

Splenic cysts: clues in sonographic differential diagnosis and a new role for twinkling artifact

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Abstract

Aim: Our aim was to investigate the frequency of various splenic cysts, to define the sonographic differential diagnostic clues and to introduce the value of twinkling artefact in the diagnosis of epidermoid splenic cysts. **Material and methods:** All the splenic cysts imaged by ultrasound in 3 university hospitals during the period of 2005 to 2022 were recorded, followed-up and analyzed. **Results:** One hundred seventy-one patients with splenic cysts were detected and these were classified and 73% of the cysts were simple. Ten cysts were epidermoid cysts as proven by post-operative final histology. **Conclusion:** Cystic splenic lesions are rare. Most of them are small simple cysts. Epidermoid cysts are larger in volume, constitute 6% of the total and can be differentiated by the help of twinkling artefact by Doppler ultrasound.

Keywords: spleen cyst; epidermoid cyst; ultrasound; Doppler; twinkling artifact

Introduction

Splenic lesions are rare and mostly incidental. Cystic splenic lesions include true congenital cysts, pseudocysts, hydatid cysts, abscesses, infarcts, cystic neoplasms, and aneurysms.

True congenital epithelial cysts can basically be classified as epidermoid cysts, mesothelial cysts, and endothelial cysts [1-4]. However, there is no clear-cut consensus on the classification; some experts believe that all three cysts are variations of epithelial cysts, whereas some others suggest that actually there are variations based on the embryological origins, internal layers, and their fluid contents [5,6]. Especially, the epidermoid and mesothelial cysts are regarded as very similar or the same

entity, and constitute the true non-neoplastic congenital cysts, whereas dermoid cysts of the spleen are extremely rare and endothelial cysts (cystic lymphangiomas and hemangiomas) are usually considered as neoplastic [1,2,7,8]. It is usually accepted that true epithelial cysts originate from invagination of capsular peritoneal mesothelium [1,3].

Differential diagnosis is very important in the management of cystic splenic lesions. Ultrasound (US) and color Doppler mode can give clues that are very helpful in the differential diagnosis. Splenic abscesses, infarcts, cystic neoplasms, and aneurysms can usually be differentiated easily by either accompanying symptoms or other Doppler features and by follow-up findings. Contrast-enhanced US, CT, or MRI can help in better defining and describing some additional features or the perisplenic relations of the lesions.

In this study, we present the differential diagnostic clues for splenic cystic lesions that we have experienced in the last 7 years in 3 large university hospitals. Among the differential diagnostic clues, we have observed that twinkling artifacts within the cyst, were almost exclusively seen in epidermoid-type cysts and can be introduced as a new pathognomonic sonographic sign.

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Materials and methods

During the period 2005-2022, all splenic epidermoid cysts were prospectively recorded and followed up at Akdeniz University Hospital. The striking sonographic twinkling artifacts in some cystic lesions which proved to be an epidermoid cyst, were realized first in 2005, and this feature was searched in any cystic splenic case, thereafter. Between 2015-2022, all epidermoid splenic cysts were retrospectively searched in the PACS of two other university hospitals (Istanbul Medeniyet University Hospital and Koç University Hospital). Retrospectively, the splenic cysts were divided into 5 groups: epidermoid cysts, simple epithelial cysts, hydatid cysts, pseudocysts, and indeterminate cysts. Only cysts were included and cystic appearing abscesses, infarcts, and neoplastic lesions (such as cystic lymphangiomas and aneurysms) were excluded. All available US, CT, and MRI of the patients with a cyst in the spleen were evaluated. Since it was technically not possible to count all the splenic cysts in all three hospitals for a period of 17 years, the annual prevalence was calculated based on the total annual records of the 3 hospitals. It was possible to record all the splenic cysts in two institutions for a period of 7 years and for the last year in the largest university hospital. We received research protocol approval from our institutional Review Board (IRB) with a number of 2022.198. IRB1.075 by HD.

US exams were performed with Applio 500 (Canon) or Logic E9 (GE) machines. Although usually convex probes (1-6 MHz) were initially used, linear transducers (5-14 MHz) were also added to see more details in some cases. The dimensions, the wall thickness, internal echoes, septations, any mural nodules or solid components were recorded for all cystic splenic lesions. Doppler mode imaging was also an essential part of the exam. The solid components, the wall, and the septa were scanned in Doppler mode to detect any blood flow. Twinkling artifacts accompanying the internal echoes were also recorded.

Statistical analysis

Only descriptive statistics were performed.

Results

Total cases and distribution

The retrospective search revealed 171 patients with splenic cysts. Since there were many uncountable tiny cysts in some patients, they were not counted separately, but counted on patient/case base. The number and percentage of cysts were as; 125 simple cysts (73%), 10 epidermoid cysts (6%), 12 hydatid cysts (7%), and 10 pseu-

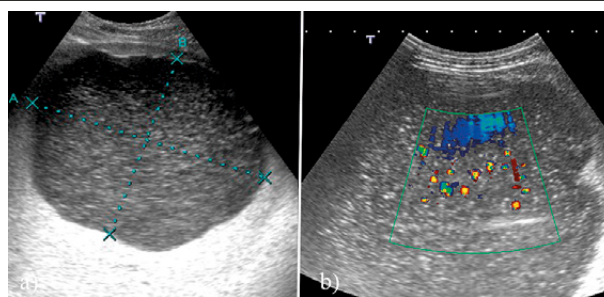


Fig 1. A 8-year-old boy with a huge 14x11 cm cystic splenic mass: a) B-mode appearance shows intense internal echoes; b) In Doppler mode, the twinkling artifacts are striking.

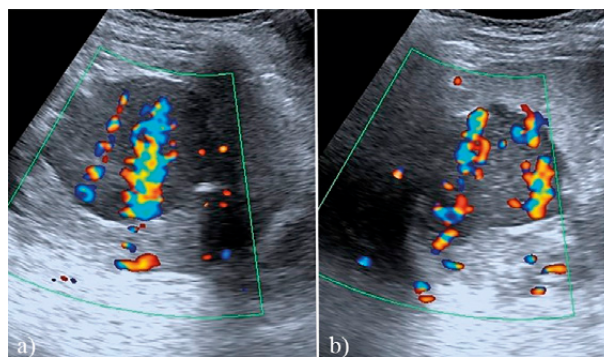


Fig 2. A 17-year-old female with an epidermoid cyst of 64 mm in diameter: a) Twinkling artifacts are seen in Doppler mode with a PRF of 3.5 k; b) With a higher PRF, more twinkling artifacts are apparent.

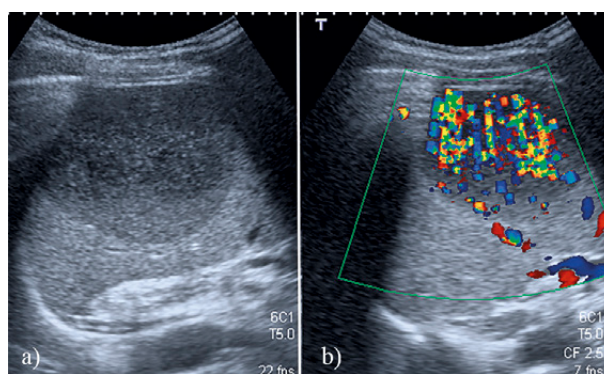


Fig 3. A 14-year-old girl with an epidermoid cyst: a) B-mode shows a 65 mm cyst with intense internal echoes; b) Color Doppler mode shows twinkling artifacts.

docysts (6%). In 14 cases (8%) the cysts were accepted as indeterminate, 12 of them probably epidermoid and 2 probably pseudocyst.

Epidermoid Cysts

A total of 22 splenic cysts in 3 hospitals, had features typical for epidermoid cysts. However, only 10 of them were pathologically proven (fig 1-4). The remaining 12 cysts were either smaller than 5 cm or the patients refused surgery. Therefore, they were categorized as inde-

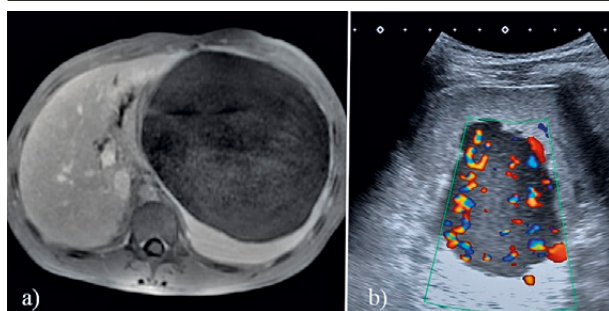


Fig 4. a) Pre-op images of a giant (16 cm) epidermoid cyst in a 15-year-old boy, Axial contrast enhanced T1 image; b) After 6 months of hemisplenectomy and cyst aspiration, US shows a recurrent 6 cm cavity with twinkling artifacts.

terminate/probably epidermoid. The biggest epidermoid cyst had a diameter of 144 mm. The smallest measured 41 mm, and the average diameter was 92 mm. All epidermoid cysts showed intense twinkling artifacts in Doppler mode; the artifacts were more prominent with increased PRF values.

Simple epithelial cysts

Simple epithelial cysts actually formed the largest group comprising 73%. In the first center (Akdeniz University Hospital) 48 simple cysts were counted among 5,026 abdominal CT/MRI cases. The prevalence was 0.0096. In the second center (Medeniyet University Hospital) 40 simple cysts were recorded among 4,092 abdominal CT/MRI cases with a prevalence of 0.0098. The third center (Koc University Hospital) had 37 simple cysts in a combined 21,342 abdominal US/CT/MRI cases. The prevalence here was 0.0017.

Average diameter of the simple cysts was 17 mm (range: 5 mm-77 mm). Interestingly, 27 cases among the total 125 cases had multiple cysts (22%). They usually stayed stable and some even disappeared in follow-ups (fig 5, fig 6). Few of them enlarged. Only one case was associated with autosomal dominant polycystic kidney disease, and a few were with Gorham Disease or Lymphangiomatosis.

Hydatid Cysts

Hydatid (Echinococcal) cysts were not rare. Especially in the Mediterranean region, the disease is endemic in some mountain villages. Therefore, hydatid cases were mostly seen in Akdeniz University, which is the largest university hospital covering a wide range of Mediterranean area. A total of 15 hydatid cysts in 12 patients were detected. Three patients each had two cysts, and all the others had single cysts. The cyst diameters ranged between 17-100 mm (average: 48 mm). The distribution of the cysts according to Gharbi classification was as follows: 4 cysts Gharbi type 1, 3 cysts Gharbi type 2, 1 cyst Gharbi type 3, 5 cysts Gharbi type 4, and 2 cysts Gharbi



Fig 5. Sixty-nine-year-old woman with an incidental simple cyst: a) The cyst was detected in February 2017 by US and measured 16 mm; b) Follow-up coronal CT image in May 2022 shows that it is 16 mm; stable within the last 5 years.

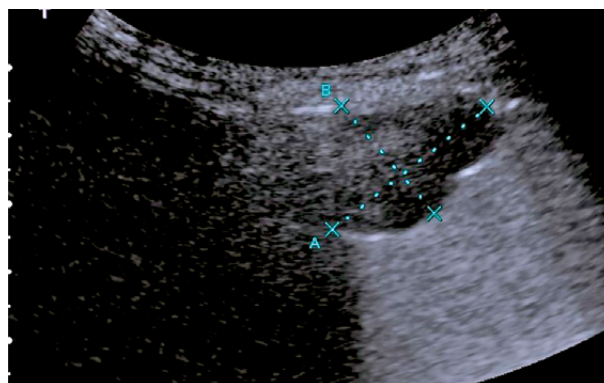


Fig 6. Five-year-old boy with a vanishing 22x14 mm simple cyst. In 2014, the lobulated cyst measured 30x21 mm, slightly larger compared to the examination 3 years before. In 2019 and 2020, after 8 and 9 years the cyst could not be detected.

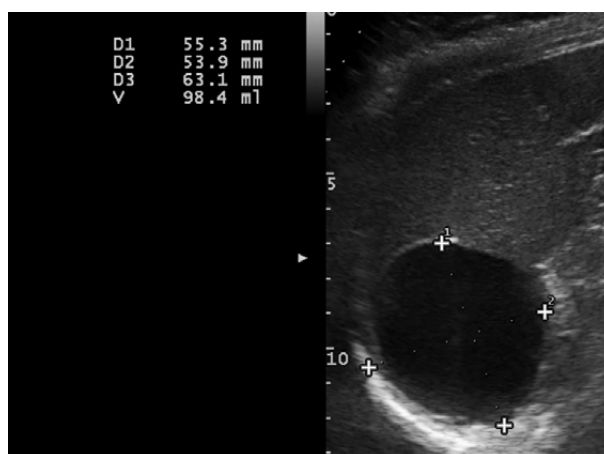


Fig 7. Sixty-year-old woman with a thick-walled Gharbi type 1 hydatid cyst; the cyst has a volume of 98 cm³.

type 5 (fig 7-10). One of the patients with multiple liver and spleen cysts who was treated by PAIR was proved to have an alveolar type hydatid cyst (fig 11).

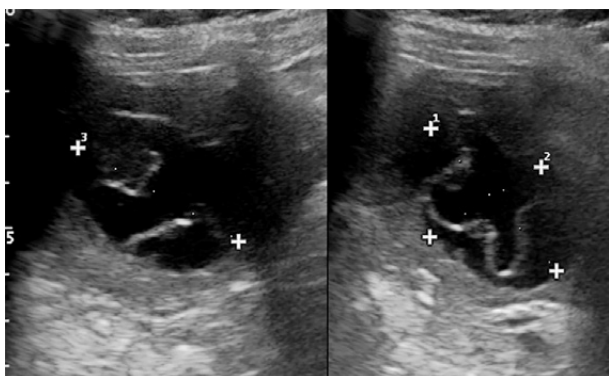


Fig 8. Eleven-year-old boy who initially had surgery for a lung hydatid cyst and then simultaneously had percutaneous treatment for liver and spleen cysts. Gharbi type 2 spleen cyst measures 42 mm.

Pseudocysts

There were 10 pseudocysts and 2 indeterminate/probably pseudocysts (fig 12). Since the patients' trauma history and its relation to the cyst is not always clear, their proof was difficult. They had varying degrees of wall calcification.

Discussion

Epidermoid cysts and twinkling artifact

In this study, our primary purpose was to describe a new observation in the differential diagnosis of splenic cysts. Our initial observation that epidermoid cysts may have internal echoes that give strong twinkling artifacts on color Doppler mode was confirmed with the following cases that had pathological proof after surgery or percutaneous treatments. During our study period, an interesting paper about dermatological epidermoid cysts, first reported the twinkling artifacts as a pathognomonic sign [9]. Twinkling artifact is well known as an artifact

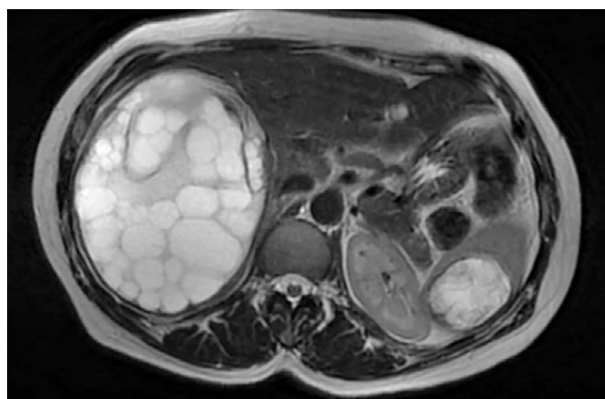


Fig 9. MR image of a 56-year-old woman with Gharbi type 3 hydatid cysts both in the liver and the spleen.

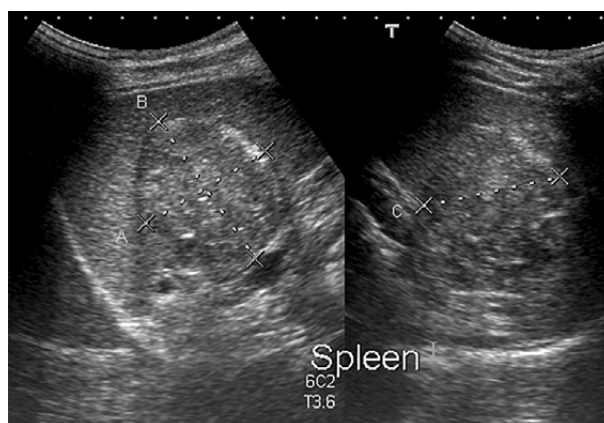


Fig 10. Seventeen-year-old girl with a Gharbi type 4 cyst of 52 mm. She has been using albendazole for the last 8 years because she has multiple liver and spleen hydatid cysts.

behind strong reflector surfaces such as gas-air, stones, and some metallic objects. It has been helpful in daily US practice by pointing out small stones-calcifications and other foreign materials and metallic objects, some

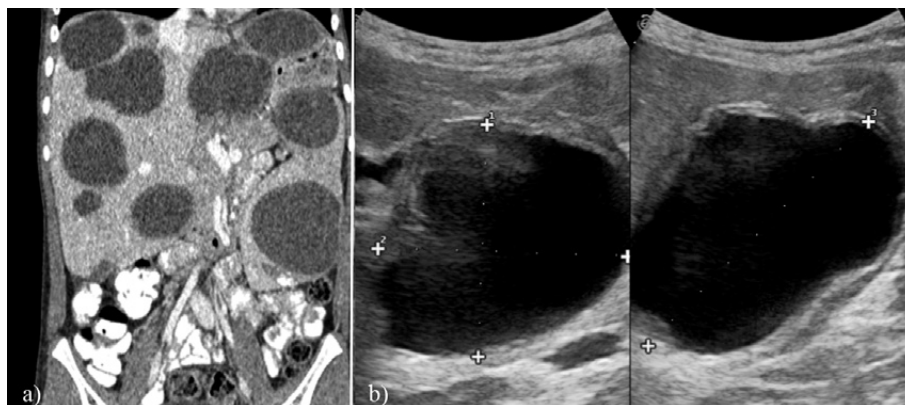


Fig 11. a) Multiple liver and spleen cysts in a 15-year-old girl on CT and b) ultrasound. After percutaneous treatments, they were proved to be echinococcus alveolaris.

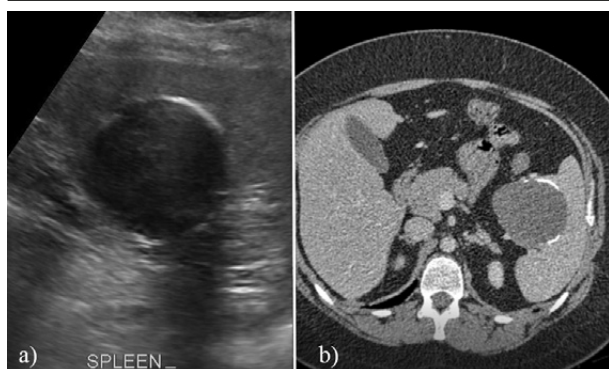


Fig 12. A probable splenic pseudocyst in a 27-year-old woman: a) and b) US and CT shows a 6 cm cyst with partially calcified walls.

of which are not easily shown by gray scale images. The twinkling artifact in epidermoid cysts are caused by the keratin substance [9].

Prevalence and distribution of splenic cysts

Our secondary purpose was to enhance our understanding of splenic cystic lesions, which are quite rare. We did this by collecting as much as possible data about splenic cysts from 3 big hospitals over a long period. Because of the rarity and the seldom need for surgery, our knowledge remains limited about splenic cysts. The incidence of splenic cysts in an autopsy study of 42,327 cases, was found to be 0.0007% [10]. The prevalence of simple cysts in our study was similarly low (between 0.0017 % and 0.0098 %).

It is often thought and cited that the most frequent cystic splenic lesion is a pseudocyst [2,11]. In this study, we have certainly realized that this is not true. Actually, the new era of widespread high-tech CT/MRI use with decreased slice thickness, enhances the chances of revealing subcentimeter simple cysts. Of the 171 patients, 73% had small simple epithelial cysts in our series. Epidermoid cysts, hydatid cysts and pseudocysts had almost equal incidences with 6-7 %. In the autopsy series, similar to our study, 12 of the 15 cysts were less than 2 cm, and 4 cases had multiple cysts (27 %) [10]. In our study, the average diameter of the biggest cysts was 17 mm and 22% of cases had multiple simple cysts. The prevalence was lower in the third center because US records were also considered. Probably CT and MRI are more sensitive in the detection of small simple cysts that may be overlooked by US, because of the shadowing ribs.

Differential Diagnosis

Simple epithelial cysts are usually small with a thin wall that may not be perceivable, and with no internal echoes. Few thin septa may be seen. On the other hand, epidermoid cysts are usually bigger and have intense internal echoes that are presumed to reflect the keratin

content and/or cholesterol crystals. Pseudocysts are acquired splenic cysts that are often post-traumatic and do not have a real epithelial wall and the wall is often calcified. They may also have internal echoes usually due to the aging blood products. We have observed that the internal echoes of epidermoid cysts always have intense twinkling artifacts whereas pseudocyst or hydatid cyst contents do not create twinkling artifacts. Gharbi classification for the liver hydatid cysts can be applied to the spleen. One of the most important points in differential diagnosis is the thick cyst wall which is composed of at least two layers; endocyst and ectocyst. However, it is not always easy to appreciate the thickness of the cyst wall; in children it may not be so thick yet or the pressure inside the cyst may be too high and compress the layers. If the internal pressure drops after a trauma or treatment, the membranes float in the cyst and the image becomes pathognomonic for a hydatid cyst. Gharbi type 3 cysts have multiple daughter cysts. Gharbi type 4 cysts are basically pseudomass lesions that may imitate neoplasms. Hydatid cysts in the liver that often accompany splenic ones, positive serology and childhood in an endemic region are helpful clues in favor of hydatid cyst.

Treatment

Although the treatment options of splenic cysts vary, it is generally accepted that 5-6 cm diameter is a cut-off value to consider intervention. Interventions include percutaneous aspiration and sclerotherapy, and laparoscopic or open surgical removal of the cyst preserving the spleen, if possible. Splenectomy may be the last option [3].

Epithelial small cysts that are less than 5-6 cm in diameter may be followed-up by US annually. Epidermoid cysts usually enlarge over time and have to be removed surgically. Percutaneous treatment may be effective but not always successful [8].

Hydatid splenic cysts may be treated by percutaneous drainage and sclerotherapy, with good results, depending on the Gharbi stage. Type 1 and 2 hydatid cysts have high success rates similar to the results obtained in the liver and kidneys [12]. Surgery is usually preferred for Gharbi type 3 and active type 4 cysts. Inactive type 4 and type 5 cysts may be followed up to ensure inactivity. Pseudocysts which are believed to result from trauma, can also be followed up. Percutaneous drainage, laparoscopic fenestration and other spleen preserving surgeries may be indicated depending on the symptoms and the growth rate of the pseudocysts [5].

Conclusion

Splenic cysts are rare; most of them are small simple cysts that can be followed-up. They usually do not

grow and do not produce symptoms. Epidermoid splenic cysts should be followed up with caution because they often grow over 5-6 cm diameter and cause pressure symptoms. Differential diagnosis of epidermoid cysts is possible by searching for the twinkling artifact in color Doppler mode US. The internal echoes of neither the pseudocyst nor the hydatid cyst is expected to create a twinkling artifact.

Conflict of interest: none

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