

Splenic cyst in children with recurring abdominal pain: Case report and literature review

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Abstract

Abdominal pain is a common symptom in pediatrics. Cysts non-parasitic splenic are a rare cause of recurrent abdominal pain, but an increase in the number of diagnoses. Originating from the mesothelial cells lining the spleen capsule, occurring in 0.5 - 2% of the population. They may remain asymptomatic for years and usually are discovered incidentally on imaging studies or during the investigation of recurrent abdominal pain. The objective of this report is to elucidate a rare cause of a frequent symptom and major morbidity.

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CASE REPORT

Ten-year-old female patient referred to as M.L.F. presented with complaints of recurrent diffuse abdominal pain for approximately one year, possibly located in the epigastric region and progressively more uncomfortable, but not enough to limit her daily activities. First, a total abdominal ultrasound was performed (Figure 1), which revealed a cystic formation adjacent to the spleen with fine debris in suspension measuring approximately 6.4 cm x 6.3 cm.



Figure 1. Abdominal ultrasound - cystic formation adjacent to the spleen.

We chose to perform a total abdomen MRI (Figure 2) for better resolution of the image, and it revealed an intraperitoneal cyst with internal, regular, and thin septations. The cyst was located in the left upper quadrant and measured 6.0 cm x 6.2 cm, with no cleavage plane with a large gastric curvature or with the spleen, exerting mass effect on these organs. Neither the physical nor the laboratory examinations revealed any abnormalities.

We chose to perform a videolaparoscopy and intraoperatively identified the cyst on the ventral surface of the spleen, in addition to a small hemangioma on the surface of the left hepatic lobe. A splenectomy was performed, but surgical conversion to open laparotomy was required because of bleeding during the release of the upper pole of the spleen.

The surgical specimen was sent to pathology, and the macroscopic examination revealed a cystoid cavity filled with mucous material and smooth inner surface. These results were histologically compatible with splenic cyst involving the mesentery and associated with squamous epithelial metaplasia. The immediate postoperative period took place in the pediatric intensive care unit without complications. Patient recovered without complications and was discharged after 48 hours to receive outpatient follow up.

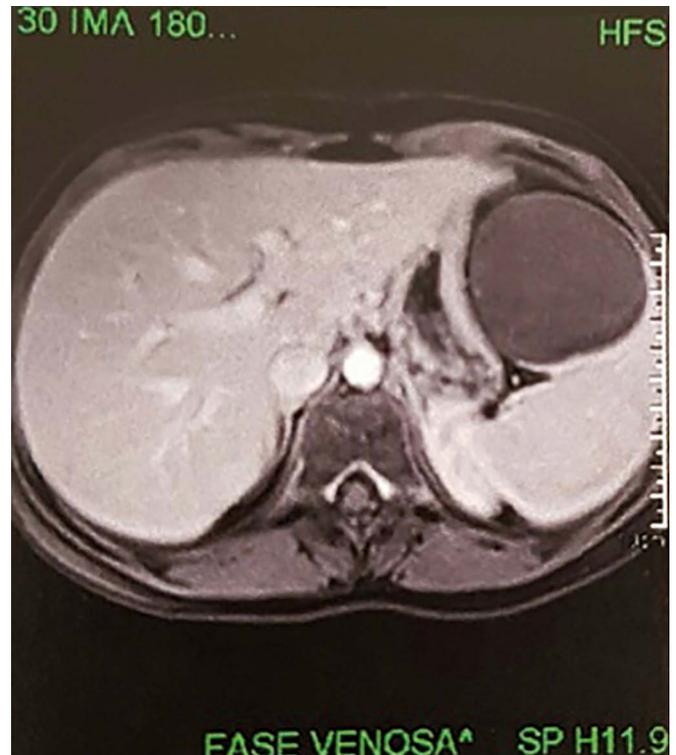


Figure 2. Magnetic resonance- intraperitoneal cyst.

DISCUSSION/REVIEW

The first case of a nonparasitic splenic cyst (NPSC) was described by Andral in 1929. As Table 1 shows, splenic cysts are currently categorized as parasitic (usually caused by *Echinococcus granulosus* and found in endemic areas) or non-parasitic, which, based on the presence of epithelial lining, can also be classified as primary cysts (epithelial/true cysts), which have an epithelial capsule, or secondary cysts (pseudocyst/false) which have no capsule. Secondary cysts tend to be the result of a trauma due to a failure in the organization of subcapsular or parenchymal hematomas; more rarely, they are the result of necrosis or abscesses¹⁻³.

Primary splenic cysts correspond to 10% of NPSCs, and may be congenital or neoplastic in origin. They occur predominantly in children and adolescents and are usually asymptomatic until they reach a large size, at which point they cause local or referred abdominal pain. They can also cause abdominal distension and the compression of adjacent structures, and, more rarely, thrombocytopenia.

Originally, primary cysts were classified as congenital, neoplastic, traumatic, or inflammatory; however, the inclusion of traumatic and inflammatory cysts in this classification has been used with progressively less frequency, because these cysts are a secondary result of traumatic and/or inflammatory processes^{1,2,4}.

Congenital NPSCs account for approximately 25% of primary cyst cases. Classification is based on the epithelial

Table 1. Classification provided by Mirilas et al.².

Classification	Criteria
Primary	
1. Congenital	Cyst lining: mesothelial, transitional and/or stratified squamous. Cyst appearance (inside): bright, white in color, with trabeculation
2. Neoplastic	
Angiomas	
Hemangiomas	Cyst lining: endothelial
Lymphangiomas	Blood content in the cyst
Dermoid cysts	Cyst lining: ectopic, mature ectodermal tissue
Secondary	
Traumatic	History of positive trauma Cyst appearance (inside): "shredded, bleeding," Normal splenic architecture Cyst wall: thick, collagenous
Necrotic	Heart attack: pain in the upper left quadrant Active or previous bacterial endocarditis Non-specific acute splenitis: for example, typhoid fever, infectious mononucleosis, hematogenous spread of streptococcus infection, generalized lymphadenopathy

lining and may be mesothelial, transitional, or squamous. Although the exact mechanisms of its etiology, pathogenesis, and development are still unknown, proposed mechanisms include invagination of the mesothelial capsular surface with subsequent cyst formation, embryonic inclusion or metaplasia of epithelial cells of adjacent structures, as well as a mechanism involving peritoneal endothelial cells¹⁻³.

In pathological examinations, NPSCs exhibit unique features. The mesothelium is usually unilocular and varies in size. The surface is whitish or grayish-white, smooth, and shiny, and the inner portion exhibits coarse trabeculations due to subepithelial fibrosis.

The cyst fluid may vary in density and color, which may range from colorless to a green, yellow, or brown hue. Microscopically, the cyst may have cholesterol crystals and macrophages. The cyst wall is usually lined with mesothelium, squamous or transitional tissue, and there may occasionally be a combination of epithelial cells. The sporadic absence of a cellular lining may lead to the erroneous diagnosis of secondary cyst unless several sections are studied. Table 1 summarizes the histopathological characteristics of CENPs^{1,2}.

Increases in tumor markers such as CA 19-9 and the carcinoembryonic antigen have been associated with primary mesothelial cysts, and CA 19-9 marker levels tend to drastically decrease after cyst removal. Therefore, it can be used as a screening test for recurrence in cases in which the spleen is preserved. CA 19-9 is also found in the analysis of the cyst wall lining, in addition to cells that stain positive for calretinin, a mesothelial marker. The origin is confirmed immunohistochemically¹.

A preoperative diagnosis is rare, but NPSCs can be suspected in cases of unilocular cysts with no previous history of trauma, infection, or exposure to hydatid infection. Some imaging exams may be useful in the investigation, such as ultrasound of the abdomen, which shows whether the cysts are anechoic or hypoechoic, in addition to showing wall thickness.

CTs and MRIs can provide additional information regarding the morphology of the cyst, the composition of the cyst fluid, the precise location of the cyst in the spleen, and its anatomical relationship with the surrounding abdominal organs. Laboratory tests are usually normal, and thrombocytopenia may occur in rare cases. The diagnosis is confirmed histopathologically^{1,5,6}.

The first attempt to excise a splenic cyst was performed by the French surgeon Jules Pe'an in 1867, but the attempt failed because of excessive bleeding, and a total splenectomy was performed. Currently, the conventional treatment for NPSCs is surgical, but conservative treatments may be an option for cysts that are up to 5 cm in diameter, completely asymptomatic, and which exhibit the most typical characteristics of NPSCs^{1,4}.

In cases with surgical indication and due to the growing popularity of spleen preservation to preserve immune function, many alternative treatment options have been suggested. These include aspiration, internal and external marsupialization, partial splenectomy, partial cystectomy (decapsulation), and partial laparoscopic cystectomy^{1,3,4}.

Cyst aspiration as definitive treatment has been described, but it has not been successful. Agents such as tetracycline or alcohol are injected into the cysts to destroy the lining, but cases of recurrence have occurred. Therefore, this procedure is intended as only a temporary measure^{2,5}.

Marsupialization is defined as the creation of an opening in the wall of the cyst in order to drain it. This opening may be internal (within the peritoneal cavity) or external (through an intentional cyst-cutaneous fistula). This treatment leads to cyst recurrence and is not recommended^{3,4}.

Cystectomy with partial splenectomy consists of cyst resection with a contiguous part of the spleen parenchyma and was first reported by Morgenstern and Shapiro in 1980. Removing the cyst wall and its interior in their entirety is the only definitive treatment that can ensure that no remaining cyst tissue remains, leading to the absence of symptoms or recurrences^{3,4}.

To avoid the difficulties of spleen partial removal, some authors have proposed total resection of the cyst, leaving *in situ* the portion of the cyst wall that is contiguous with the parenchyma spleen. This has been referred to as the “decapsulation of the spleen,” but the use of this term is misleading, because the spleen is not decapsulated.

Its advantages include a simpler and faster procedure with less blood loss. Disadvantages include the possibility of recurrence of the cyst, since a portion of the cyst lining is left intact. Recurrence may also occur due to smaller “derivative” cysts in the vicinity of the larger cyst, as has been seen in some patients^{3,4}.

Among the procedures described above, the best results and lowest rates of recurrence have been achieved with partial cystectomy with splenectomy and partial cystectomy. With the advent of laparoscopic surgery, it was inevitable that his method would become part of the surgical treatment for splenic cysts. Laparoscopic methods are now being used ever more frequently, and outcomes have been positive^{1,3,4}.

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