Non-parasitic splenic cysts

Cistos esplênicos não-parasitários

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A B S T R A C T

This article gives an overview of the topic, with emphasis on current concepts and management of the clinical situations in question, in particular the concepts related to treatment. An extensive review of the etiology and monitoring of patients with non-parasitic splenic cysts is also made. These reports are derived from major clinical studies published in the current medical literature.

Key words: Spleen. Cysts/therapy. Cysts/etiology.

INTRODUCTION

The splenic lesions that have cystic presentation include a range of disorders. We discuss these non-parasitic ones, which are: the primary or true cysts (epithelial, epidermoid), pseudocysts (serous or hemorrhagic), vascular cysts (post-infarction, peliosis) and cystic neoplasms (hemangioma, lymphangioma, lymphoma and metastases).

Splenic cysts are more frequent in the second and third decades of life, but they may appear in other age groups[1,2].

In 1829, Andral was responsible for the first description of a non-parasitic cyst of the spleen[3]. Robbins (1978) reviewed a series of 42,327 autopsies over 25 years, founding 32 patients with splenic cysts. Subsequently, isolated cases have been reported and, in 1978, approximately 600 cases were confirmed in the literature[4,5].

Splenic anatomy and physiology

The splenic size and configuration vary. Typical parameters include dimensions of 12 x 7 x 4 cm and a weight of 150 grams (range 100-200 grams) and an anatomic relationship with the bottom of the stomach, the upper pole of left kidney and tail of the pancreas[6].

Splenic trabeculae originate in the internal capsule and divide the body into compartments made of a mesh of lymph follicles and reticuloendothelial cells (white cells) interspersed with blood vessels[6].

The spleen plays an important role in hematopoiesis, immune function, and protection against infections and malignant diseases[6,7]. The main hematologic function of the spleen is to act as a filter to remove use erythrocytes, leukocytes, and older platelets from the bloodstream. Its hematopoietic function occurs almost exclusively in fetal life[8].

The immunologic function of the spleen is complex. In summary, encapsulated bacteria and parasites are removed from the bloodstream, a cellular response to infection is generated and production of antibodies to face the disease ensues[8].

Clinical Presentation

Most cysts are asymptomatic and are diagnosed incidentally during abdominal imaging. The number of splenic cyst diagnosed is increasing, probably due to the large number of radiological examinations currently performed[9].

Large cysts of the spleen (greater than 8 cm) may cause pain and weight sensation in the left hypochondrium, whether by splenic capsule distension or compression of adjacent structures[10,11]. The symptoms of compression on surrounding organs, such as nausea, vomiting, flatulence and diarrhea gradually emerge. In some cases, the effect on the cardiorespiratory system may cause pleuritic pain, dyspnea and persistent cough[4,10,12].

Many diseases that affect the spleen have similar appearance on imaging studies. Thus, history becomes very important to help reduce the number of unnecessary tests and diagnostic hypotheses. If a diagnostic uncertainty persists, percutaneous biopsy is useful for elucidation.

Classification

Splenic cysts were classified by Martin[13]: Type I (primary or real) – are cysts with epithelial capsule, which can be either parasitic or not. Non-parasitic type I cysts may be congenital, vascular or neoplastic; and Type II (secondary

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or pseudocysts)[14], which have no capsule. From the radiological point of view, it is usually impossible to distinguish between primary and secondary cysts.

Congenital cysts (epithelial)

Congenital splenic cysts or epithelial cysts comprise approximately 25% of true splenic cysts[11,13], are diagnosed primarily in children and young adults and are usually solitary.

Although the exact mechanism of their etiology, pathogenesis and development is still unknown, proposed mechanisms include: involution of pluripotent cells from the splenic parenchyma during development with subsequent squamous metaplasia; origin from endothelial cells or peritoneal coelomic mesothelium.

At microscopic analysis there is normally a large, smooth, encapsulated cyst and occasionally trabeculae or septa[15,16]. Microscopically, the cyst is internally lined with columnar, cuboidal or squamous epithelium. They can be subdivided into dermoid, mesothelial, and epidermoid[14,17].

Usually congenital splenic cysts are asymptomatic and have a good prognosis. In some cases they may become symptomatic due to increase in size secondary to trauma, cyst wall hemorrhage with an increase in osmolality of cyst fluid, or the presence of stomata in the cyst wall.

Dermoid cysts

These cysts are extremely rare, with few cases reported in the literature. They may contain skin appendages and squamous epithelium in the interior[14] and their existence is challenged[18] because many of these cysts contain well differentiated tissues. In ectopic location, they can be considered benign teratomas[19,20]. In Addition, they share the same histological appearance of ovarian dermoid cysts that originate from primordial germ cells[19,20].

Pseudocysts

They are called pseudocysts because they do not have capsule[21]. They correspond to about 75% of parasitic cysts of the spleen. They are secondary to trauma, infection or infarction, trauma being the most common etiological factor. Most are solitary and asymptomatic.

They are believed to be the final stage of organization of an intra-splenic hematoma[16,21,22]. A remote history of trauma to the upper left quadrant can be often verified.

Macroscopically, they are mostly smaller than true cysts and may contain internal debris. Microscopically, these cysts are composed of dense fibrous tissue, often calcified, with no epithelial lining. They contain a mixture of blood and debris in their interior[14].

The appearance on imaging studies is similar to that of true cysts, with density varying with the amount of protein and fibrin existing in the interior[22]. Some may rupture spontaneously, 70% of ruptures occurring during the two weeks after trauma and 90% within the first month. The interval between trauma and rupture is called "latent period"[23]. Some pseudocysts increase in size spontaneously or after novel local traumas.

Vascular Cysts

Peliosis

Peliosis is a rare disease characterized by multiple blood-filled cysts located inside the parenchyma of solid organs, mainly in the liver. Exclusively splenic peliosis is a very rare phenomenon. The disease is more common in men[24,26].

Although patients are usually asymptomatic, peliosis may become a potentially lethal condition, given that spontaneous rupture of the organ may occur[27]. If the diagnosis is confirmed, further investigations should be considered to detect the presence of disease in other organs[28].

It was initially thought that peliosis occurred exclusively in the organs belonging to the mononuclear phagocytic system, i.e., liver, spleen, bone marrow and lymph nodes[28]. However, other organs like lungs, kidneys and parathyroid glands may also foster it[24]. Many etiologic agents have been associated with the occurrence of peliosis[28,30], such as toxins (including chronic alcoholism), corticosteroids, oral contraceptives, tamoxifen, azathioprine and androgens.

HIV-positive patients may have opportunistic infectious diseases as an etiological factor[31,32]. Infectious agents such as hepatitis B or C viruses, *Staphylococcus aureus* and tuberculosis may be highlighted[28-33]. Multiple myeloma, Waldenstrom's macroglobulinemia and other malignancies such as Hodgkin's disease, hepatoma and seminoma were also related to the etiology of this disease[30,33].

The identification of peliosis can usually be done macroscopically at operation or during radiological investigation. The surface of the spleen may be nodular[26]; numerous cysts with their cavities filled with blood are found at sectioning of the specimen[25]. Arteries occasionally protrude into the lumen of the cysts. This may contribute to the lethality of the disease. On CT it is seen as a hypodense cyst, causing no mass effect.

Physicians should be alert to the spontaneous rupture of the spleen, especially in the case of anticoagulation or thrombolysis. In literature there is no indication of surgical exploration of patients in whom the diagnosis was incidental splenic peliosis. Patients should be encouraged to refrain from practicing high-risk activities, such as contact sports. It seems prudent to avoid oral contraceptives. In case of visceral rupture emergency splenectomy should be performed[27].

Cystic neoplasms: Hemangioma, Lymphangioma, Lymphoma and Metastases

Lymphoma

Primary splenic lymphoma represents 1-2% of linfomas[1,2,4]. About 60 cases have been reported till 1983[4].

Angioma

The most common type of splenic cystic neoplasm is the blood vessel (hemangioma) or lymph vessel (lymphangioma) angioma. Splenic hemangiomas may be capillaries or cavernous[34], where the capillaries are mostly composed of blood vessels that are in accordance with the normal caliber of the capillary and cavernous hemangiomas are composed of larger caliber blood vessels[35]. Lymphangiomas of the spleen are often cavernous[36,37]. The angiomas are considered congenital, as they are usually present at birth[35,36].

The cystic tumors (eg, cystadenomas) are derived from epithelial cells, usually glandular[38].

In the case of hemangiomas, the cystic cavity may contain blood. The wall covering consists of endothelium, i.e., the normal lining of the vessels of origin, which was demonstrated by positive immunohistochemical staining with antibodies against factor 8 (endothelial antigen) and by negative staining with antibodies against keratin (epithelial and mesothelial antigen)[38].

Metastases

Splenic metastases of solid tumors occur in the terminal phase of the illness, so they seldom have an indication for resection. Tumors arising from the ovaries, lung, breast, stomach, skin and colon may affect the spleen[39]. The number of cases of isolated splenic metastasis is less than 25 in the world literature.

The lesions may be solid or cystic; on ultrasound, they show varying degrees of echogenicity, but are generally hypoechogenic[40]. On Computed Tomography (CT) they typically appear hypodense. On Magnetic Resonance Imaging (MRI) they are predominantly hypointense on T1-weighted, and hyperintense in T2-weighted, images. MRI is more accurate for the diagnosis of splenic metastases with necrotic or hemorrhagic contents, which will display heterogeneous images[40].

Differential Diagnosis

When a nodule is detected in the upper left quadrant of the abdomen, it is necessary to exclude diseases associated with splenomegaly as mononucleosis, hemolytic anemia, chronic leukemia, collagen diseases and liver diseases that cause portal hypertension[41].

Imaging tests

It is difficult to distinguish between true and false cysts both radiologically and histologically.

A plain abdominal radiograph may reveal a mass, which may be calcified, in the left upper quadrant.

Ultrasound can differentiate solid and cystic lesions in most cases. Typically, in ultrasound, the splenic cyst appears as a homogeneous, anechoic mass with thin walls. Septations, irregular walls, or a heterogeneous pattern of internal echogenicity, with debris or hemorrhage and peripheral hyperechoic foci with posterior acoustic shadowing due to calcifications in the wall, can be seen in a complex cyst. Calcifications are useful for differentiating cysts from other causes of splenomegaly[4,9,11]. An epidermoid cyst has a complex pattern with irregularity and thickening of the posterior wall caused by epithelial trabeculae with peripheral and internal echoes in the interior due to the presence of blood clots[11,42].

The spleen is usually heterogeneous on TC, especially during the contrast phase. It is postulated that the reason for this heterogeneity is due to the histological features of the vascular system of the organ, with different rates of blood flow in the red pulp[43]. One must exercise caution to avoid misinterpretation of this heterogeneity.

Cystic lesions on CT are spherical, well-defined, with attenuation equivalent to that of water, with a thin or imperceptible capsule[42] (Figure 1). In one series, cyst wall trabeculation or peripheral septations were found in 86% of true cysts and in 17% of false cysts[43]. There was also wall calcification in 14% of true cysts and in 50% of false ones[42].

The splenic cyst may contain areas with high density due to hemorrhage, increase of protein content or purulent material. This high density can occur in up to 33% of pseudocysts.

Computed tomography is more sensitive than ultrasound in the identification of septa (more common in true cysts) or calcifications (more common in false cysts)[44].

In MRI the cyst is hypointense on T1-weighted images, with strongly hyperintense images on T2, with signal intensity equal to water without reinforcement after contrast injection. However, depending on the contents of the cyst, the signal intensity on T1 can be increased (hemorrhagic cyst), while the signal intensity on T2 remains elevated[42].

Angiography can be useful for differentiating a splenic cyst, which is normally avascular, from a solid malignant mass (lymphoma, sarcoma), which is usually vascularized and has a disorganized pattern[43].

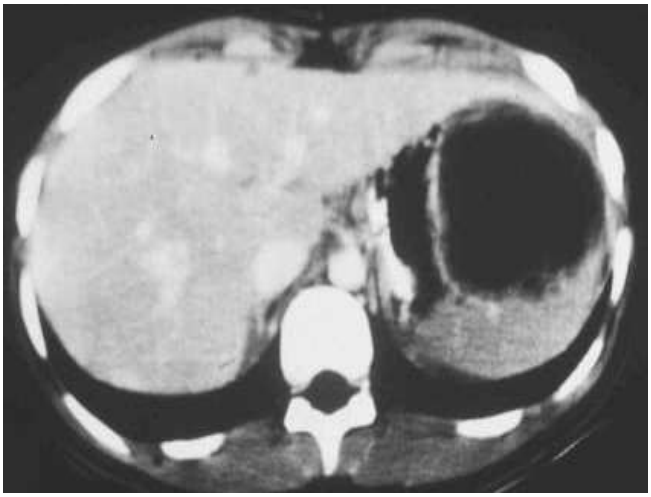

Figure 1 - *Computed tomography of abdomen after administration of intravenous contrast showing a septated splenic cyst with low density content, causing compression of adjacent structures.*

Treatment

Laparotomy with splenectomy has been the method of choice for the treatment of many splenic cysts[11]. Today, more conservative surgical procedures have wider application, especially in children and young adults, in order to avoid serious postoperative infections[17].

Due to the increased risk of complications, splenic cysts with a diameter greater than 4-5 cm should be surgically treated[10,12], as conservative treatment options, such as percutaneous aspiration or sclerosis, do not result in good long term control[11]. Some studies have shown that splenic cyst sclerosis with alcohol was successful for small lesions (up to 11 mm) but not for large ones[44].

Treatment options are: partial splenectomy, total cystectomy, cyst marsupialization or decapsulation, with access either by laparotomy or laparoscopy[11,12,43]. Partial splenectomy is defined as the operation that preserves more than 25% of the splenic parenchyma, (minimum of tissue in order to preserve the immune defense without increasing the risk of cyst recurrence)[11].

Laparoscopic partial splenectomy can be safely performed[14, 20]. This procedure is recommended if the cyst is superficial and is located at the poles of the spleen due to increased risk of recurrence[11]. The incision of the splenic capsule and hemostasis are conducted with electronic cautery or monopolar scissors[11,12]. A more conservative approach would be the cystotomy, however, this technique is still in development, as the amount of tissue surrounding the lesion that should be resected to prevent recurrence is not certain [12].

The major complications associated include infection, rupture and bleeding [10,12,42].

Final Thoughts

Splenic cysts larger than 5 cm or symptomatic should be treated surgically, trying to preserve as much splenic parenchyma as possible. If the cyst is very large and almost entirely covered by parenchyma or located in the splenic hilum, total splenectomy is recommended because there is risk of intractable bleeding. Partial splenectomy is an acceptable procedure in most other cases. The laparoscopic approach appears to be a safe procedure, with all the advantages of minimally invasive surgery.

R E S U M O

Este artigo apresenta uma revisão geral do tema, com ênfase em conceitos atuais e no manejo das situações clínicas em questão, em especial a conceitos referentes ao tratamento. Também é feita uma revisão extensa quanto à etiologia e seguimento dos pacientes com cisto esplênico não parasitário. São citadas informações derivadas dos principais estudos clínicos publicados na literatura médica atual.

Descritores: *Baço. Cistos/terapia. Cistos/etiologia. Literatura de revisão como assunto.*

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