Clinical Case Report

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Giant epidermoid cyst of the spleen in a pediatric patient

A case report

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Abstract

Rationale: Splenic cysts (SCs) are rare findings in children, particularly the youngest. Here, we discuss a case that is useful for the differential diagnosis and treatment of SCs.

Patient concerns: A 9-year-old Albanian boy was admitted for severe abdominal pain localized mainly in the left hypochondrium for approximately 24 hours. His medical history was without significant clinical problems.

Diagnosis: Splenomegaly was diagnosed during the first clinical examination, and laboratory tests showed an increase in CA 125 and CA19-9. Abdominal ultrasonography showed splenomegaly with a large hypoechoic oval formation with well-defined margins and the presence of internal fine suspension spots; abdominal magnetic resonance imaging revealed a well-defined SC. The cystic lesion caused major effects on the neighboring organs, shifting them from their normal sites.

Interventions: Considering the mass's volume, an open splenectomy was performed. Upon histopathological examination, the lesion was characterized by a stratified squamous keratinized thick lining and brownish liquid contents consisting of lymphocytes, erythrocytes, and hemosiderin-rich macrophages. These features informed the diagnosis of a giant epidermoid SC.

Outcomes: No complications occurred in the post-operative period, and blood exams revealed the quick normalization of CA 19.9 and CA 125 levels. The boy was discharged on the eighth post-operative day. No complaints were documented during the regular follow-up.

Lessons: This case shows that modern imaging techniques are useful for the differential diagnosis between epithelial mass and SCs of different origins. Open splenectomy has been the treatment of choice for years, but future studies should clarify whether more conservative methods are associated with positive long-term outcomes and if they can also be used for large SCs.

Abbreviations: MRI = magnetic resonance imaging, SCs = splenic cysts, US = ultrasonography.

Keywords: abdominal mass, abdominal pain, giant epidermoid cyst, splenectomy, splenic cysts, splenomegaly

1. Introduction

Splenic cysts (SCs) are rare findings in children, particularly the youngest, although their prevalence is increasing due to the widespread use of abdominal imaging and nonoperative treatment for splenic trauma.[1] They are traditionally classified as primary or true SCs and secondary or false SCs or pseudo-SCs. The presence of an epithelial lining distinguishes the 2 types. True SCs include parasitic cases, mainly due to *Echinococcus granulosus*, and non-parasitic cases, which are in turn divided into congenital or epithelial, vascular and neoplastic (splenic haemangioma, littoral cell angioma, and lymphangioma).[1] False SCs are secondary to trauma or splenic infarcts.[2,3] To simplify the classification, some authors consider all non-parasitic SCs as congenital and do not include trauma as a cause of SCs because post-traumatic lesions are simple intrasplenic hematomas that have failed to organize in the usual manner.[4] The differential diagnosis is not easy because other non-SC conditions involving the spleen, such as inflammatory pseudotumors of the spleen, splenic hamartoma, and splenic abscesses, should be considered.[4] Moreover, when an SC has been identified, the choice of the best therapeutic approach is challenging. Here, we discuss a case that is useful for the differential diagnosis and treatment of SCs.

2. Case

2.1. Presenting concerns

A 9-year-old Albanian boy was admitted to the Emergency Department and had been suffering from severe abdominal pain

Editor: N/A.

The authors report no conflicts of interest.

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Medicine (2019) 98:27(e15653)

Received: 6 January 2019 / Received in final form: 5 April 2019 / Accepted: 17 April 2019

http://dx.doi.org/10.1097/MD.0000000000015653

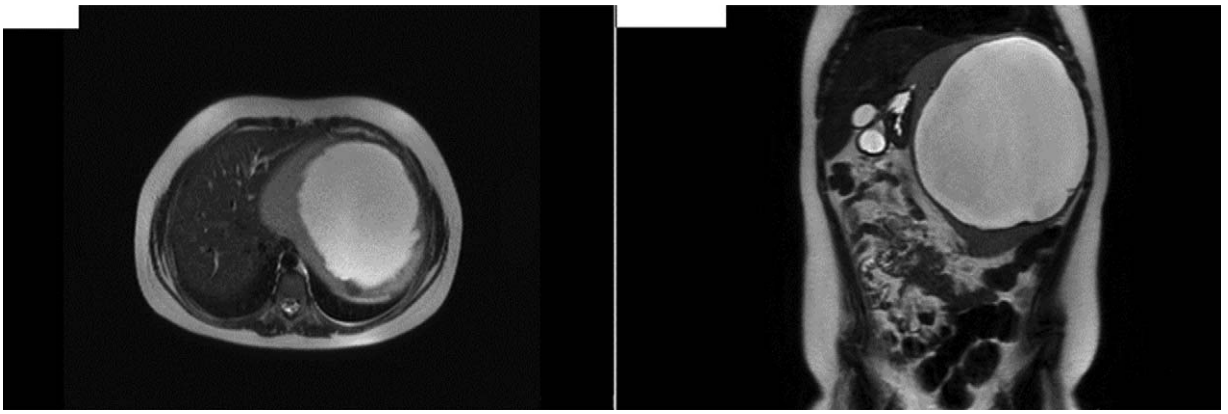

Figure 1. Axial (A) and coronal (B) magnetic resonance imaging of the child abdomen, showing splenomegaly (20 ×15 × 16 cm) with a polylobulated well-defined cystic mass (16.8 × 15 × 15.5 cm) with homogeneous fluid contents, hypointense T1-weighted images and hyperintense T2-weighted images without contrast enhancement.

localized mainly in the left hypochondrium for approximately 24 hours. The patient's medical history was without significant clinical problems. No previous hospitalization was reported.

2.2. Clinical findings

No signs or symptoms of disease were detected during the first clinical examination, with the exception of a significant tenderness to palpation of the abdominal wall, particularly in the left upper quadrant, where a hard mass was detected and protruded approximately 4 cm from the costal arch. The mass had a smooth surface and was mobile with movements associated with breathing. Splenomegaly was diagnosed and laboratory tests were conducted to exclude hematological, autoimmune and infectious diseases associated with the condition.

2.3. Diagnostic focus and assessment

At the same time, an abdominal ultrasonography (US) was performed. All the laboratory test results were negative. Neither anemia nor thrombocytopenia was demonstrated. Moreover, the patient's white blood cell count and morphology excluded blood proliferative disorders. Acute phase reactants (C reactive protein, procalcitonin and erythrocyte sedimentation rate) were in the normal range. Infectious mononucleosis tests were negative. Malaria was excluded. Serological tests for echinococcosis (ELISA and passive hemagglutination test) were negative. While CEA, βHCG and alpha-fetoprotein were negative, CA 125, and CA 19-9 showed high values of 41.6 U/mL (normal range, <5) and 181.4 U/mL (normal range <35), respectively. US showed splenomegaly with a large hypoechoic oval formation with a diameter of 17 cm, largely well-defined margins and the presence of internal fine suspensions spots.

To better define the mass's characteristics, abdominal magnetic resonance imaging (MRI) was performed. It revealed a well-defined SC (16.8 × 15 × 15.5 cm) with a homogeneous fluid content that was hyperintense (T2-weighted images) without contrast enhancement (Fig. 1). The cystic lesion caused major effects on the neighboring organs (left kidney, stomach, and left hepatic lobe), shifting them from their normal sites. The residual splenic parenchyma was significantly compressed and hypo-vascularized. As a collateral finding, the presence of a small accessory spleen was also reported.

2.4. Therapeutic focus and assessment

Considering the mass's volume and the very reduced amount of splenic tissue, an open splenectomy was performed using a xifo-umbilical approach. Figure 2 shows the intraoperative image of the removed spleen. The small accessory spleen was not removed. Upon histopathological examination, the 16 × 14 × 9 cm spleen with parenchyma was largely replaced by a cystic neoformation with a maximum size of 14 × 13 cm. The lesion was characterized by a stratified squamous keratinized thick lining and brownish liquid contents consisting of lymphocytes, erythrocytes, and hemosiderin-rich macrophages. These features informed the diagnosis of a giant epidermoid SC.

2.5. Follow-up and outcomes

No complications occurred in the post-operative period, and blood exams revealed the quick normalization of CA 19.9 and

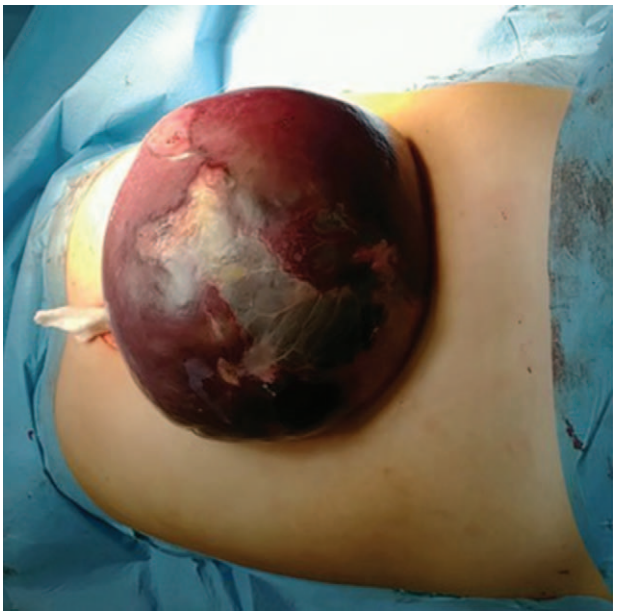

Figure 2. Intraoperative image of the removed spleen.

CA 125 levels. The boy was discharged on the eighth post-operative day. He had already been vaccinated against *Haemophilus influenzae* and Meningococcus C, and had taken the 13-valent pneumococcal conjugate vaccine in early childhood. Therefore, the follow-up vaccine reinforcement program included the 23-valent pneumococcal polysaccharide, meningococcal B and meningococcal A, C, Y, and W vaccines. Furthermore, antibiotic prophylaxis with benzathine penicillin was performed for 1 year after intervention. No complaints were documented during the regular follow-up.

3. Discussion

Epithelial SCs are the most common type of SCs in children.[5] The global incidence of epithelial SCs is 7 per 10,000 as reported in a review of 42,327 autopsies.[6] Females are more frequently diagnosed than males. Histologically, these cysts are differentiated based on the type of epithelial lining, as epidermoid (squamous lining), mesothelial (cuboidal lining), and dermoid (squamous lining with sebaceous glands, hair follicles, and skin appendages). In most cases, congenital SCs remain asymptomatic and are diagnosed incidentally. In some children, such as in the patient described here whose histology revealed a stratified squamous keratinized thick lining, they can become large enough to become symptomatic.

However, the differentiation of epithelial SCs from pseudo-SCs can be difficult. Certain characteristics, such as echogenicity from the debris and calcification, are helpful for identifying pseudo-SCs. For example, in our case, US revealed unilocular anechoic lesions, a smooth cyst wall appearance, and a glistening trabeculated internal lining.[7] Calcification was detected. Via MRI, the cyst was hypointense in the T1-weighted images; in contrast, it was hyperintense in the T2-weighted images, with an intensity signal equal to water. No reinforcement was observed after contrast injection.[7] Similar to the case for our patient, elevated serum and cystic fluid tumor markers, including CA 19-9, CEA, and CA 125, have been described. Interestingly, these markers return to baseline after splenectomy.[8]

Other causes of SCs are very rare, and the diagnosis is generally made based on accompanying signs and symptoms or US and MRI results, not on histological findings.[9] This child had emigrated from Albania only a few months before hospital admission. As Albania is a country where echinococcosis is endemic, it cannot be excluded that the SC the patient was suffering from was of parasitic origin. However, biochemical and serological tests, physical examination and the abdominal US were not consistent with this hypothesis, and echinococcosis was excluded. In this regard, it must be highlighted that echinococcal cysts tend to be unilocularly filled with clear fluid that contains many daughter cysts and brood capsules with protoscolices.[10] Moreover, as no trauma was reported, secondary SC was not considered as a possible diagnosis.

Splenic haemangiomas and hamartomas were excluded by MRI. In haemangiomas, dynamic gadolinium-enhanced imaging demonstrates a progressive centripetal pattern of enhancement that becomes uniform after delayed images.[7] In hamartomas, diffuse heterogeneous enhancement can be demonstrated from images obtained early after administration of contrast material.[7] Moreover, enhancement becomes uniform in the delayed images.

Splenic abscess should be considered in patients with fever, severe abdominal pain, and laboratory tests and a history of an underlying clinical condition favoring severe infections. Abscesses appear as anechoic to hypoechoic, poorly defined lesions via US. High echogenicity is usually related to gas formation.[11] However, CT scan is the best technique to diagnose splenic abscess as it can detect small collections and even small amounts of gas within the abscess.[11]

Another possible diagnosis is primary or secondary lymphoma. In the last case, the signs and symptoms of systemic disease-directed the diagnosis. In primary cases, contrast-enhanced US can be useful, though histology is the best means to confirm diagnosis.[12]

Treatment of SCs, even if asymptomatic, is considered to require surgical resection due to their potential for infection, rupture following trauma, massive hemorrhage, or abscess formation. Open total splenectomy has been considered the treatment of choice. However, spleen removal leads to an increased risk of severe, life-threatening infections. Moreover, compliance with suggested measures to prevent infections frequently produces poor results. Finally, among patients that follow recommendations, the real efficacy of prophylactic measures is not fully observed.[13] For all these reasons, the use of more conservative methods to preserve as much parenchyma as possible has been suggested in recent years. Partial open splenectomy has been proven safe and effective.[14] Moreover, laparoscopic partial techniques or robotic approaches were found to be even more beneficial for patients as they are equally effective as conventional open surgeries but reduce morbidity, complications and patient recovery.[14] However, partial splenectomy is only recommended only when the SC is relatively small and the size of the remaining spleen suggests that adequate immunological function will be maintained. In adults, an SC with a diameter ≤ 5 cm is considered ideal for partial removal. This was not the case for the child described here, for whom total splenectomy was performed due to the large lesion size. Moreover, when partial splenectomy is possible, anatomical variations in the splenic hilar vasculature should be considered. When the main splenic artery is long and divides near the hilum, partial splenectomy is more difficult. Other therapeutic measures such as aspiration of the SC contents, sclerotherapy, and percutaneous drainage are not currently recommended, although they have been found to be effective and long-lasting alternative treatments in some children.[15] Reimplantation of healthy splenic tissue for the preservation of splenic function after splenectomy is a possible effective solution for the future.[16]

In conclusion, this case shows that the differential diagnosis between epithelial mass and SCs of different origin can be difficult, although modern imaging techniques can be useful in establishing pre-operative diagnosis. SCs are rare diseases, particularly in children. In most cases, they are benign congenital epithelial masses that remain asymptomatic throughout the individual's life or are incidentally diagnosed. However, some SCs can cause problems, particularly when they become large enough to compress the neighboring organs or to significantly reduce the functioning splenic parenchyma. Thus, treatment is mandatory in symptomatic cases. Open splenectomy has been the treatment of choice for years. Currently, more conservative methods such as partial splenectomy have been suggested, especially when the SC is relatively small and the residual spleen parenchyma suggests conservation of spleen immunological function. Future studies with adequate follow-up could clarify whether conservative methods are associated with positive long-term outcomes and if they can be used for large SCs.

3.1. Patient's parents perspective

We are very grateful to the pediatricians and surgeons for their diagnostic and therapeutic approach, which was associated with a favorable clinical evolution.

3.2. Informed consent

This case report was approved by the Ethics Committee of the University of Perugia (PED-2018-21) and both parents provided written informed consent for the evaluation of themselves and their child as well as for the publication of this manuscript, including photography. The patient's parents provided their written informed consent for the publication of this study.

Acknowledgment

We would like to thank all the surgeons, pediatricians and nurses involved in the management of this child, as well as the patient's parents.

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