Giant splenic cyst in a teenager girl. Case report

Quiste esplénico gigante en una adolescente. Caso clínico

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Abstract

Giant nonparasitic splenic epidermoid cysts are relatively uncommon. These lesions can lead abdominal pain, but most of them are asymptomatic, and they are discovered incidentally. We report a 13-y old female with a giant splenic epidermoid cystic, given the special interest of diagnostic and therapeutic decision-making of this rare entity. Case report: A 13-y old female with clinical history of abdominal pain since the last two months. On physical examination a firm, hemotender mass was palpable in left hypochondrium. Diagnosis of a large cystic splenic mass was made based on ultrasound and abdominal computed tomography scan. Splenectomy was performed, and histopathological- immunohistochemistry studies revealed findings suggestive of primary epithelial cyst. The post-operative clinical course was satisfactory and uneventful. Conclusions: Treatment of giant nonparasitic splenic cysts is surgical. Preserve splenic parenchyma must be the aim in an individualized decision-making. The different types of surgical modalities will be according to the diagnosis and clinical situation (cyst size, age, comorbidities).

Introduction

Giant and non-parasitic solitary splenic cysts are infrequent. They are incidentally diagnosed in infancy or in young people younger than 20 years old. The case can range from painless abdominal mass to abdominal pain, discomfort or sensation of fullness in the upper left quadrant due to splenomegaly and the pressure exerted by the cyst on adjacent structures, even left omalgia and arterial hypertension can be found in case of compression of the kidney. If the lesion is diffuse and compromises the entire spleen (e.g. tumor), the phenomenon of splenic sequestration and hypersplenism may occur with pancytopenia. Different classifications of non-parasitic cysts according
Clinical case

A 13-year-old female patient who came to the emergency room due to abdominal pain located in the left, intermittent, hypochondrium of two months of evolution. She had no relevant medical-surgical history or history of trauma.

According to the physical examination, a mass of hard consistency in the left hemi abdomen was palpated, which could correspond to a grade 4 spleen according to the Boyd scale (table 2), but because of its large size it was difficult to determine the organ on which it was based.

Blood tests included a biochemical profile within normal limits, and hemogram showed mild normochromic normocytic anemia with a slightly elevated index of red blood cell dispersion. A mass effect on the left hypochondrium was observed in the chest and abdomen x-rays. Abdominal ultrasound showed a giant avascular complex cystic lesion of probable splenic origin. In order to obtain a better characterization of the lesion as possible surgical treatment, an abdominal computed Tomography (CT) with contrast was performed, which revealed a giant cystic mass (17 x 16 x 14 cm) suggestive of splenic lymphangioma or epithelial cyst (figure 1). In addition, a nodule of 20 mm in the right ovary, compatible with a teratoma or hemorrhagic follicle, was observed as a second option.

Following the administration of the first dose of the 13-valent pneumococcal conjugate vaccine PCV13 V (the remainder of vaccination prior to possible splenectomy was administered according to the official immunization schedule), surgical intervention was scheduled.

Due to the large size of the tumor and the doubts about the nature and origin of the tumor prior to the intervention, it was decided to directly approach the

Table 1. Classification of nonparasitic splenic cysts

<table>
<thead>
<tr>
<th>Category</th>
<th>Description</th>
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<tbody>
<tr>
<td>Congenital</td>
<td>With mesothelial, transitional, or epidermoid epithelial cell lining</td>
</tr>
<tr>
<td>Neoplastic</td>
<td>Neoplasms of endothelial origin</td>
</tr>
<tr>
<td>Lymphangioma</td>
<td>Hemangioma</td>
</tr>
<tr>
<td>Degenerative</td>
<td>Cystified splenic infarcts</td>
</tr>
</tbody>
</table>


Table 2. Classification of splenomegaly after physical examination, by Boyd

<table>
<thead>
<tr>
<th>Grade</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>0</td>
<td>Tap the spleen is possible, not palpable</td>
</tr>
<tr>
<td>1</td>
<td>Palpable spleen that exceeds ribs plane</td>
</tr>
<tr>
<td>2</td>
<td>Palpable spleen that goes to the half of an imaginary line that goes from the rib cage to the navel</td>
</tr>
<tr>
<td>3</td>
<td>Palpable spleen that goes just to the navel</td>
</tr>
<tr>
<td>4</td>
<td>Palpable spleen that goes beyond the navel even to get to the right iliac fossa</td>
</tr>
</tbody>
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Figure 1. Abdominal computed Tomography: giant cystic mass (17 x 16 x 14 cm) suggestive of splenic lymphangioma as a first option.
middle laparotomy. Splenectomy was performed; obtaining a spleen that measured 220 x 155 x 120 mm and weighed 2,588 g (figure 2). An anatomopathological study of the splenectomy specimen was performed, containing a subcapsular cyst of 210 x 150 x 120 mm, which had a unique cavity with fluid content with cloudy aspect, limited by a fibrous wall with a trabeculated internal surface (figure 3). The histopathological study showed that the cyst was covered by a non-keratinizing squamous epithelium without atypia, which rested on a band of fibrous tissue of irregular thickness (figure 4).

In zones, the epithelium was markedly thinned, being reduced to one or two layers of cells. The immunohistochemical study revealed that the coating epithelium expressed cytokeratin AE1/AE3 and was negative for calretinin, even in the monolayer zones, thus ruling out a mesothelial differentiation. The final diagnosis was epithelial cyst (also called epidermoid).

A 2 cm cyst was found in the ovary; Considering that tumor markers within the range of normal, and that the cyst disappeared over time, confirmed suspicion of ovarian follicle.

The postoperative evolution and the subsequent ambulatory follow-up were uncomplicated up to 12 months after surgery, when discharge was discharged.

Discussion

Non-parasitic splenic cysts, unique and large, are delimited (at least in part) by epithelium or mesothelium. The splenic cyst may be epidermoid, transitional or mesothelial, or all three at once (the metaplasia of the mesothelium is well known). There are secondary non-parasitic cysts, usually to traumatisms, delimited by dense fibrous tissue with or without calcification and haemosiderin deposition4-6. Within splenic cysts or tumors, lymphangiomas are the rarest in frequency, usually subcapsular, having calcified mural lesions3.

Immunohistochemical identification also helps differentiate lymphangiomas from cystic lesions7. At the moment of the origin of the epithelial cyst, there could be an invagination of pluripotent sectors of the splenic capsule into the splenic pulp during the embryonic period, which would later experience squamous metaplasia.

The classification of splenic cysts can be seen in Table 1, other authors propose to classify according to the extension in the spleen and its relation to the splenic hilum4, which may be useful for the surgical decision.

Splenomegaly becomes evident if the cyst size is greater than 5 cm, as seen in the exploration and
simple radiology of our patient. Abdominal ultrasound is useful as an initial diagnostic exam in splenic cysts and allows a rapid diagnosis, on the same day of the emergency room visit, as in the case presented; but they are not useful in order to delineate the topography of the lesion. CT scans show topography, size, probable nature and anatomical features.

CT and magnetic resonance imaging are of choice in the diagnosis and planning of the surgical strategy. In large cysts, CT with 3-D reconstruction may show the relationship with hilum vessels and remaining spleen size. Although, these data can only be determined at the time of surgery.

Conventional treatment in splenic cysts greater than 5 cm was splenectomy, open, with medial or subcostal incision. Since 2001, laparoscopy has been proposed as an approach of choice, if possible. Surgical resection is performed with the risk of infection (e.g. Salmonella) or spontaneous bleeding or rupture or trauma, which can be life-threatening. Exceptionally, it is described as malignant transformation. The most widely used technique in giant cysts, with limited splenic parenchyma, especially in children older than 6 years, is total splenectomy. Since the 1980s, partial splenectomy with resection of the cyst has been proposed as the first option, although it is not defined how much splenic tissue needs to be preserved to maintain function (perhaps 25% of the spleen).

The consequences of total splenectomy are the microbiological risk of severe infections by encapsulated bacteria (pneumococcus preferentially, also Neisseria meningitidis, even severe malaria). The risk of vascular complications such as atherothrombosis and possible pulmonary hypertension in the long term, in patients with splenectomy, mainly due to hematological disorders (in certain hemoglobinopathies, hemolytic anemias and others), is being studied. In any case, it seems advisable to inform the splenectomized patients and to advise them to avoid cardiovascular risk factors (such as smoking, hypercholesterolemia, sedentary lifestyle, obesity).

In this case presented, we opted for the open approach due to the large size and little remnant splenic tissue, as described in other publications. The ovarian finding initially raised special diagnostic needs and made it advisable to expand the surgical exploration.

Although splenic tissue may appear to be significant in CT, and prior to surgery, partial splenectomy with resection of the cyst was considered as the first option. Finally, total splenectomy had to be performed because of the large size of the cyst and the scarce actual splenic tissue found during the surgical procedure. The family gave the appropriate written informed consent, after receiving information on the possible options, to be confirmed at the intraoperative time.

Conclusions

Surgical treatment of giant non-parasitic splenic cysts should be individualized, taking into account various factors such as size, diagnostic possibilities, residual splenic tissue, patient age and comorbidities. The aim of this procedure, which is not always feasible, is the resection of the cyst retaining as much of the spleen as possible.

Ethical Responsibilities

Human Beings and animals protection: Disclosure the authors state that the procedures were followed according to the Declaration of Helsinki and the World Medical Association regarding human experimentation developed for the medical community.

Data confidentiality: The authors state that they have followed the protocols of their Center and Local regulations on the publication of patient data.

Rights to privacy and informed consent: The authors have obtained the informed consent of the patients and/or subjects referred to in the article. This document is in the possession of the correspondence author.

Financial Disclosure

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Conflicts of Interest

Authors state that any conflict of interest exists regards the present study.
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