes back the left kidney and pancreas.
- All criteria suggest a hydatid cyst.

We completed the assessment with hydatid serology and tumor markers (CA19.9, AFP), which returned negative. The absence of any evidence of splenic trauma and hematic sediment rule out a post-traumatic cyst. The absence of a daughter cyst and the absence of associated liver lesions in this child argues against a hydatid cyst. Furthermore, the thin, regular walls rule out cystic lymphangioma, whose edges are classically scalloped. Finally, the content of fine echogenic spots that may correspond to cholesterol crystals suggests the diagnosis of an epidermoid cyst of the spleen [4].

#### Treatment

A total splenectomy was performed en-bloc *via* a midline laparotomy given the large volume of the cyst and the tight anatomical relations after vaccination prophylaxis.

The pathological examination confirmed the diagnosis of an epidermoid cyst (Figures 2-4). The outcome was favorable [5].



Figure 2: Ultrasound image shows a large splenic cyst.

 Table 1: Classification table of splenic cysts.



Figure 3: Ultrasound image shows a large splenic cyst.



Figure 4: Intraoperative image showing the large splenic cyst.

## **Results and Discussion**

Splenic cysts, although rare (approximately 1000 cases reported), have been the subject of multiple classifications which distinguish (Table 1) [6].

Clinical classification of splenic cysts (martin)
Primary-True
Parasitic
Non-parasitic
Congenital
Neoplastic
Secondary-False

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Epidermoid cysts of the spleen are rare benign tumors. They represent 2.5% of all splenic cysts and 25% of non-parasitic splenic cysts. Their cause is undetermined; their occurrence possibly results from inclusions of mesothelial remnants in the spleen. This is a congenital pathology that is found mainly in children and young adults, with a female predominance. They are generally small and asymptomatic, discovered incidentally. They can gradually increase in size. Cysts larger than 5 cm may present with splenomegaly, pain in the left hypochondrium, shoulder or chest pain, dyspnea, persistent cough, nausea, vomiting, constipation, dyspepsia, dysphagia or sometimes a palpable mass with signs of compression of nearby organs.

This is the case of our patient who presented with a palpable mass (cyst larger than 5 cm) that was painful and showed signs of compression of neighboring organs. In very rare cases, when the diameter exceeds 8 cm, other complications may occur, such as intracystic hemorrhage or rupture, which induce peritoneal signs such as hemoperitoneum or peritonitis. Superinfections remain exceptional. In the literature, the symptoms and complications of cysts are proportionally linked to their diameter. The growth of the NPSC is slow and occurs over several years, culminating at around the ages of 11 and 12 [7].

On abdominal ultrasound, the most characteristic signs were:

- The existence of a fluid-filled mass with posterior enhancement and finely mobile echogenic content. This content, highly suggestive of the diagnosis of epidermoid cyst, is attributed to cholesterol crystals, keratin lamellae, hemorrhage or intracystic superinfection.
- The existence of a single, well-defined mass with peripheral trabeculae ("festoon sign").

The CT appearance is less specific than the ultrasound appearance. The epidermoid cyst is a benign cystic lesion whose ultrasound appearance is practically pathognomonic. A pathological study of the surgical specimen confirms the diagnosis. On macroscopic examination: The internal wall of the cyst is composed of a network of thick, fibrous trabeculations and areas covered with squamous epithelium. The fluid content is chocolate-colored (Figures 5 and 6). The definitive diagnosis can only be established based on the cytopathology studies with the presence of a cuboidal, columnar or squamous epithelial lining [8].



**Figure 5:** Macroscopic aspects: Of an internal wall composed of a network of trabeculations.



**Figure 6:** Macroscopic appearance of a total splenectomy with chocolate-like appearance.

The spleen has an immunological function, so its preservation should be a therapeutic concern, especially in children. The management of Non-Parasitic Splenic Cysts (NPSCs) remains controversial. Several conservative treatment strategies can be proposed for cysts smaller than 5 cm, such as surveillance, aspiration with or without sclerotherapy or cystectomy.

Conservative treatment for cysts larger than 5 cm increases the risk of recurrence. Partial or total splenectomy is performed conventionally or laparoscopically. Different surgical options have been suggested depending on the patient's age, symptoms, size, nature of the cyst and its anatomical relationships with neighboring organs. Surgical treatment involves partial splenectomy preferably, but this requires the metameric arrangement of the vascularization and polar position of the epidermoid cyst (Figure 7) [9].

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**Figure 7:** Diagram showing the suggested protocol for the management of non-parasitic splenic cysts.

Partial splenectomy is technically more difficult to perform and carries a risk of bleeding during division of the splenic parenchyma. Our case presented a 10 cm cyst with intimate connections to the vessels and adhesions to neighboring organs. In this situation, we recommend a total splenectomy.

## Conclusion

Splenic epidermoid cysts are rare benign tumors. They are often discovered incidentally, despite their sometimes-large size, as in our case. Ultrasound and CT scans can guide the diagnosis, which is confirmed by pathological examination. The surgical treatment of choice is conservative which might sometimes be impossible. If the cyst is large or if its anatomical relationships with the surrounding tissues prevent partial splenectomy, a total splenectomy is recommended.

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