Review Article

Treatment of splenic cysts

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Abstract: Splenic cysts are a relatively rare entity with variable etiology. They are typically classified as primary or secondary cysts, and primary cysts are divided into parasitic and non-parasitic cysts. Parasitic cysts are typically caused by *Echinococcus granulosis* or *multilocularis*. Secondary cysts lack an epithelial lining and are therefore considered psuedocysts. They are frequently discovered incidentally and otherwise typically present with symptoms related to mass effect of the cyst. The management is greatly dependent upon size and symptomatology. Cysts that are parasitic, large, or symptomatic require operative intervention. Cysts that present with rupture or infection also require operative intervention. Laparoscopic surgery is safe and effective in surgical management of splenic cysts, and spleen preservation is preferable whenever it is possible, though splenectomy vaccines should be given if spleen preservation seems unlikely. There are a variety of techniques in operative management of splenic cysts. They can be managed by partial splenectomy, decapsulation, cyst fenestration, or unroofing of the cyst. The other technique that can be used in patients who are of prohibitive surgical risk is "PAIR" which stands for Puncture of cyst, Aspiration of cyst contents, Injection to sterilize the cyst, and Re-aspiration. This has a greater recurrence rate as does any technique that leaves any remaining cyst wall behind.

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Overview

Given the rarity of splenic cysts, a majority of the literature is composed of various case series studies with little high-quality evidence on optimum management. There are different classification systems of splenic cysts, and the etiologies are variable. The first distinction is between primary, or "true" cysts which is approximately 25% of splenic cysts, and secondary cysts or pseudocysts which are the other 75%. The primary cysts all possess an epithelial lining, and are further sub-divided into parasitic and non-parasitic cysts (1,2).

Parasitic primary cysts

The spleen is the third most frequently affected site of hyatid cysts after liver and lung. Hyatid cysts caused by *Echinococcus granulosis* or *mulitlocularis* (3). This is the most common cause of primary splenic cysts globally, and is seen more in endemic areas like the Mediterranean and Eastern Europe. Hyatid cyst should be suspected if the cyst has calcified walls or there are other intra-abdominal cysts with intraluminal daughter cysts. Their typical appearance is an inner germinal layer, the endocyst, and an outer laminated layer, the ectocyst.

Non-parasitic primary cysts

The remainder of primary cysts are congenital and appear as simple cysts. They have a trabeculated lining with epidermoid, transitional, or mesothelial epithelium, and can contain more than one type of epithelial lining (4). A majority are congenital epidermoid cysts which are caused by a defect in mesothelial migration leading to unfolding of the peritoneal mesothelium or from entrapment within the splenic sulci. The epithelial cells are often positive for CA 19-9 and CEA on immunohistochemistry, and this can lead to elevated serum levels of these tumor markers, though the cysts have no malignancy potential (5,6). They also stain positive for keratin which is characteristic of epithelium, and negative for factor 8 which is characteristic of endothelium (4). On gross appearance they have a smooth white or gray lining, and contain fluid with variable characteristics. Another exceedingly rare form of non-parasitic primary cysts are dermoid cysts which contain dermal appendages like hair follicles and sweat glands.

Secondary cysts/pseudocyts

Secondary cysts are defined by their lack of epithelial lining, making them pseudocysts. Secondary cysts typically have smooth borders, are unilocular, and can be thick-walled with focal calcifications. Most are hypothesized to be post-traumatic and form after a hematoma liquefies and fails to resorb. Traumatic cysts often have a shaggy hemorrhagic interior (4). Secondary cysts can also occur after a splenic infarct when that portion of spleen liquefies.

Splenic peliosis

This is a rare entity that is characterized by multiple cystic, blood filled cavities in the spleen that occur as a result of sinusoidal dilation. They are found incidentally but do pose a risk of splenic rupture and intra-abdominal hemorrhage (7).

Presentation

Splenic cysts are frequently found incidentally on abdominal ultrasound, computed tomography (CT) scan, or magnetic resonance imaging (MRI) (8). If patients do present with symptoms, they are typically vague and a result of mass effect of the cyst. In symptomatic patients the most common symptoms are vague abdominal pain, early satiety, nausea, vomiting, left shoulder pain, abdominal distension, pleuritic chest pain, shortness of breath, splenomegaly, or even hypertension as a result of compression of the left kidney (4,8).

Work up

Often patients will present to a surgeon after an imaging study has already shown a splenic cyst. On ultrasound cysts are anechoic or hypoechoic intrasplenic lesions, and can have internal echoes from debris. There may be shadowing from calcifications in the cyst wall, internal septation, and wall trabeculation. On CT the wall is thin, with sharp definition to the parenchyma of the spleen, and the rim of the cyst cavity does not enhance with intravenous contrast (9). On MRI they appear as well-defined cysts with a homogeneous fluid cavity. Parasitic cysts are calcified and have daughter cysts. When a patient presents with a splenic cyst, it is imperative to perform serologic testing to confirm whether it is a parasitic cyst prior to proceeding to surgery. Otherwise the type of cyst is typically not known until it is examined by pathology (9). There is little utility in percutaneous sampling of the cyst for diagnosis, as management of non-parasitic cysts is the same, and the determination of cyst as parasitic does not require sampling.

Occasionally patients will present with cyst rupture or infection, which requires operative intervention (10).

Management

For parasitic splenic cysts, the management is typically surgical. If patient factors put them at prohibitive risk for surgery, and the cyst is less than 5 centimeters in diameter, a technique called “PAIR” has been described. This stands for puncture of cyst, aspiration of cyst contents, injection with 3% hypertonic saline, alcohol, or 0.5% silver nitrate to sterilize the cyst, and re-aspiration, in conjunction with anthelminthics (11). Typically, parasitic cysts should not be drained percutaneously due to risk of seeding or anaphylaxis. Spleen preserving surgery can be attempted if there is a small solitary cyst on the periphery (3), but care should be taken to ensure that there are 1-centimeter margins from the cyst due to the invasive nature of the infection (11). If spleen preservation does not seem possible, splenectomy should be performed with great care not to rupture the cyst, which could again lead to anaphylaxis or intra-abdominal spread. Epinephrine should be on hand in the operating room to treat anaphylaxis in the case of cyst rupture.

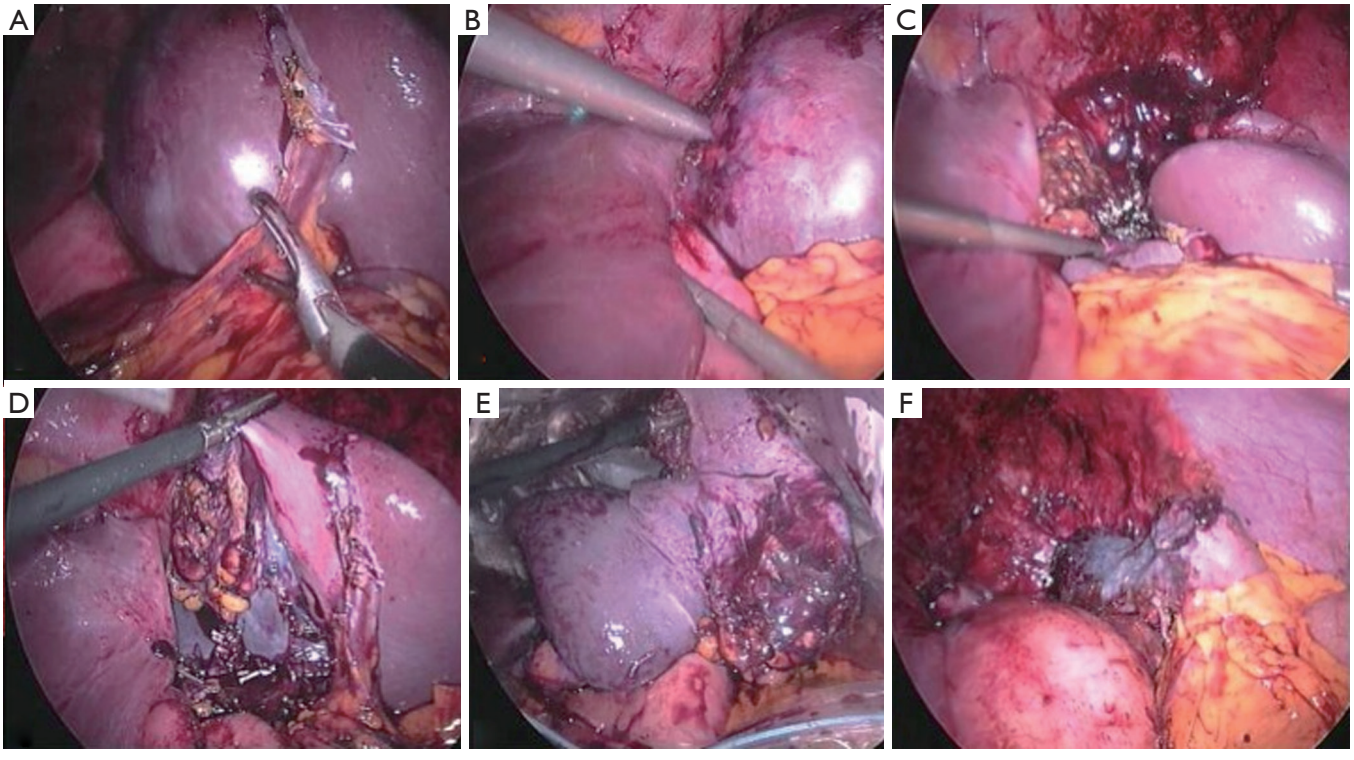

Figure 1 Laparoscopic approach to excision of splenic cyst. (A) Identification of splenic cyst and taking down adhesions; (B) entering and decompressing cyst; (C) decompressed cyst; (D) inside of cyst cavity; (E) dissection off spleen; (F) spleen post resection.

Small, asymptomatic non-parasitic splenic cysts or pseudocysts can be monitored. Previously a cutoff of 5 centimeters was used to determine operative versus non-operative management (4,12). If smaller than 4 or 5 centimeters the cysts could be observed with yearly ultrasounds to monitor for growth or involution over time. Kenney *et al.* found that using size as a cutoff was irrelevant, and that treatment should be based on symptoms (2). Asymptomatic cysts, when followed, would shrink over time and therefore not require surgery, whereas symptomatic cysts would typically enlarge, necessitating surgery.

When operative, simple cysts can be managed in a variety of ways from total splenectomy to simple deroofing of the cyst (13,14). These can all be performed laparoscopically (13,15). The trend is moving towards spleen preservation whenever possible, with partial splenectomy, decapsulation, cyst fenestration, or deroofing (8,12). There is also the option for percutaneous drainage. This topic is controversial, and some recommend it, while others have found that it should not be attempted due to high recurrence rates (2).

The technique for decapsulation involves suctioning out the cyst, opening the anterior cyst wall and then circumferentially excising the cyst wall using an energy device such as the LigaSure or Harmonic Scalpel. This technique leaves the portion of the cyst wall that is stuck to the spleen, which leads to less blood loss when compared with excising a portion of contiguous spleen parenchyma (*Figure 1*). This is preferred over simple unroofing, which had a higher recurrence rate (13,16). Any remaining cyst wall can be a nidus for reformation of cyst. If the back wall is left in place, omentum can be tacked to the area, though some propose that this can increase the risk of recurrence.

Partial splenectomy is preferred in patients who have recurrent splenic cysts or primary splenic cysts, as the recurrence rate is less than that after decapsulation (17), and generally speaking partial splenectomy lead to less recurrence than more conservative tactics (18). If based on location of the cyst there is a high likelihood that the patient will require a splenectomy, they should receive splenectomy vaccines (pneumococcal, meningococcal, and hemophilus influenzae) 2 weeks prior to surgery, or 2 weeks after surgery if the splenectomy was not expected and they did not receive pre-operative vaccines. In the rare patient

who presents with cyst rupture or infection, operative intervention is typically performed using any of the described techniques (10).

Splenic cysts are rare, and most often incidentally found. Surgery is indicated in parasitic cysts and symptomatic non-parasitic cysts. The trend is toward minimally invasive, spleen-preserving surgery. If 25% of the spleen parenchyma is retained, then patients should have intact function. There are many operative options available, but less recurrence is seen when the complete cyst wall is removed by partial splenectomy.

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