American Journal of Case Reports

e-ISSN 1941-5923
© Am J Case Rep, 2023; 24: e941585
DOI: 10.12659/AJCR.941585

Received: 2023.06.26
Accepted: 2023.08.29
Available online: 2023.11.02
Published: 2023.12.10

A Large Splenic Epidermoid Cyst Initially Misdiagnosed as a Hydatid Cyst

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Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
Literature Search F
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Financial support: None declared
Conflict of interest: None declared

Patient: Male, 22-year-old
Final Diagnosis: Splenic epidermoid cyst
Symptoms: Abdominal discomfort • abdominal distension
Clinical Procedure: —
Specialty: Gastroenterology and Hepatology

Objective: Unusual clinical course

Background: Splenic cysts are classified as either parasitic or non-parasitic cysts, with both types sharing similarities in their clinical presentation and imaging findings. Most splenic cysts are of parasitic origin, while non-parasitic splenic cysts are less common. Splenic epidermoid cysts (SECs) are a rare subtype of non-parasitic cysts and compromise only around 10% of them.

Case Report: In this paper we present a case of 22-year-old man with no significant past clinical history, who presented with non-specific, vague symptoms, including persistent left upper-quadrant pain and discomfort for the last 2 years. A physical examination and extensive laboratory tests were inconclusive. Subsequently, the patient underwent multiple imaging studies including ultrasonography and computed tomography (CT) scan of the abdomen. His ultrasonographic findings were consistent with the diagnosis of hydatid cyst, which was further emphasized by its frequent occurrence in clinical practice, as our country is considered an endemic region. In light of this, he underwent laparoscopic splenectomy following percutaneous cyst drainage. The consequent histopathological examination revealed the diagnosis of splenic epidermoid cysts.

Conclusions: When encountering splenic cysts in regions where parasitic infections are endemic, special attention is needed, as physical examination, laboratory tests, and imaging studies alone are insufficient to differentiate among the types of cysts. Histopathological examination remains the diagnostic tool of choice, particularly when imaging findings are inconclusive. Splenectomy, with either a laparoscopic or open approach, is the treatment of choice for splenic cysts to prevent recurrence as well as other potential catastrophic complications.

Keywords: Echinococcosis • Splenic Neoplasms • Ultrasonography, Doppler • Drainage • Splenectomy

Full-text PDF: https://www.amjcaserep.com/abstract/index/idArt/941585

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Background

Cystic lesions of the spleen are rarely encountered in clinical practice. The incidence of splenic cysts is yet to be determined, and the incidence of all splenic cysts was estimated to be around 0.07% of 43 000 autopsies in a review [1]. Splenic cysts can be divided into primary and secondary based on the presence of epithelial lining. Moreover, primary cysts were further divided into parasitic and non-parasitic splenic cysts (NPSCs), with the former being the most common [2].

Splenic epidermoid cysts (SECs), which compromise only around 10% of all NPSCs, are even rarer [2]. These lesions are discovered incidentally after patients present with non-specific symptoms such as left upper-quadrant discomfort and fullness [3]. The diagnosis of SECs cannot be done solely based on the clinical presentation and imaging findings, as histopathological examination remains the definitive diagnostic modality [4]. In regard to management, surgical splenectomy is considered the mainstay of treatment to minimize recurrence and complications such as bleeding or infection [3].

Herein, we report a case of 22-year-old patient who presented with a 2-year history of abdominal pain with a presumptive diagnosis of splenic hydatid cyst that was managed conservatively, which turned out to be splenic epidermoid cyst upon histopathological examination.

Case Report

A 22-year-old male patient, who was a smoker with a non-significant medical history apart from septoplasty, lipoma excision, and contrast allergy, presented to the clinic with abdominal pain in the left upper-quadrant region (LUQ) for 2 years. It was sudden in onset, intermittent (1 episode per week, each episode lasting 30 minutes), and colicky. Nevertheless, no radiation, exacerbating, or relieving factors were identified. He sought medical care and underwent investigations, outside our institution, for the pain 1 year ago, which revealed the presence of a large splenic hydatid cyst that was managed only conservatively at that time. Furthermore, he had a new, sudden-onset right upper-quadrant (RUQ) pain that started 6 months ago, with 3 episodes per week (each lasting 3-4 hours), radiating to the right shoulder and back, and increased by eating fatty meals. Upon detailed review, he denied any other gastrointestinal or other systematic symptoms such as nausea, vomiting, diarrhea, fever, or other symptoms.

On examination, the patient was looking well and was vitally stable. He was conscious, alert, and oriented to time, place, and person, with a Glasgow coma scale of 15/15. The systemic exam was unremarkable, and an abdominal examination revealed the presence of a non-tender LUQ mass without RUQ tenderness. Importantly, the results of all laboratory tests were negative.

He was admitted to the hospital for further evaluation and underwent an abdominal and pelvis contrasted computed tomography (CT) scan, which confirmed the presence of splenomegaly and multiple large splenic cysts that were rounded and lobulated, with the largest measuring 12×11.3 cm, findings suggestive of hydatid cysts. A large gallbladder stone, as well as numerous bilateral small renal cysts, were also noted (**Figures 1-3**).

Laparoscopic cholecystectomy and splenectomy (following percutaneous ultrasound-guided cysts drainage) was done after discussing the treatment options with the patient, followed by receiving the required vaccines. Fortunately, the surgery was uneventful, and no complications evolved postoperatively apart from a slight elevation in white blood cells (WBCs) and C-reactive protein (CRP), which came back to normal as he was covered with the appropriate antibiotics. He was discharged and planned for follow-up visits.

The histopathological sections that were done on the splenic cysts revealed, grossly, the parenchyma of the spleen contained multiple cysts of variable sizes ranging from 0.7 to 9.5 cm in diameter. The inner surface of the cysts was glistening white in areas, hard and calcified in some, and irregular in others (**Figure 4**). Histopathologic examination revealed multiple cystic spaces, some of which had denuded and calcified lining while other areas were focally lined by keratinizing stratified squamous epithelium, resembling the epidermis of the skin (**Figure 5**). The previously described findings were negative for malignancy as well as strongly suggesting the diagnosis of multiple epidermoid cysts rather than hydatid cysts.

Discussion

Splenic cysts are considered a disease of rare occurrence. They were originally classified by Fowler and modified by Martin based on the presence of epithelial lining, in which they divided the cysts into primary (also known as true) cysts or secondary (also known as false or pseudo) cysts. Furthermore, the primary cysts were divided into parasitic and NPSCs. Secondary cysts compromise 75% of all splenic cysts, with trauma being the most common cause [2]. Nevertheless, primary cysts are even less prevalent in the literature, with the parasitic variants occupying a larger proportion of them, while splenic epidermoid cysts (SECs) compromise the majority of NPSCs and only around 10% of all splenic cysts [5].

However, a more recent study done by Morgenstern on 23 splenic cysts cases suggested that the aforementioned

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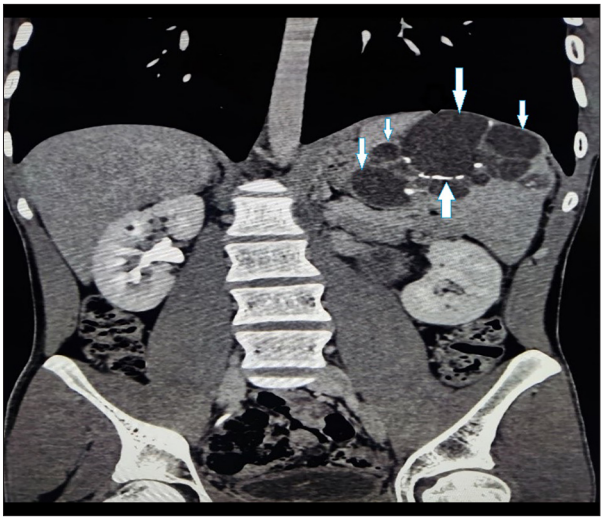

Figure 1. Multiple, round, lobulated, fluid-density (20-40 HU) cystic lesions, most of them showing wall calcifications seen at the spleen, and multiple, small, round cystic lesions seen at the bilateral kidneys.

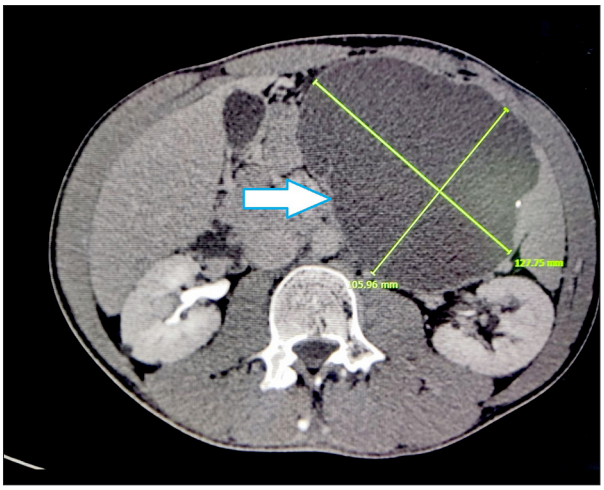

Figure 2. Well-encapsulated, round, lobulated, fluid-density (20-40 HU) cystic lesion in the spleen, measuring 12.7×10.5 cm.

classification is misleading, as the epithelial lining alone is not the defining feature. He suggested a new classification system based on the true pathogenesis of cysts. This classification divides the NPSCs into congenital, traumatic, neoplastic, and degenerative [3].

Congenital epithelial splenic cysts have a characteristic gross appearance, with the interior of the cyst being white or grayish in color with coarse fibrous trabeculations, the presence of epithelial lining, and the absence of preceding trauma history. In addition, the epithelial lining can be epidermoid (most common), mesothelial, transitional, or a mixture of them [1,3]. This is in accordance with our case, as the patient did not have

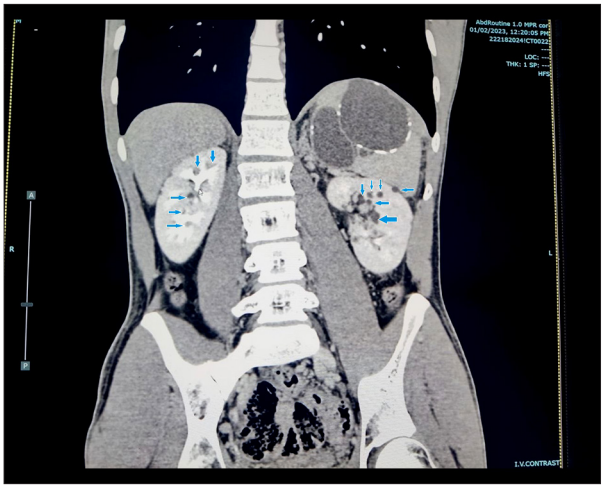

Figure 3. Multiple small, round, lobulated, cystic lesions seen at bilateral kidneys. In addition, there were 2 circumscribed cystic lesions with wall calcifications seen at the spleen.

a history of trauma, and had the characteristic gross and histological findings. Moreover, he had concomitant renal cysts, which may support a congenital cause.

SECs are usually diagnosed in adolescents and young adults, with a predilection for females. The cysts can be single or multiple and can be uni- or multi-locular [2,5]. Although most patients are asymptomatic, left upper-quadrant discomfort and fullness are the most common presenting symptoms. Other symptoms, such as referred shoulder pain and splenomegaly, can occur [3,5]. This is consistent with our case, as the patient was in the typical age group, with left upper-quadrant pain and splenomegaly. Splenomegaly is always evident when the cyst size exceeds 6 cm [3].

The benign nature of the epidermoid cyst makes the diagnosis incidental in most cases during ultrasonography (USG) [5,6]. On USG, the presence of cystic mass with or without septations warrants further workup to differentiate between infectious, neoplastic, and congenital cysts [6]. Serological workup and tumor markers are essential to exclude parasitic etiologies [7], especially in the epidemic regions, and neoplasms [8], respectively. Further imaging techniques, including computed tomography (CT) or magnetic resonance imaging (MRI) give more information about cyst morphology, components, exact location, and relationship to the adjacent structures, which is needed to guide the treatment decision [6].

The definitive diagnosis of SEC is not straightforward, and needs histopathological examination with certain immunohistochemical analysis, as none of the imaging findings or blood tests are pathognomonic. The presence of keratinizing stratified squamous epithelium encapsulated by a fibro-collagenous

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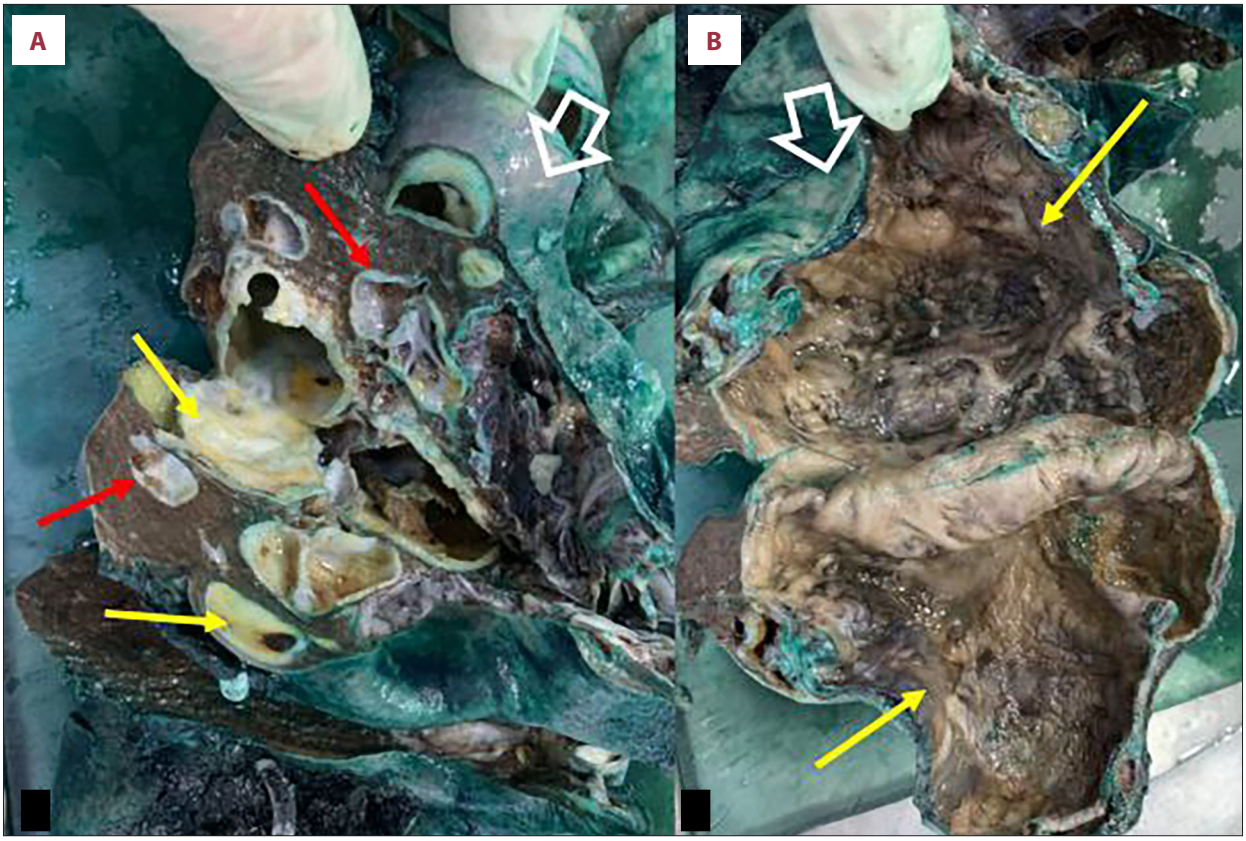

Figure 4. (**A**) The outer capsule of the spleen shown here is inked green (empty white arrow) and the cut sections reveal multiple cysts of variable sizes present within the splenic parenchyma, with glistening, white, shiny lining (red arrows) and calcified hard areas (yellow areas). Superior view. (**B**) The capsule of the spleen is shown again (empty white arrow) and the largest cyst within the spleen is opened, revealing a denuded, coarse, fibrotic surface (yellow arrows). Superior view.

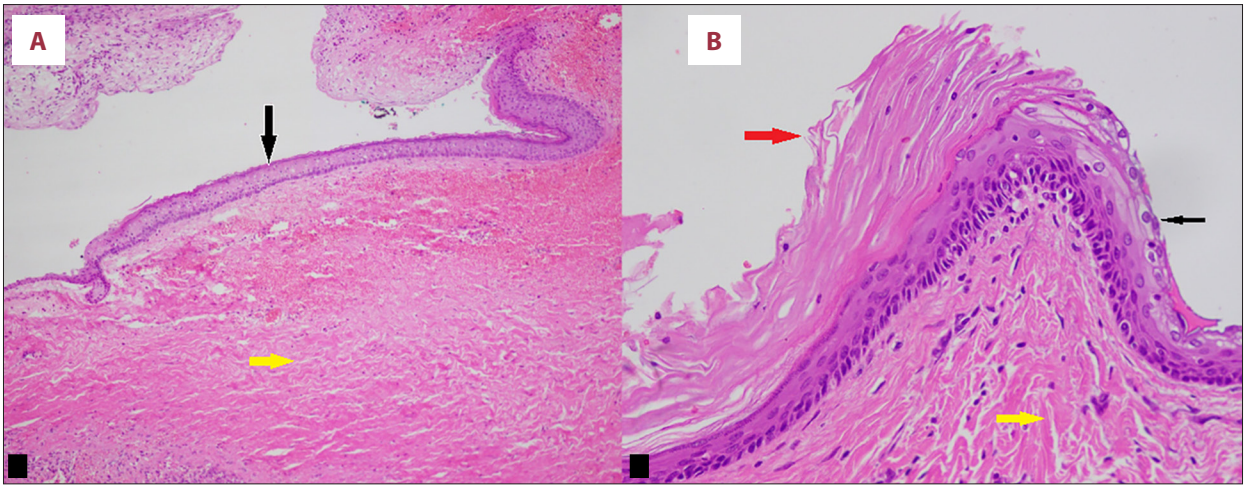

Figure 5. (**A**) The lining of some of the cysts is composed of keratinizing squamous epithelium resembling the epidermis of the skin with the red arrow pointing to the keratin layer, the black arrow pointing to the multilayered squamous epithelium and the yellow arrow pointing at the collagen bundles of the underlying fibrous tissue. Original magnification, 400× (Hematoxylin and eosin stain). (**B**) Lower magnification of the cyst wall shows the stratified squamous lining (black arrow) and the underlying fibrous connective tissue (yellow arrow). Original magnification, 100× (hematoxylin and eosin stain).

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wall that is positive for cytokeratin immunohistochemical stain is consistent with the diagnosis of splenic epidermoid cyst [1]. Given the non-specific USG findings and the high pretest probability of hydatid cyst in Jordan, our case was highly consistent with the diagnosis of hydatid cyst [9] until the definitive diagnosis of the epidermoid cyst was made based on the histopathological findings postoperatively.

Splenic cysts may be complicated by infection, bleeding, or/and rapture if untreated or mistreated, which mandates their removal [3,6]. However, given the scarcity of cases in the literature, there is no clear evidence or specific criteria for the management of SEC. Accordingly, the management of splenic cysts varies depending on their size, location, and surgeon skills. Consequently, and as a rule, a minimally invasive splenectomy should always be adopted to alleviate the symptoms and decreases the risk of recurrence [3,5,10]. In addition, our patient had concurrent gallbladder stones that warranted cholecystectomy, which supported the surgical management. Percutaneous drainage of the cyst is associated with higher recurrence rates, especially when used alone. However, using it as a bridging technique before cyst removal seems to decrease the cyst size, and makes the spleen accessible for laparoscopic removal instead of the open approach [11]. Our patient had a large cyst that hindered total splenectomy via laparoscope, so he was bridged successfully for laparoscopic excision via percutaneous drainage.

Generally, the prognosis is good, and patients should receive the appropriate vaccination and follow-up for recurrences, which varies depending on the surgical approach [12] and the histopathological findings [8], especially if complete removal was not achieved [6].

Conclusions

We report a case of a SEC that was diagnosed histopathologically following the presumptive diagnosis of hydatid cyst. Special attention is needed in areas endemic for parasitic infections, as preoperative imaging studies alone are insufficient to differentiate among the types of cysts and postoperative histopathological examination remains an essential part of diagnosis. Splenectomy, with either a laparoscopic or open approach, is the treatment of choice to prevent recurrence and potential catastrophic complications.

Acknowledgment

The authors give special thanks the patient and his family, as well as the medical staff, for their contribution to the case report.

Declaration of Figures' Authenticity

All figures submitted have been created by the authors who confirm that the images are original with no duplication and have not been previously published in whole or in part.

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