A review of the cysts of the spleen

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Abstract

Cystic lesions of the spleen are relatively uncommon and often incidental findings. Given the rarity of these lesions, when encountered clinically, a wide range of differential diagnosis are considered. This review highlights the spectrum of neoplastic and non-neoplastic cystic lesions of the spleen and summarizes the pathogenesis, clinical presentation, key radiologic and histopathological features, associated complications and various available treatment options.

Cystic lesions of the spleen can be classified broadly as non-neoplastic (pseudocysts, congenital, infectious or vascular cysts) and neoplastic (primary or metastatic cystic neoplasms). With frequent use of abdominal imaging techniques such as ultrasonography (US) and computed tomography (CT), the detection of splenic cysts has increased. Total splenectomy has been the preferred treatment for large, multiple, deeply located splenic cysts. However, because of immunologic role of the spleen and increased risk of post-splenectomy infections, currently spleen-preserving minimally invasive procedures have been advocated for superficially located splenic cysts, especially in children and young adults.

Keywords lesions; neoplastic; non-neoplastic; spleen

Introduction and general comments

Cystic lesions of the spleen are infrequent findings, encompassing a wide range of differential diagnosis. In an autopsy series of 42,327 cases, the reported incidence of the splenic cyst is 0.07%.[1] Splenic cysts can be classified as primary or secondary; parasitic or non-parasitic and true or false cysts.[2–5]

In this review, various non-neoplastic and neoplastic cystic lesions of the spleen (Table 1) will be discussed. Non-neoplastic splenic cysts include post-traumatic (pseudocysts), congenital, inflammatory and vascular cysts, whereas, neoplastic splenic cysts can be either benign or malignant.

Congenital cysts are true cysts lined by various types of epithelium. Pseudocysts (false cysts) are usually post-traumatic in origin and lack epithelial lining. Spleen can be affected by various bacterial, fungal or parasitic organisms; resulting in abscess formation. Furthermore, both benign and malignant (primary or secondary) cystic neoplasms can affect the spleen. Clinically, small splenic cysts are usually asymptomatic. Larger cysts however, may present with epigastric fullness, palpable mass, splenomegaly, abdominal pain or discomfort. In addition, cysts can be symptomatic secondary to adhesions or pressure on adjacent structures such as the diaphragm, stomach, liver, pancreas, kidney and omentum.[3] Other non-specific symptoms include nausea, vomiting, flatulence and constipation.[2] Occasionally, splenic cysts may present with complications such as infections, anaphylactic shock, fistula to hollow organs, acute peritonitis, haemoperitoneum and empyema as a consequence of a ruptured cyst.[5,6] Laboratory tests such as elevated serum carbohydrate antigen (CA) 19-9 and carcinoembryonic antigen (CEA) levels[5]; serological tests and several imaging modalities such as US, CT or magnetic resonance imaging are valuable in approaching the diagnosis.[7] Total splenectomy has been the preferred treatment for larger, multiple, deeply located splenic cysts. However, given the immunologic role of the spleen and increased risk of post-splenectomy infections, currently spleen-preserving minimally invasive procedures such as partial splenectomy (laparoscopic or open), total cystectomy, percutaneous sclerotherapy, marsupialization and fenestration have been recommended for superficially located cysts, especially in children and young adults.[5]

Cystic lesions of the spleen

Non-neoplastic	***Neoplastic***
Post-traumatic:	*Benign cystic neoplasms:*
• Pseudocyst	• Haemangioma
Inflammatory:	• Lymphangioma
• Hydatid cyst	*Malignant cystic neoplasms:*
• Bacterial or pyogenic abscess	• Primary
• Fungal abscess	○ Mucinous cystadenocarcinoma
Congenital:	○ Primary splenic lymphoma
• Epidermoid	• Metastatic
• Mesothelial	○ Melanoma
• Dermoid	○ Breast, ovarian, gastric, colon and pancreatic cancers
Vascular:	
• Post-infarction	
• Peliosis	

Table 1

Pseudocyst

Splenic pseudocysts, also known as false or secondary cysts, are devoid of epithelial lining and often post-traumatic in origin.[2] Clinical evaluation of the patient with pseudocyst often reveals history of trauma to the left upper quadrant. It is believed that the pseudocysts are the most common non-parasitic splenic cysts.[5] Following splenic trauma, a subcapsular or intraparenchymal haematoma may form which with, subsequent organization, liquefaction, resorption and encapsulation may transform into a pseudocyst. The formation of haematoma is dependent upon several factors, including type, severity, site of trauma, as well as integrity of the splenic capsule and blood coagulation status of the patient.[5] On US imaging, it is difficult to distinguish between a true epithelial lined cyst and a pseudocyst. However, certain characteristics such as echogenicity from the debris and

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calcification are helpful in identifying a pseudocyst. On CT, pseudocysts appear as sharply demarcated, unilocular, low density lesions. Macroscopically, pseudocysts are often unilocular containing mixtures of blood and necrotic debris. Microscopically, pseudocysts are filled with blood and debris and are surrounded by dense fibrous wall and also lack epithelial lining. Calcifications and hemosiderin deposition may be seen within the fibrous wall.[3]

Splenic abscess

Splenic abscess is relatively a rare clinical condition characterized by localized collection of pus within the splenic parenchyma. Given its non-specific clinical presentation, it poses a great diagnostic challenge. Splenic abscess affects males and females equally and usually seen in middle-aged and older patients. Several etiologic factors for splenic abscess include; infections (infective endocarditis, pneumonia, otitis media, osteomyelitis, urinary tract infections, diverticulitis), recent surgery, splenic trauma, immunodeficiency (diabetes mellitus, congenital or acquired immunodeficiency syndrome, use of immunosuppressive therapy), haematological disorders (thalassaemia, sickle cell anaemia, leukaemias) and intravenous drug abusers or alcoholics.[8,9] Occasionally, these may be encountered in patients lacking underlying risk factors.[10] Though nonspecific, the clinical features include fever, left upper quadrant pain, anorexia, nausea and vomiting. A wide variety of pathogens (bacterial, fungal and protozoa) have been isolated from splenic abscesses. Solitary abscesses usually harbour bacterial organisms (*Staphylococcus, Streptococcus, Klebsiella, Pseudomonas, Salmonella, Escherichia coli* and *Mycobacterium*) whereas; multiloculated splenic abscesses predominantly contain fungal organisms (*Candida, Aspergillus* and *Cryptococcus*) and are more frequent in immunosuppressed patients[9,11]. Delay in diagnosis or treatment can be detrimental to the patient and associated with high mortality. Both US and CT scan are the effective diagnostic imaging tools. However, CT scan is the most reliable and sensitive imaging technique, which can detect small collections and even small amounts of gas within abscess.[9,12] On US imaging, abscesses appear as anechoic to hypoechoic, poorly defined lesions. High echogenicity is usually seen in relation to gas formation. While, CT scan may show well-delineated, low attenuation masses.[7] Grossly, abscesses have irregular borders and may have a pseudocapsule or no capsule. Histologically, abscesses contain dense inflammatory infiltrate with abundant neutrophils and necrotic debris. Open splenectomy with antimicrobial therapy is the preferred treatment. Other minimally invasive treatment modalities include laparoscopic splenectomy and percutaneous image-guided drainage.[8]

Parasitic cyst

Hydatidosis (hydatid disease) is a parasitic infection caused by the genus *Echinococcus*. *Echinococcus granulosus* (EG) is the most common species causing hydatidosis in humans. Commonly affected organs are liver (75%) and lung (15.4%) but hydatid disease can be seen in any organ of the body. Splenic involvement is rare with reported prevalence ranging from 0.5 to 8%.[13–15] Concomitant involvement of multiple organs has also been reported in about 5–13% of cases.[16] Splenic hydatid disease can be an isolated finding or can occur as a part of disseminated disease secondary to ruptured liver hydatid cyst (Figure 1). These slow growing cysts are usually asymptomatic. Most frequently reported symptoms are palpable mass, fever, dull pain or splenomegaly. Occasionally, patients may present with complications such as anaphylactic shock secondary to the cyst rupture, secondary infections and fistula formation with adjacent hollow organs. Adult forms of EG are found in the small bowel of the definitive hosts (dogs and other carnivores). The definitive host passes eggs in the feces that are ingested by the intermediate hosts (sheep, goat, cattle and pigs) and humans (accidental fecal-oral transmission). Oncospheres from the hatched eggs penetrate the intestinal mucosa; enter the circulation and lodge in the liver or other organs resulting in cyst formation.[15] Definitive hosts are infected by ingesting visceral organs of the intermediate hosts. There is no direct transmission from humans to humans. Many available diagnostic serological tests include complement fixation, enzyme-linked immunosorbent assay (ELISA) or indirect haemagglutination.[15] Imaging techniques such as US and CT are the useful diagnostic tools for evaluation of splenic hydatidosis, but may show nonspecific features. Grossly, cysts tend to be unilocular filled with clear fluid that contains many daughter cysts and brood capsules with protoscolices. Histologically, the cyst wall shows three distinct layers:

- Inner layer (germinal layer) is composed of daughter cysts and brood capsules with protoscolices.
- Middle layer (laminated membrane) is eosinophilic, avascular and refractile.
- Outer layer consists of dense fibrovascular tissue.

Antihelminthic drugs (Albendazole, Mebendazole) with splenectomy are the treatment of choice. Total splenectomy is preferred when cysts are larger, multiple or located more centrally or in the hilar region. Other available conservative modalities include enucleation of the cyst, partial cystectomy, percutaneous aspiration irrigation and re-aspiration (PAIR) and intraoperative use of scolicidal agents.[15]

Congenital cysts

Congenital cysts (aka epithelial cysts) are the true cysts lined by the epithelium (Figure 2). These are typically seen in patients in their second and third decade of life. The exact histogenesis of the congenital cysts is still unclear. Some authors have postulated origin of these cysts from peritoneal mesothelial cells, since mesothelial cells have tendency to undergo squamous metaplasia in the face of chronic irritation. Other suggested aetiologies include: displacement of the epithelium during embryogenesis or teratomatous differentiation.[6] Clinically, epithelial cysts are usually asymptomatic, found incidentally on imaging. However, in symptomatic patients the most common symptom is right upper quadrant pain. Elevation of tumour markers such as CA 19-9 and CEA in the serum and the cystic fluid has been reported, which returns to baseline after splenectomy.[17,18] On imaging, these cysts are usually unilocular anechoic lesions. On gross examination, the cyst wall appears smooth and glistening with trabeculated internal lining. Histologically, based on the type of epithelial lining these are categorized into epidermoid cyst with stratified squamous epithelial lining, mesothelial cyst with

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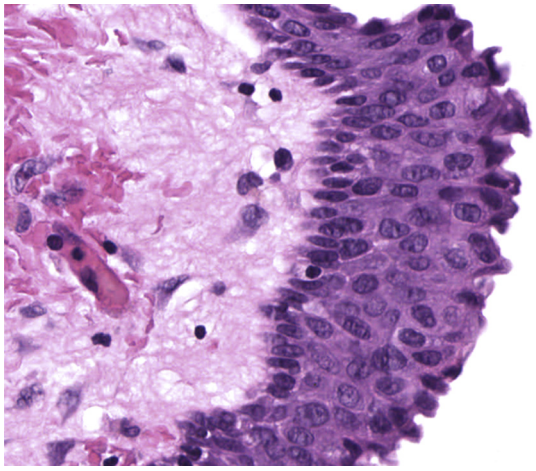

Figure 1 Hydatid cyst of the tail of the pancreas and spleen showing atrophic pancreatic tissue and a cyst composed of laminated eosinophilic material. (H&E ×25).

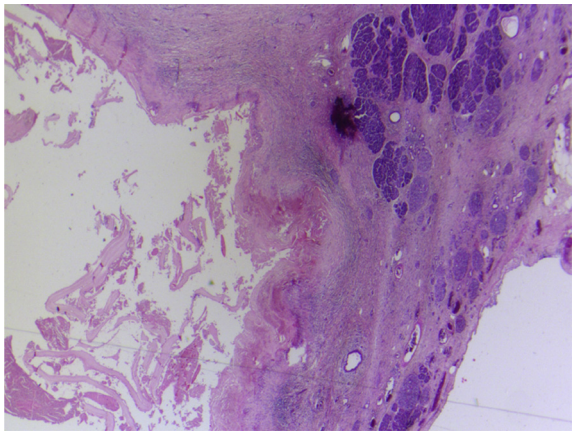

Figure 2 Epithelial cyst wall lined by a stratified squamous epithelium (H&E ×400).

cuboidal epithelial lining and dermoid cyst lined by squamous epithelium with hair follicles, sebaceous glands and skin appendages. By immunohistochemistry, the epidermoid cysts are CA 19-9, CEA and cytokeratin positive but show no immunoreactivity for calretinin; whereas mesothelial cysts are calretinin and cytokeratin positive but show no staining for CA 19-9 and CEA[6] (Figure 3). The differential diagnosis includes benign vascular tumours such as lymphangioma and haemangioma. Epithelial cysts are negative for endothelial immune markers. The treatment options vary considerably depending upon the size and location of the cyst, patient's age and presence or absence of symptoms. Various treatment modalities include splenectomy (total or partial), partial cystectomy, splenic decapsulation.[6]

Post-infarction cyst

Splenic infarction is caused by occlusion of either arterial supply or venous drainage that leads to tissue ischaemia and subsequent necrosis. Several etiologies associated with infarction include; sickle cell disease, thromboembolic events such as infective endocarditis, inherited clotting disorders, connective tissue disorders, vasculitides, myeloproliferative disorders, haematological and non-haematological malignancies. Patients with splenic infarcts are often asymptomatic, however occasionally may present with left upper quadrant pain, fever and left shoulder pain secondary to irritation of the diaphragm. On ultrasound imaging areas of acute infarct with cystic change may appear hypoechoic. On CT scan an acute splenic infarct appear as an area of low attenuation.[7] Macroscopically, infarcts are wedge-shaped and subcapsular in location. Acute infarcts appear hemorrhagic whereas old infarcts tend to be paler. On microscopic evaluation, the splenic parenchyma in acute infarction may show coagulative-type necrosis, oedema, haemorrhage and occasionally cystic changes. In chronic infarction there is usually loss of parenchymal volume secondary to fibrosis. Haemorrhage, rupture, pseudocyst and abscess formation are potential complications of splenic infarcts.

Peliosis

Isolated splenic peliosis is a rare finding, as most of the cases show simultaneous liver involvement. It is characterized by blood-filled cystic spaces within the splenic parenchyma. The exact aetiology of peliosis is unknown. It has been associated with several conditions and illnesses such as tuberculosis, haematological malignancies such as Hodgkin disease and multiple myeloma, acquired immunodeficiency syndrome (AIDS), post-transplant immunodeficiency, intravenous drug abuse, chronic alcoholism, intake of anabolic steroids or oral contraceptives, viral infections and prior thorium contrast injections.[7,19] Patients with splenic peliosis often have no symptoms; therefore, their detection is usually incidental on imaging or at autopsy. Peliosis frequently involves the entire spleen. On US imaging, splenic parenchyma shows multiple hypoechoic or hyperechoic lesions. Similarly, CT scan shows multiple low-attenuation lesions of different sizes throughout the splenic parenchyma. Grossly, multiple intraparenchymal blood-filled cystic cavities of variable size are seen. Thrombosis within the blood spaces can also occur. On microscopic examination, these blood-filled cystic cavities frequently show well-demarcated margins usually in parafollicular distribution.[20] The cystic cavities are closely spaced without intervening connective tissue septa.[7] Because of rarity of this condition, there are no guidelines regarding management of these patients. However, rupture of the cysts whether secondary to trauma or spontaneous, can lead to fatal intraperitoneal haemorrhage; therefore splenectomy is considered to be a reasonable approach.

Haemangiomas

Haemangiomas are benign proliferation of the blood vessels that range from capillary to cavernous in size. These slow growing neoplasms are rare but considered to be the most frequent primary benign neoplasms of the spleen.[21] Splenic haemangiomas are frequently seen in young adults (age range; 35–45 years). These lesions are usually small and mostly detected incidentally on imaging. Clinically, symptomatic splenic haemangiomas may present with non-specific symptoms such as palpable mass, pain or splenomegaly. Splenic haemangiomatosis which is characterized by diffuse involvement and replacement of splenic parenchyma by multiple haemangiomas is frequently seen in

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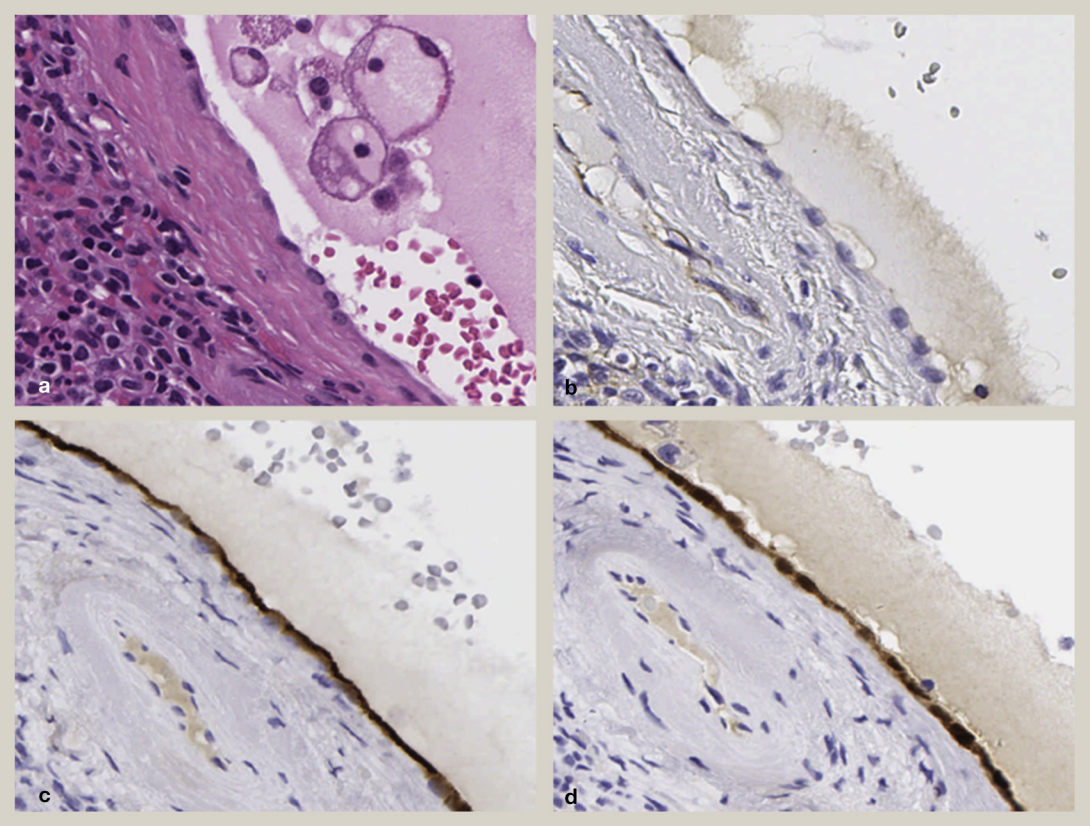

Figure 3 Mesothelial cyst. (**a**) Cyst wall lined by flattened mesothelial cells (H&E ×400). Immunohistochemical stains (×400) show that the mesothelial cells are: (**b**) CD31 negative; (**c**) D2-40 positive; (**d**) Calretinin positive.

association with Klippel–Trenaunay syndrome.[22] Kasabach–Merritt syndrome (KMS) has been reported in association with large (cavernous) haemangiomas. KMS is characterized by anaemia, thrombocytopenia, and consumptive coagulopathy in the presence of diffuse cavernous splenic haemangiomas.[23] Depending on the size, percentage of solid or cystic component the radiologic findings of splenic haemangiomas may vary. On ultrasound imaging haemangiomas are well-delineated echogenic masses that may be cystic or solid. On CT imaging, haemangiomas may appear as well-demarcated low-attenuation lesion involving the splenic parenchyma.[7] Grossly, these are well-defined unencapsulated, solitary or multiple nodular lesions. These lesions are variable in size ranging from a few millimeters to several centimeters and may appear predominantly solid or cystic; mixed-types are also seen. Larger cystic haemangiomas may show degenerative changes and fibrosis or may undergo infarction or thrombosis.[21] Histologically, cystic haemangiomas are composed of multiple cystically dilated blood-filled vascular spaces lined by endothelial cells with intervening delicate fibrous septa or splenic parenchyma. Differential diagnoses include splenic hamartoma, which is a well-circumscribed lesion within the splenic parenchyma containing disorganized vascular channels. By immunohistochemistry, the endothelial cells stain positive for Factor VIII, CD31 and CD34. Large haemangiomas are at risk of spontaneous rupture resulting in haemoperitoneum. Splenectomy (open or laparoscopic) is the treatment of choice. Partial splenectomy is effective when the lesion is small and more superficially located. Other available treatment options include; arterial embolization, antiangiogenic therapy and radiotherapy.[24]

Lymphangiomas

Splenic lymphangiomas are rare benign vascular neoplasms of the splenic lymphatic vessels. The lesion is frequently observed in children, mostly younger than 2 years of age and rare in adults.[25] The lesion may be seen confined to the spleen exclusively or may involve multiple organs in association with lymphangiomatous syndrome.[21] Regarding the pathogenesis of splenic lymphangiomas, authors have failed to agree on whether it is a hamartomatous lesion, true neoplasm or developmental malformation of the lymphatic vessels.[26,27] Based on the size of the lesion, the clinical presentation is quite variable. Small lesions tend to be asymptomatic and incidentally discovered on imaging whereas, larger lesions are symptomatic and may present with pain, mass or splenomegaly. The lesion appears as hypoechoic or anechoic, multiple thin-walled cysts of varying size on US imaging. Internal septations and peripheral linear calcifications may be identified.[7,27] CT imaging shows multiple well-defined areas of low attenuation with no evidence of contrast enhancement.[27] On macroscopic examination, the splenic lymphangiomas are usually cystic, composed of unilocular or multilocular dilated cysts, typically seen in subcapsular location within the splenic parenchyma or within large trabeculae. Histologically, the lesion is composed of multiple cystically dilated lymphatic channels lined by a single layer of endothelial

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cells and filled with eosinophilic, proteinaceous material. By immunohistochemistry, the endothelial cell lining of the lymphatic channels stain positive for D2-40 (podoplanin). Surgical resection of the spleen is the standard of care and known to have better prognosis. Recurrence after incomplete resection has been reported.[27] The differential diagnosis includes haemangiomas, epithelial splenic cysts (epidermoid, mesothelial) and parasitic cysts. Complications associated with larger lymphangiomas include infection, acute peritonitis secondary to rupture, hypersplenism, consumptive coagulopathy, bleeding and portal hypertension.[28]

Primary splenic mucinous cystadenocarcinoma

Primary mucinous cystadenocarcinoma of the spleen is exceptionally rare with only a few cases reported in the literature.[29,30] Regarding pathogenesis, these tumours can originate from; invaginations of splenic capsular mesothelium with subsequent mucinous metaplasia, heterotopic pancreatic or heterotopic intestinal tissue within the splenic parenchyma. Association of these tumours with pseudomyxoma peritonei has also been described. The clinical features are usually non-specific. Elevated serum CEA and CA 19-9 levels have been reported which normalize postoperatively. On imaging, these tumours may appear as multicystic, multiloculated septated masses. Grossly, the tumour is composed of multiple cysts containing abundant gelatinous mucin. Histologically, these mucin filled cystic spaces are lined by tall to stratified columnar mucinous epithelium containing hyperchromatic atypical nuclei. Papillary projections and back to back gland arrangement can also be seen.

Primary splenic lymphoma

Lymphoma is the most frequently seen malignant tumour of the spleen. Splenic involvement in lymphomas can be either primary or secondary as a part of systemic lymphoma. Secondary involvement of spleen in lymphoma (Hodgkin and non-Hodgkin) is more common than primary splenic lymphoma (PSL), which is a rare clinical entity. Distinguishing PSL from secondary involvement by lymphoma arising elsewhere can be quite challenging. PSL are defined as lymphomas confined to the spleen and hilar lymph nodes with six months relapse-free period following splenectomy.[31] However, the definition of PSL varies considerably in the literature.[32–34] Clinically, patient may present with splenomegaly and other non-specific symptoms such as fever, left upper quadrant pain, weight loss or night sweats. On imaging, splenic involvement in PSL may be seen as homogenous diffuse involvement; small miliary deposits; multiple nodular (>1 cm) deposits or a large solitary mass.[35] PSL may mimic a benign cyst[36] or an abscess[34] and has a potential of being misdiagnosed. Contrast enhanced ultrasound imaging is an effective diagnostic tool.[36] Histologically, small B cell lymphomas are the most commonly encountered PSL. However, aggressive large cell lymphomas such as diffuse large B cell lymphomas (DLBCL) have also been reported. Various treatment options for PLS include total splenectomy, combination chemotherapy and radiation therapy. Total splenectomy with combination chemotherapy is considered to be more effective treatment for primary splenic non-Hodgkin lymphomas.[34,37]

Cystic metastasis

Metastasis to spleen is relatively uncommon but can be seen in association with widespread malignancy. Melanoma is the most frequent primary tumour metastasizing to the spleen.[7] Other epithelial tumours metastasizing with splenic metastasis include carcinomas of breast, endometrium, ovaries, lung, stomach, colon, and prostate. Smaller lesions tend to be asymptomatic whereas the larger lesions often present with left upper quadrant pain or discomfort. Cystic metastatic lesions are mostly hypoechoic on ultrasound imaging and appear hypodense on CT scan.[7] Grossly, a solitary lesion or multiple small metastatic deposits can be seen within the splenic parenchyma or on the capsular surface. Cystic transformation is frequently observed in rapidly growing large metastatic lesions as a result of infarction or tumour necrosis.[38] Microscopic appearance varies based on the type of the primary tumour. ◆

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Practice points

- Splenic cysts should be adequately sampled to reduce the risk of erroneous diagnosis.
- Splenic abscess should be suspected clinically in a febrile patient presenting with left upper quadrant pain or tenderness and leucocytosis.
- Hydatid disease should be considered in differential diagnosis of splenic cystic lesions in patients presenting with; a travel history to endemic regions, simultaneous cystic lesions in the liver or other organs, daughter cysts and cyst wall calcification.
- Given the rarity of the primary splenic mucinous neoplasms, the possibilities; metastasis from elsewhere or direct extension from the adjacent organ should be excluded.
- Primary splenic lymphoma may mimic a benign cyst or an abscess and has a potential of being misdiagnosed.

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