

Epidermoid cyst of the spleen

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Cysts of the spleen are not common, nevertheless, can occur. When they occur, they pose diagnostic and management challenges. Most of these cysts are benign and when they are small in size, these are usually asymptomatic. Large cysts can rupture, causing varying degrees of morbidities. However, most are discovered incidentally during work up for other medical conditions. Although it is mere speculations, it seems that more cysts are expected to be diagnosed, since the improvements of radiological imaging techniques and its more frequent use. We present here a case of epidermoid cyst of the spleen, and offer short discussion on the differential diagnosis, clinical and pathological features, and a brief literature review.

A 36-year-old woman who suffered unexplained upper abdominal pain for the preceding 4-6 weeks, and had unremarkable physical exam. She denies any history of recent trauma, travel abroad, or taking medications other than regular pain medications. Computed tomography (CT) examination with contrast of the abdomen was performed. The radiologic imaging revealed 5-cm hypodense well-circumscribed cystic mass of the spleen with multiple smaller cystic lesions at the periphery. A subsequent splenectomy was performed. The specimen consisted of a 370-gm fresh spleen with a large subcapsular multilocular cyst measuring 6.7 x 6.5 cm. The cyst had a thick wall and was filled with a yellow semi-solid grumous material without bone or hair. The inner surface of the cyst was glistening and shiny with no nodules. Adjacent to the largest cyst, 2 other smaller cysts measuring 1.8 and 1.0 cm and filled with white gray material were also identified. The remaining of the spleen parenchyma was grossly unremarkable, and devoid of solid masses or nodules. Histologically, the major cyst was surrounded by a thick fibrous capsule with no connection to the surrounding splenic parenchyma. The cysts were lined by a stratified squamous epithelium with associated keratin, cholesterol cleft, and focal calcifications (Figures 1). There were neither skin appendages in the wall of the cyst, nor additional solid tissue inside the cavity. The diagnosis was "epidermoid cyst of the spleen."

Splenic cysts can be categorized into 3 subtypes: pseudocysts (false or secondary cysts), true (epithelial or primary) cysts, and the parasitic category. Pseudocysts comprise approximately 75% of all non-parasitic cysts of the spleen.^{1,2} Parasitic cysts are most commonly due to *Echinococcus* infestation, and more frequent in endemic areas. True splenic cysts are relatively rare. Of these, epidermoid cysts are the most common and comprise

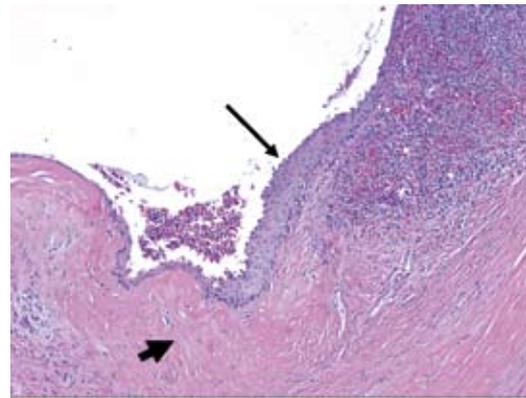


Figure 1 - Low powered views of the cyst with mature squamous lining (long arrow) devoid of skin appendages, hair or bone. Sub-epithelial sclerotic band separating the cyst from splenic parenchyma is noted (short arrow). In other areas cholesterol clefts are also noted inside the lumen (hematoxylin and eosin 100x).

approximately 90% of the cases.³⁻⁴ Most of the reported cases occur in children, or young adults and they present with vague abdominal pain, and splenic mass related to cyst enlargement similar to our patient. Often, these cysts are asymptomatic, and found incidentally on radiological imaging for other unrelated reasons. They can involve accessory spleen, and cases originating from an intrapancreatic accessory spleen had been reported. They typically appear as a hypodense mass on CT scan that does not enhance with contrast. The radiological differential diagnosis includes epidermoid/dermoid cysts, parasitic cysts, pseudocyst, splenic infarction, splenic abscess, and cystic neoplasms. The epidermoid cysts are common in the United States, however, parasitic cysts are more common worldwide. Although the exact pathogenesis of the epidermoid and other epithelial cysts is not known, speculations about embryonic epithelial inclusions, and invaginations of the surface mesothelium followed by metaplasia are possible explanations. Interestingly, the epidermoid splenic cysts can be associated with elevated level of carcinoembryonic antigen and carbohydrate antigen 19-9 in the aspirated cystic fluid and serum.⁵ Using immunohistochemical stains, Higaki et al⁵ documented the presence of these markers in the squamous epithelium, and suggested that the squamous epithelium is the site of production. In addition, they noticed a drop of serum markers after surgical removal.

Total splenectomy, partial splenectomy, and even cystectomy are adequate treatment. Laparoscopic surgical fenestration of the cyst had been attempted to save the spleen however, was found to be associated with post operative splenic infection, and higher rate of recurrence. Ruptured cyst and related size of the mass are the few complications.

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References

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