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Case report

Large splenic epithelial cyst: A rare presentation

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ABSTRACT

Introduction and importance: Splenic cysts are infrequent and most of them are related to parasitic diseases. Splenic epithelial cyst (SEC) is the most common type of primary non-parasitic splenic cysts. Its pathogenesis is not yet clear. Splenic cysts are usually asymptomatic and are detected incidentally during imaging exams or an exploratory laparotomy.

Case presentation: Our case is about a primary SEC discovered incidentally. An 18-year-old male patient without any personal and family medical history, presented with symptoms of upper urinary tract infection. Renal ultrasound was performed and found incidentally a solitary cystic lesion in the spleen. A laparoscopic partial splenectomy was made then. The specimen was sent for pathological examination and a diagnosis of primary (epidermoid) epithelial splenic cyst was given.

Clinical discussion: SEC is a rare pathology that could mimic other splenic cysts like hydatid cyst. The preoperative diagnosis of SEC can be suspected by ultrasonography, computed tomography or magnetic resonance imaging. However, histopathological examination is mandatory to confirm the diagnosis. A wide range of differential diagnoses is considered when we suspect clinically SEC because of its lower incidence.

Conclusion: Actually, spleen-preserving surgery with minimally invasive methods such as laparoscopy is the gold standard for the treatment of SEC despite the risk of recurrence encountered with these techniques. However, different surgical modalities are discussed depending on the size and the location of the cyst and the patient's age.

1. Introduction

Cystic lesions of the spleen are rarely seen in routine surgical practice. The true incidence is unknown, however an incidence rate of 0.07% is reported in a review of 42,327 autopsies [1]. According to the causative agent, splenic cysts can be divided into parasitic and non-parasitic cysts. Non-parasitic cysts are classified, based on the presence or absence of cellular epithelial lining, into primary (true) and secondary (false) cysts. Epithelial cyst is the most common type of non-parasitic cysts and constitutes approximately 10% of all splenic cysts [2,3]. SEC is usually asymptomatic and fortuitously discovered. It may nevertheless lead to abdominal discomfort, pain or complications [4]. Various imaging modalities could be used to confirm its cystic nature but the final diagnosis can only be made via pathological examination [5].

The treatment of a non-parasitic splenic cyst is controversial. However partial splenectomy seems to be the most effective one in terms of preserving spleen function and avoiding recurrence [6]. We here report a case of an 18-year-old male with a large SEC discovered incidentally and managed in a multidisciplinary university hospital in Tunisia. This work has been reported in line with the SCARE criteria [7].

2. Case report

An 18-year-old male without any personal and family medical history presented with painful urination, lower abdominal pain and fever. A urine culture was performed confirming urinary tract infection with *Klebsiella pneumoniae*. Abdominal ultrasonography was carried out and revealed a well-defined echogenic splenic cyst, which measured 10.3 cm (Fig. 1, A). Computed tomography confirmed the presence of a solitary multilocular splenic cyst, measuring 10 cm in diameter with parietal calcifications, suggestive of hydatid origin (Fig. 1, B). Abdominal magnetic resonance imaging showed multilocular cyst hyper-intense T1,

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located in the medial portion of spleen measuring 92 × 90 × 80 mm, suggestive of lymphangioma or hamartoma. All routine hematological and biochemical investigations were normal. The indirect hemagglutination (IHA) test for Echinococcus granulosus was negative. The patient received pneumococcal and meningococcal vaccine before surgery. He underwent laparoscopic partial splenectomy by a university assistant in general surgery. The intervention was successful and post-operative follow-up was uneventful. The patient was discharged home three days later. The splenectomy specimen was sent for pathological examination. On gross examination, the specimen measured 12 × 11 × 8 cm. Cut section showed a multilocular cyst of 10 × 8.5 cm with thick white wall, which was filled with brown colored fluid (Fig. 2, A). Microscopic examination revealed a thick fibrous wall lined by squamous epithelium (Fig. 2, B, C, D). Focally, the lining epithelium was flattened and the cyst wall was composed of fibro collagenous tissue with macrophages. Focal parietal calcifications were also present. Based on the above features, the diagnosis of splenic epidermoid cyst was given. The patient is on regular follow up and is doing very well one year after surgery with no evidence of recurrence.

3. Discussion

Splenic cysts are rare and epithelial ones are more infrequent [8]. Since the first splenic cyst reported in 1929 by Andral, the classification of these lesions has evolved into the present system [9]. The two main principal classifications made by Fowler and Martin are based on the presence or absence of cyst epithelium lining [10]. Primary (True) cysts with epithelial lining while secondary (False) cysts are without epithelial lining. True cysts can be further classified into parasitic and non-parasitic.

False splenic cysts account for 80% of all splenic cysts and usually occur because of trauma, disorganized hematoma, splenic infarction, intrasplenic pancreatic pseudo cyst, and splenic abscess. True splenic cysts represent only 20% of splenic cysts and can be neoplastic in origin (hemangioma and lymphangioma), infectious (mostly hydatid disease), or congenital (primary epithelial cysts) [11].

In 2002, Morgenstern with series of 23 cases propose a new classification based on a unified theory of the true pathogenesis cyst. This classification subdivides non parasitic splenic cysts into congenital, neoplastic, traumatic and degenerative [12].

Parasitic cysts are the most common splenic cysts especially in endemic areas, such as Africa, South America and Australia and most of them are caused by Echinococcus granulosus [6]. Since we are in endemic country, the main differential preoperative diagnosis was hydatid cyst.

Echinococcosis detection test is positive in approximately 90% of patients with hydatid cysts and the definitive diagnosis is made only by histopathology [13].

Epithelial cyst is the most common type of non-parasitic cysts and constitutes approximately 10% of all splenic cysts [2,3]. It is common in second and third decades of life with slight female predilection, but it can occur at any age. Most of them are asymptomatic clinically or they can manifest by mild pain and palpable mass in the left upper abdomen [14]. Very few cases present with complications like acute abdomen due to intracystic bleeding, infection or rupture [15,16]. They can manifest also with hypersplenism or elevated systemic blood pressure due to renal artery compression [7,17]. Most of primary cysts are diagnosed incidentally, and their prevalence is increased because of the increased use of non-invasive imaging techniques [9]. On ultrasonography, epithelial cysts appear usually as well-defined, thin-walled anechogenic and unilocular or multilocular lesions. They may contain echogenic debris in cases of intracystic bleeding or infection and the echogenicity may increase depending on these complications. On Computed tomography, they appear as rounded and well-demarcated lesions. However, all of these diagnostic modalities are unable to differentiate the splenic epithelial cyst from other splenic cysts [17].

Histologically, epithelial cysts are classified basing on the type of epithelial lining into epidermoid cyst with stratified squamous epithelial lining, mesothelial cyst with cuboidal epithelial lining and dermoid cyst lined by squamous epithelium with dermal appendages, hair follicles and sebaceous glands [1]. Immunohistochemical study shows that epithelial lining in epidermoid cysts is positive for cytokeratin and negative for endothelial markers and calretinin. Some cases demonstrate that epidermoid cysts are immunoreactive for carcinoembryonic antigen (CEA) and carbohydrate antigen 19-9 (CA19-9), which may be elevated in blood serum and usually return to normal levels after removal of the cyst [11,14,18]. The histogenesis of epithelial cysts is still controversial, Burrig [19] and Ough et al. [20] suggested that these cysts originated from invagination of capsular peritoneal mesothelium or collection of peritoneal mesothelial cells trapped in splenic sulci, which subsequently undergoes squamous metaplasia due to chronic irritation. Furthermore, other studies suggested that epithelial cysts have a teratomatous derivation or inclusion of fetal squamous lining instead of metaplasia [5], while others considered splenic epithelial cysts as a result of secondary spontaneous intra-splenic bleeding or collection of fluid after injury [15,21].

Management of non-parasitic splenic cysts is controversial and there is a limited data to determine the appropriate time to interfere [6]. In the past, it has been recommended to treat symptomatic cysts, or when they are larger than 5 cm due to an increased risk of spontaneous rupture or infection. However, some authors reported that small cysts may be also at risk of rupture after a simple trauma [8]. Surgical modalities are multiple for the treatment of these cysts and the choice of the suitable one vary depending on the size, number, pathogenesis, relation with splenic hilar vessels and parenchyma, the amount of remaining healthy splenic tissue and patient's age [22]. Total splenectomy has been the preferred treatment for years in order to decrease the risk of bleeding and other complications of the cyst. Currently, more conservative

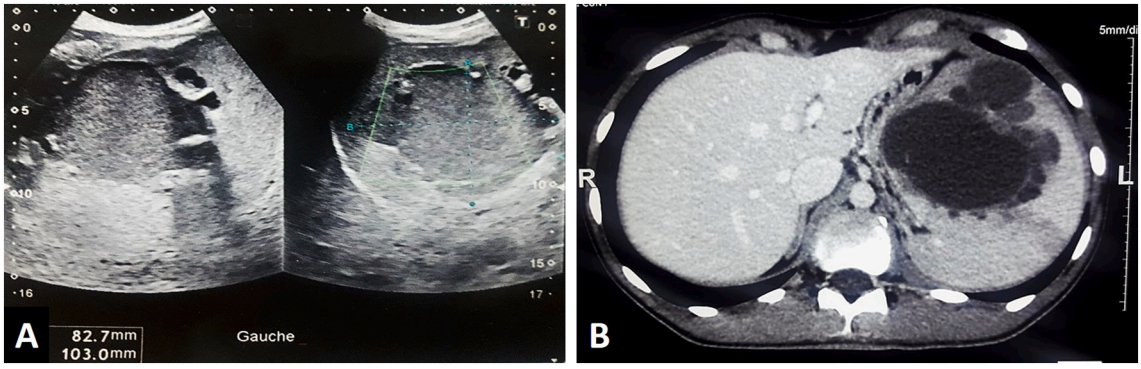

Fig. 1. A: Abdominal ultrasound showing a well-defined echogenic splenic cyst measuring 103 × 82.7 mm. B: Axial computed tomography image of the abdomen showing multilocular splenic hypodense cyst, measuring 10 cm in diameter with parietal calcifications, suggestive of hydatid origin.

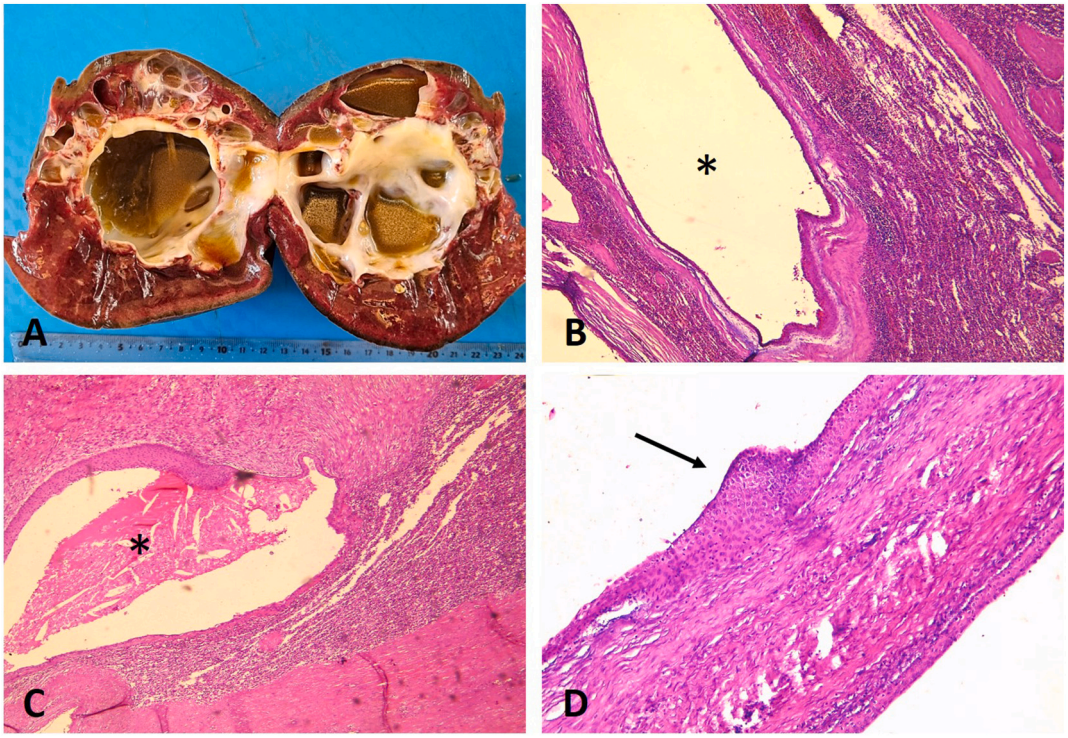

Fig. 2. A: Gross examination of the specimen showing at cut section a multilocular cyst within the spleen with thick white wall. B, C: Histological findings showing a splenic multilocular epithelial cyst (*black asterisk*) (HE ×4). D: The cyst has a thick fibrous wall that is lined by squamous epithelium (*black arrow*) (HE ×10).

methods have been suggested, such as partial splenectomy with the awareness of immunologic role of the spleen, particularly in young age and because of the increased risk of post splenectomy infection [23]. Some authors reported other conservative methods like laparoscopic decapsulation of congenital splenic cysts, and it was demonstrated that it could be an adequate and safe surgical option in selected patients. Termos et al. reported in a review of literature that 10 of 11 cases treated by decapsulation were managed successfully and only one has a post-operative complication [11].

4. Conclusion

Epithelial cyst is the most common type of non-parasitic cysts that is difficult to diagnose preoperatively and is usually diagnosed fortuitously. Imaging can facilitate or guide the diagnosis. However, careful histopathological interpretation along with immunostaining is important to lead to correct diagnosis. Today the optimal treatment option is partial splenectomy with minimally invasive methods such as laparoscopy is the gold standard for the treatment of SEC in order to avoid complications.

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Ethical approval

This study is exempt from ethical approval at our institution.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author contribution

Seifeddine Ben Hammouda and Samiha Mabrouk: Writing - original draft. Ahlem Bellalah, and Mezri Maatouk: Reviewing and Editing. Abdelfatteh Zakhama and Leila Njim: Investigation, Visualization.

Registration of research studies

Not applicable.

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Declaration of competing interest

None.

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