Algorithmic Approach to the Splenic Lesion Based on Radiologic-Pathologic Correlation

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Abbreviations: H-E = hematoxylin-eosin, IPT = inflammatory pseudotumor, LCA = littoral cell angioma, SANT = sclerosing angiomatoid nodular transformation

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SA-CME LEARNING OBJECTIVES

After completing this journal-based SA-CME activity, participants will be able to:

- Explain the imaging features of splenic masses on the basis of the underlying pathologic features.
- Recognize important clinical, pathologic, and imaging features that can help differentiate the various splenic pathologic conditions.
- Formulate a narrow differential diagnosis for splenic lesions based on number and consistency.

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Splenic lesions are commonly discovered incidentally at imaging, without clinical signs or symptoms that may aid in diagnosis. As such, the differential diagnosis and subsequent management are based primarily on imaging characteristics. Much has been written about the myriad pathologic conditions that can occur in the spleen; however, there is little guidance on the approach to an incidental splenic mass. Applying an approach frequently used in imaging to the splenic mass—based on the number and consistency of lesions and refined by supplementary imaging features—allows formulation of a useful differential diagnosis. Solitary cystic masses include true cysts, pseudocysts, and parasitic cysts. When multiple cystic lesions are present, the differential diagnosis expands to include infectious lesions (abscess or microabscesses) and lymphangioma (a benign cystic neoplasm). Hemangioma is the most common solitary solid mass, although other vascular lesions (hamartoma, sclerosing angiomatoid nodular transformation) and nonvascular lesions (inflammatory pseudotumor, lymphoma) manifest as solitary and solid. When multiple solid masses are present, diffuse inflammatory disease (sarcoidosis), littoral cell angioma, and lymphoma should be considered. Malignancies, such as angiosarcoma or metastasis, can manifest as solitary or multiple and solid or cystic masses but are typically associated with symptoms or widespread primary malignancy. Careful assessment of the multimodality imaging characteristics of splenic lesions based on this approach aids the radiologist faced with the incidental splenic lesion.

Online supplemental material is available for this article.

Introduction

The largest lymphatic intraperitoneal organ, the spleen is the first line of defense against blood-borne pathogens. It is composed of two distinct types of tissue: the red pulp contains vascular structures, which filter blood and remove foreign material and damaged erythrocytes; the white pulp is composed of lymphatic tissue and initiates responses to blood-borne antigens.

Technologic advances and the dependence on imaging as an integral component of the clinical evaluation of patients have resulted in increased detection of splenic abnormalities, including splenic masses. Splenic masses are not common, and this rarity contributes to the diagnostic challenge most radiologists face when they are encountered. Most splenic masses are identified incidentally in asymptomatic patients, without laboratory findings to aid diagnosis. As such, the differential diagnosis and management are frequently based entirely on imaging characteristics.

Most incidental splenic lesions are benign; however, the biologic behavior can range from entirely benign to aggressively malignant, and making this distinction is crucial when possible. Not surprising

TEACHING POINTS

- A solitary splenic cyst is almost always benign and usually classified as parasitic or nonparasitic; nonparasitic cysts are then subclassified as primary true or secondary false cysts, as determined by the presence or absence of an epithelial lining.
- Many patients with multiple cystic masses are symptomatic or clinically suspected to have splenic disease. Bacterial and fungal splenic abscesses can be seen in immunosuppressed patients or can follow trauma or emboli.
- Solitary solid splenic lesions include benign vascular entities (hemangioma, hamartoma, sclerosing angiomatoid nodular transformation [SANT]) and malignant vascular entities (angiosarcoma). Of these, hemangioma is by far the most common.
- Lymphoma is the most common malignant tumor involving the spleen. Primary splenic lymphoma—defined as lymphoma confined to the spleen and perisplenic lymph nodes—is rare, comprising less than 1% of all lymphomas.
- Splenic lesions are often unexpected findings at imaging and can present a diagnostic challenge. Using a stepwise approach by first determining if they are cystic or solid, then further characterizing them as solitary or multiple lesions, can help in diagnosis or providing a proper differential diagnosis.

considering the contents of the red and white pulp of the spleen, most primary splenic masses are of vascular or lymphatic origin. The most common benign primary tumor found in the spleen is hemangioma, and the spleen is commonly involved in leukemia and lymphoma owing to its lymphatic components. Beyond these entities, rarity and overlap of imaging features can present a diagnostic challenge. A straightforward approach based on the number and consistency of lesions can best aid the radiologist evaluating the splenic lesion in the clinical setting.

This article highlights the key clinical, pathologic, and radiologic features of splenic masses, with emphasis on radiologic-pathologic correlation, on the basis of an image-based algorithm for characterizing splenic lesions (Fig 1).

Solitary Cystic Mass

A solitary splenic cyst is almost always benign and usually classified as parasitic or nonparasitic; nonparasitic cysts are then subclassified as primary true or secondary false cysts, as determined by the presence or absence of an epithelial lining. Secondary cysts, also known as nonpancreatic pseudocysts, are more common and are associated with trauma, including hemorrhage, infarction, or inflammation (Table 1).

Epithelial Cyst

Clinical Features.—Epithelial cysts, also known as true or epidermoid cysts, are congenital nonneoplastic lesions most commonly seen in the pediatric and young adult population, with a predilection for females (1). Epithelial cysts are rare, with a reported prevalence of 0.07% (1). However, the rate of incidentally detected epithelial cysts is increasing with more common use of noninvasive imaging.

Symptoms correlate with the size of the cyst; many are asymptomatic, while others can manifest with splenomegaly or left upper quadrant pain. The pathogenesis is unclear, with support for the theories of congenital mesothelial invagination, endodermal inclusion, and acquired subclinical trauma with mesothelial displacement (1,2). An elevated level of carbohydrate antigen (CA) 19-9 and carcinoembryonic antigen (CEA) has been noted in patients with large cysts (1).

Pathologic Features.—Grossly, epithelial cysts have a smooth glistening wall with internal coarse fibrous trabeculae (1) (Fig 2). The fluid content can be thin serous or turbid viscous. Histologically, the cyst wall is composed of fibrous tissue, which can contain calcification, and an epithelial lining, composed of stratified squamous, columnar, or cuboidal (mesothelial) epithelium (1) (Fig 2). Immunohistochemistry will show reactivity to cytokeratins (epithelial markers) or calretinin (a mesothelial marker).

Imaging Features.—Epithelial cysts are solitary, unilocular, fluid-filled cysts at all imaging modalities (Fig 2). US and MRI can show internal contents as more complex than simple fluid when the cyst contains cholesterol crystals or hemorrhage (Fig 2). Trabeculae are seen at US and MRI as thick hyperechoic or T2-hypointense bands when compared with the cyst content, respectively. The cyst wall is imperceptible or thin; calcifications are uncommon and more likely seen with pseudocysts (3). There is no contrast enhancement (Fig 2) (4).

Pseudocyst

Clinical Features.—Nonpancreatic pseudocysts, another term for secondary false cysts, the most common nonparasitic splenic cyst, result from liquefaction of posttraumatic hematoma or infarction. Rarely, pseudocysts result from a resolving abscess (1). Most are incidental, although patients with large pseudocysts can present with left upper quadrant pain or fullness (3).

Pathologic Features.—Most pseudocysts are unilocular and contain cloudy fluid. The internal surface is shaggy and hemorrhagic. By definition, there is no epithelial lining; instead,

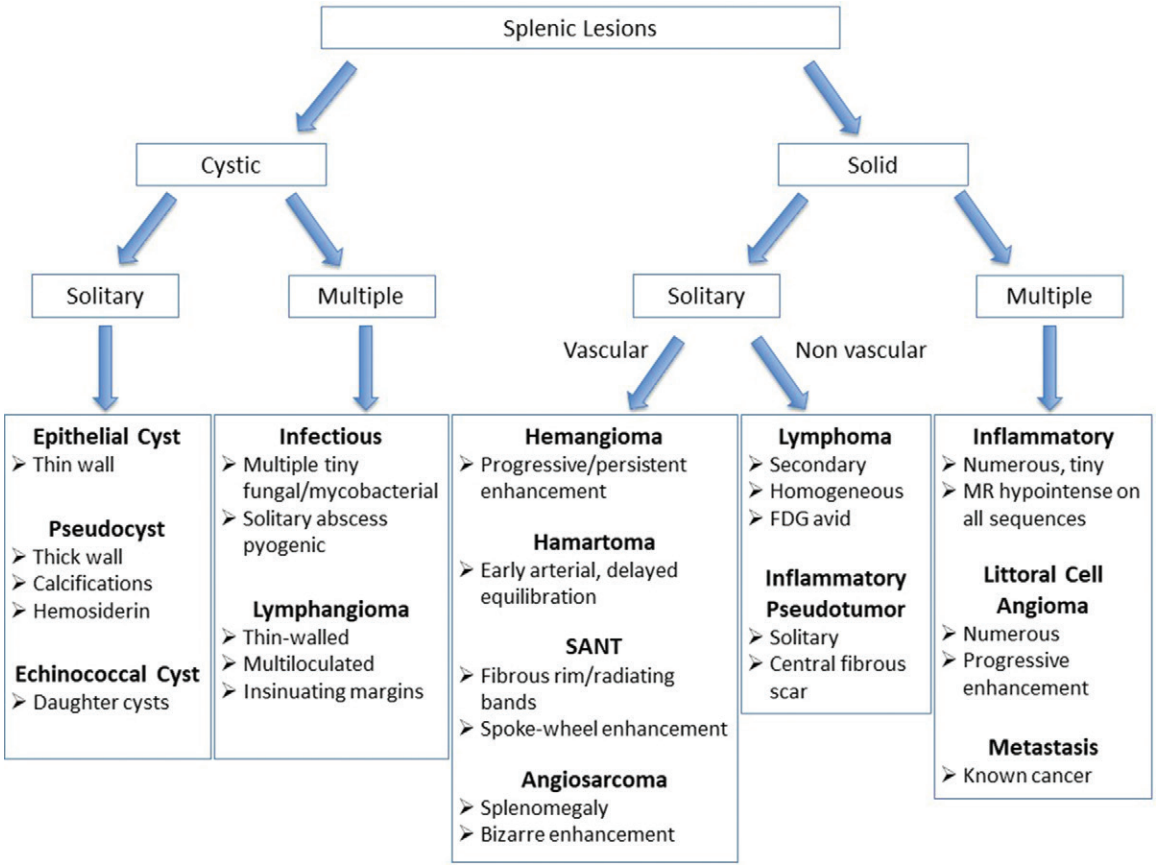

Figure 1. Algorithmic approach to the splenic lesion, by consistency and number. *FDG* = fluorodeoxyglucose, *SANT* = sclerosing angiomatoid nodular transformation.

the wall is composed of dense fibrous tissue, which can contain calcifications, hemosiderin, or cholesterol crystals (Fig 3). Histologically, the internal contents are composed of blood and necrotic debris (1). Immunohistochemistry for pancytokeratin can be performed to confirm the absence of an epithelial lining.

Imaging Features.—Multimodality imaging of splenic pseudocysts reflects their complex pathologic appearance, with a thick fibrous wall, internal debris, and septa. The thick fibrous wall, frequently containing calcification, is well demonstrated at imaging (Fig 3). Most pseudocysts are unilocular, and complex internal contents manifest as internal echogenic debris at US and variable T1 signal intensity at MRI (Fig 3) (3,5). Pseudocysts usually have fluid attenuation at CT. Thin septa may be seen at US, CT, and MRI.

Parasitic (Echinococcal) Cyst

Clinical Features.—Hydatid cyst, the most common parasitic splenic cyst, is caused by *Echinococcus granulosus* tapeworm larvae. Other *Echinococcus* species rarely cause human infection. Hydatid disease is a significant health problem in underdeveloped countries in South America, the Mediterranean region, the Middle East, Africa, and Australia (6).

Although the majority of hydatid disease involves the liver, splenic involvement can occur; the reported prevalence ranges from 0.9% to 8% (1). Splenic echinococcal cysts develop through systemic dissemination or intraperitoneal spread of a ruptured hepatic echinococcal cyst. Patients can present with no symptoms, abdominal pain, splenomegaly, or fever (6,7).

Pathologic Features.—Hydatid cysts associated with *E granulosus* typically form unilocular cysts; rare species, such as *Echinococcus multilocularis* and *Echinococcus vogeli*, form multilocular cysts. Composed of an outer fibrous laminated acellular membrane lined by an inner germinal layer composed of daughter cysts, the hydatid cyst may be surrounded by an outer dense fibrous pericyst layer, frequently with calcification. Grossly, hydatid cyst fluid is clear or pale yellow. When present, the daughter cysts resemble a bunch of grapes (Fig 4) (7). Histologic demonstration of the scolices, the mouth portion of the larvae, is diagnostic (1).

Table 1: Cystic Splenic Lesions—Key Radiologic and Pathologic Features, by Number

Lesion	Nature	Key Imaging Features	Key Pathologic Features
Solitary cystic lesions			
Epithelial cyst	Benign	Well circumscribed, unilocular *Thin imperceptible wall, rare calcifications (Ca^{2+}) *Nonenhancing Simple to complex fluid	Smooth wall Fibrous trabeculae True cellular epithelial lining Contents: serous or turbid viscous
Pseudocyst	Benign	Well circumscribed, unilocular *Thick progressively enhancing wall ± Ca^{2+}, hemosiderin (low T2 SI) Simple to minimally complex fluid	No epithelial lining Encapsulated by thick fibrous wall ± Ca^{2+}, hemosiderin, or cholesterol Contents: blood, necrotic debris
Echinococcal cyst	Benign	Well circumscribed Unilocular (*Echinococcus granulosus*) >> multilocular (*E multilocularis*, *E vogeli*) *Daughter cysts *Condensed maternal matrix: nonenhancing; hyperechoic, hyperattenuating, and T1 hyperintense to simple fluid	Outer dense fibrous pericyst (plus Ca^{2+}) Fibrous lamellated membrane Inner germinal layer composed of daughter cysts
Multiple cystic lesions			
Infectious	Benign	Fungal or mycobacterial: multiple tiny fluid lesions or hypoechoic or hypoattenuating *US: target pattern (central necrotic nidus, hyperechoic viable fungus, hypoechoic inflammatory rim) *MRI: iron-laden macrophages in inflammatory rim; low T1 and T2 SI CT and MRI: peripheral rim enhancement for treated subacute or chronic lesions Treated lesions: calcifications (CT > MRI [blooming artifact]) Pyogenic: solitary or few; ± gas, rim enhancement	Pyogenic: solitary, large Fungal or mycobacterial: multiple, tiny Thick irregular wall Leukocyte infiltration
Lymphangioma	Benign	*Well-defined thin-walled multiloculated fluid-containing structures Intervening fibrous stroma or scarring can have progressive enhancement, ± calcifications Insinuating margins	Thin-walled cysts Content: lymphlike fluid Stroma: smooth muscle plus Ca^{2+} Central scarring D2-40 antibody positive

Note.—> = more common, >> = much more common, SI = signal intensity, ± = with or without.
*Key findings for each entity.

Imaging Features.—Usually solitary, a spectrum of hydatid cyst multimodality imaging appearances depending on the complexity of the cyst has been described: type 1 is a simple cyst with no internal architecture, type 2 has daughter cysts and condensed maternal matrix (Fig 4), type 3 has mural calcifications, and type 4 has features of complications, such as rupture or superinfection (6). Rupture occurs in 50%–90% of cases owing to age and degeneration of the parasitic membranes (Fig E1) (6,8).

Multiple Cystic Masses

Many patients with multiple cystic masses are symptomatic or clinically suspected to have splenic disease. Bacterial and fungal splenic abscesses can be seen in immunosuppressed patients or can follow trauma or emboli. Although lymphangiomas can manifest as a solitary cystic lesion, they are more frequently multiple or diffuse (Table 1).

Infectious Lesions

Clinical Features.—Splenic abscesses can be bacterial, mycobacterial, or fungal and can result from hematogenous seeding, direct extension, sequelae of trauma, or prior infarcts (1). Common causes of bacterial abscess include endocarditis, pneumonia, gastrointestinal perforation, or arteriovenous malformation. The most common bacterial microbes include *Escherichia coli*, *Staphylococcus*, *Streptococcus*, and *Salmonella* (9,10). Although uncommon in the United States, granulomatous infection with

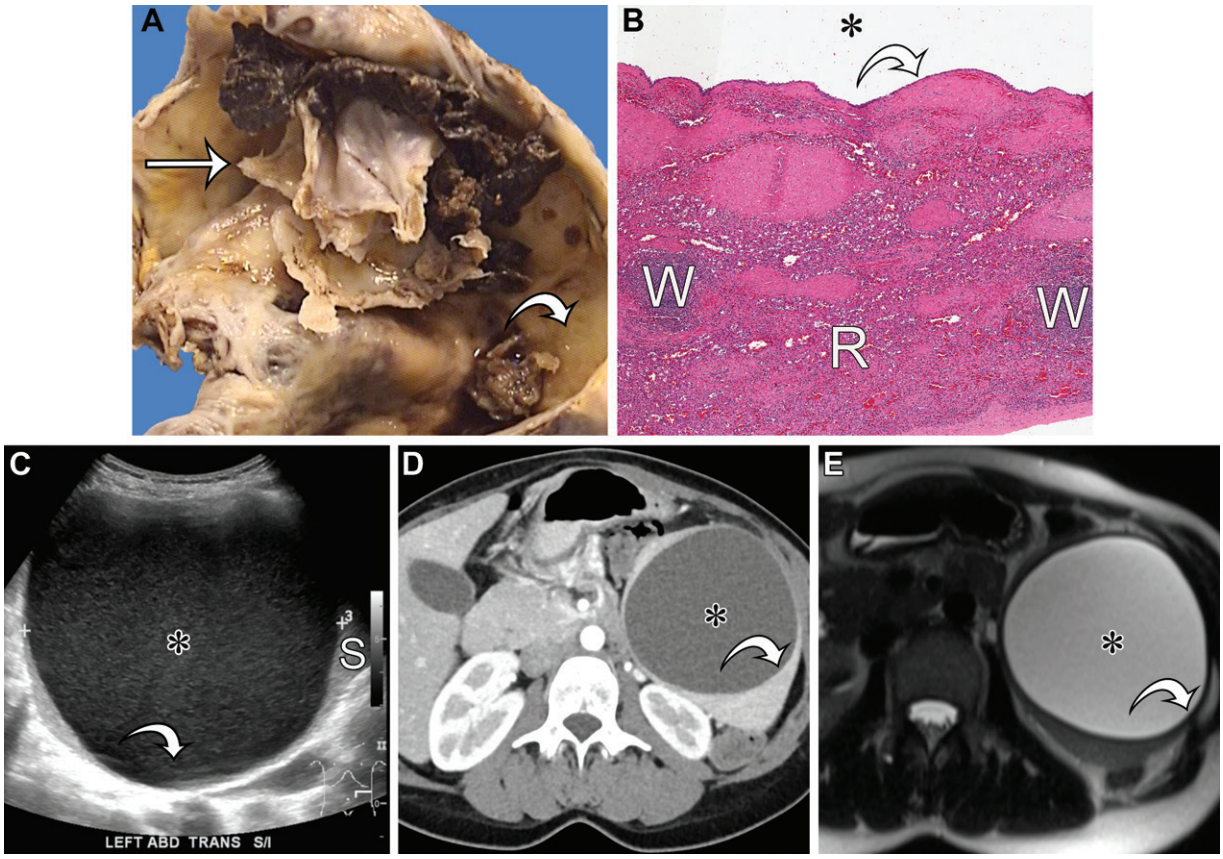

Figure 2. Epithelial cyst in a 28-year-old woman with left upper quadrant pain. **(A)** Photograph of the sectioned gross specimen shows a smooth glistening wall (curved arrow) with thick trabecular septa (straight arrow). **(B)** Photomicrograph shows the bland epithelial cellular lining (arrow) of the cyst. Note the normal spleen with red pulp *(R)* and white pulp *(W)*. ∗ = cyst lumen. (Hematoxylin-eosin [H-E] stain; original magnification, ×20.) **(C)** Transverse US image of the spleen shows a solitary, thin-walled, unilocular, hypoechoic cystic lesion with low-level echoes (∗) and a thin peripheral septum (arrow). *S* = normal spleen. **(D)** On an axial contrast-enhanced CT image, the cyst is well circumscribed with a thin imperceptible wall (arrow) and homogeneous fluid attenuation (∗). **(E)** On an axial T2-weighted image, the cyst is well circumscribed and thin walled (arrow) with homogeneous hyperintensity (∗).

Mycobacterium tuberculosis can result in abscesses due to hematogenous disseminated miliary spread.

Fungal microabscesses usually occur in patients with prolonged neutropenia. Predisposing factors include immunocompromise due to HIV infection, chemotherapy, immunosuppression for organ transplantation, or immunodeficiency. The most common fungal pathogens are *Candida*, *Aspergillus*, and *Cryptococcus* (10). Clinically, patients present with fever, abdominal pain, chills, constitutional symptoms, and sometimes septic shock (11).

Pathologic Features.—Grossly, abscesses are solitary or multiple, variably sized, red-yellow, rounded cystic lesions; pyogenic abscesses are usually larger than fungal or mycobacterial microabscesses (Fig E2). Mycobacterial nodules may contain caseous regions containing cheesy-white soft material. Abscess walls are thick and irregular (Fig E2). Infectious agents typically induce an acute inflammatory response, seen histopathologically as focal or diffuse leukocyte infiltration with large areas of necrosis (1). Fungal abscesses have a characteristic microscopic appearance, with central necrotic hyphae surrounded by viable hyphae and a peripheral inflammatory infiltrate rim. Specific microbiology is identified with culture and by using special stains.

Imaging Features.—Abscesses have ill-defined irregular margins with a central complex fluid component, due to the presence of pus, hemorrhage, or debris (Fig E2). Splenic pyogenic abscesses can be solitary or multiple. Gas appears as echogenic foci with "dirty" acoustic shadowing at US but is best documented at CT.

Fungal and mycobacterial microabscesses are usually multiple. US findings of fungal abscesses correlate with the histologic appearance, with a central hypoechoic nidus of necrotic hyphae surrounded by a hyperechoic concentric band (viable fungal elements), encased by a hypoechoic rim (zone of inflammation), with a so-called target or wheel-within-a-wheel pattern (3). At CT and MRI, disseminated fungal microabscesses appear as multiple small (<1 cm) hypoattenuating or

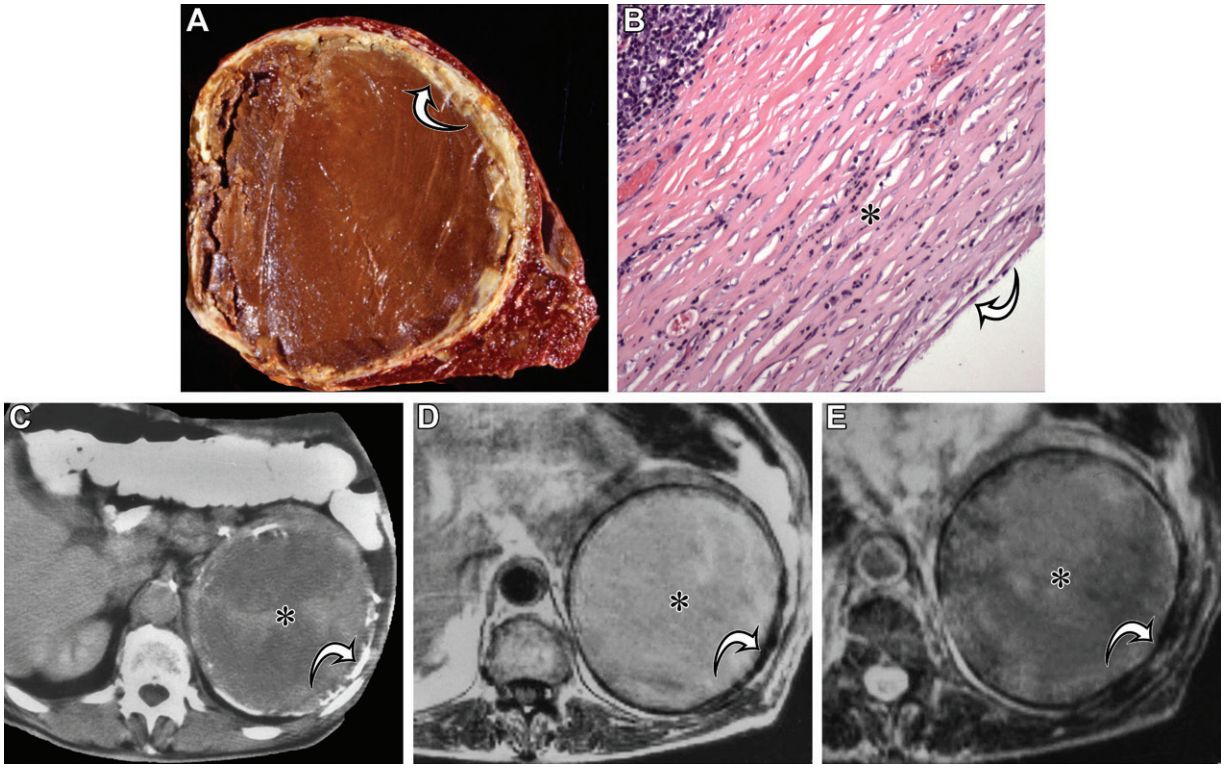

Figure 3. Pseudocyst in a 67-year-old man. **(A)** Photograph of the sectioned gross specimen shows a thick fibrous wall containing calcifications (arrow) surrounding thick debris. **(B)** Photomicrograph of the wall shows exuberant fibrosis and inflammatory infiltrate (*) without a true epithelial lining (arrow). (H-E stain; original magnification, ×200.) **(C)** Axial contrast-enhanced CT image clearly shows the calcifications (arrow); the pseudocyst is unilocular and contains heterogeneous mostly hypoattenuating fluid without enhancement (*). **(D, E)** On axial T1-weighted **(D)** and T2-weighted **(E)** images, the pseudocyst contains mixed-signal-intensity proteinaceous or hemorrhagic material (*). The thick wall (arrow) has circumferential hypointense signal, corresponding to the calcifications.

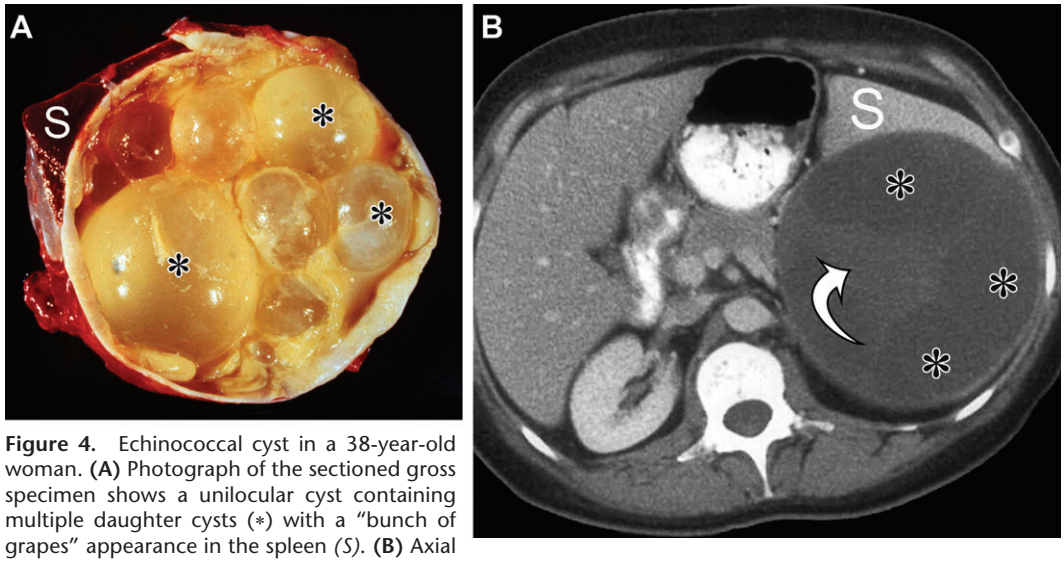

Figure 4. Echinococcal cyst in a 38-year-old woman. **(A)** Photograph of the sectioned gross specimen shows a unilocular cyst containing multiple daughter cysts (*) with a "bunch of grapes" appearance in the spleen *(S)*. **(B)** Axial contrast-enhanced CT image shows a well-circumscribed unilocular cyst containing multiple peripherally oriented hypoattenuating daughter cysts (*) separated by relatively hyperattenuating debris (arrow), also known as hydatid sand or condensed maternal fluid. This fluid will not enhance, which can help differentiate echinococcal cysts from septated multilocular cysts. *S* = normally enhancing spleen. (Case courtesy of Angela D. Levy, MD, Georgetown University Medical Center, Washington, DC.)

fluid-signal-intensity lesions scattered throughout the spleen (Fig 5) (10).

The subacute or chronic inflammatory infiltrate can be seen as a hypointense rim around these lesions with all MRI sequences, corresponding to iron-loaded macrophages (12). Acute microabscesses are not associated with avid rim enhancement, presumably owing to in-

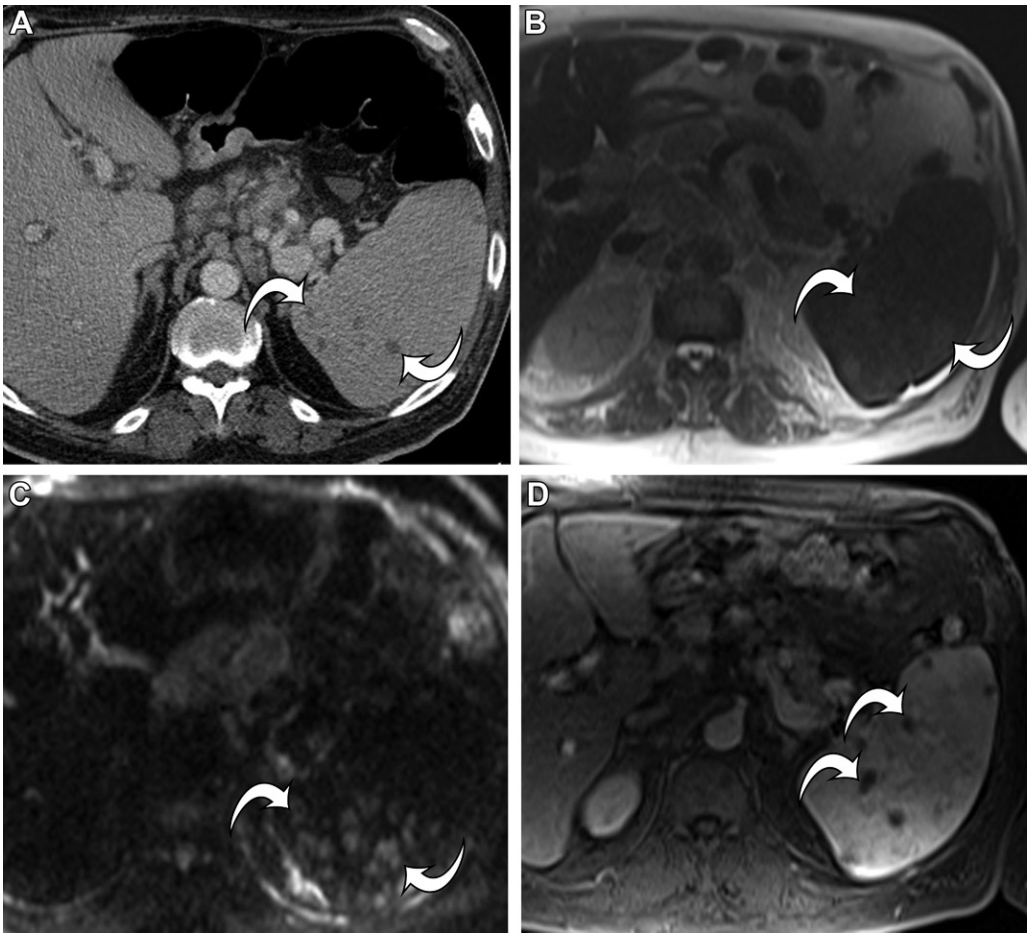

Figure 5. Fungal microabscesses in a 62-year-old patient with neutropenia. **(A)** Axial contrast-enhanced CT image shows multiple tiny hypoattenuating cystic lesions (arrows) scattered in the spleen, with central celiac axis lymphadenopathy. **(B)** Axial T2-weighted image shows subtle increased signal intensity in the lesions (arrows). **(C)** On an axial diffusion-weighted image, the lesions (arrows) are more readily visualized as hyperintense. **(D)** On an axial contrast-enhanced T1-weighted image, the lesions (arrows) do not enhance. (Case courtesy of Angela D. Levy, MD, Georgetown University Medical Center, Washington, DC.)

ability of the neutropenic patients to mount a robust inflammatory response; however, a subacute or chronic treated abscess can show peripheral rim enhancement at CT and MRI (12). Treated microabscesses appear as punctate calcifications at CT or foci of blooming artifact at gradient-echo (GRE) T1-weighted imaging (4). Abscesses resulting from endocarditis-related septic emboli can have a wedge-shaped appearance (10).

Mycobacterial involvement of the spleen most commonly manifests as splenomegaly and numerous tiny splenic lesions with ill-defined margins. Splenic involvement is seen in the context of widespread disease, with hypoattenuating adenopathy, nodular peritoneal thickening, high-attenuation ascites, and pleural effusions commonly present (10).

Lymphangioma

Clinical Features.—Lymphangiomas are rare malformations of splenic lymphatic channels characterized by anastomosing, dilated, thin-walled, fluid-filled cystic structures, similar to hemangiomas. Most occur in the pediatric population and are considered to reflect abnormal congenital development (13). An alternative hypothesis suggests that lymphangiomas reflect telangiectasia of lymphatic channels obstructed by hemorrhage or inflammation (14).

Like hemangiomas, splenic lymphangiomas can be solitary, multifocal, or diffuse. The clinical manifestation depends on the size; most are incidental and asymptomatic. However, large or multifocal lymphangiomas can result in splenomegaly, left upper quadrant pain, and nausea (14,15).

Pathologic Features.—The spleen can be normal or enlarged, depending on the size and number of lymphangiomas. Most isolated lymphangiomas are subcapsular; multifocal or diffuse lymphangiomas are more commonly seen in children (1). Grossly, lymphangioma looks like multiple thin-walled cysts of varying sizes filled with clear fluid

(Fig 6). Solid areas can be seen, reflecting central scarring (14,16).

Histologically, the cystic spaces are lined by bland flat endothelial cells, which may form papillary projections. The cysts are filled with watery eosinophilic proteinaceous fluid, which may contain histiocytes, cholesterol clefts, and lymphocytes (Fig 6). The intervening collagenous stroma can contain smooth muscle or calcification. Immunohistochemistry is necessary to confirm lymphatic derivation; the lymphatic endothelial lining will stain positive for D2-40 (podoplanin) antibody (14), less likely for CD34, CD31, and factor VIII.

Imaging Features.—Multimodality imaging reflects the gross appearance, with well-defined thin-walled unilocular or multilocular cysts (Figs 6, 7). Intervening fibrous stroma can appear as septa, with calcifications or progressive enhancement (Fig 7) (17). Intracystic debris or hemorrhage can appear as echoes at US or increased T1 signal intensity at MRI (13,15,17,18).

Solitary Solid Mass

Solitary solid splenic lesions include benign vascular entities (hemangioma, hamartoma, sclerosing angiomatoid nodular transformation [SANT]) and malignant vascular entities (angiosarcoma). Of these, hemangioma is by far the most common (1). However, lymphoma and benign nonvascular tumors and tumorlike lesions can also manifest as a solitary solid mass. Although definitive diagnosis may be difficult with imaging alone, it is important to differentiate the benign lesions from the malignant ones, such as lymphoma and angiosarcoma. Notably, benign lesions are almost always asymptomatic, whereas malignancies are rarely incidental or isolated findings (Table 2).

Hemangioma

Clinical Features.—Hemangioma is the most common benign lesion in the spleen (1). Most are found incidentally in asymptomatic patients. Usually solitary, hemangiomas can be multiple or diffuse; hemangiomatosis can reflect a manifestation of systemic angiomatosis in Klippel-Trénaunay syndrome (4,19). Kasabach-Merritt syndrome (anemia, thrombocytopenia, and coagulopathy) has been associated with large hemangiomas (Fig E3) (19).

Pathologic Features.—Grossly, hemangiomas are unencapsulated, red-purple, spongy nodules with irregular borders (Fig E3). Smaller hemangiomas can appear solid, whereas larger lesions can show prominent cystic changes. Fibrotic areas may contain calcium deposits.

Histologically, hemangioma is composed of variably sized vascular spaces lined by a single layer of bland flat endothelial cells (Fig E3). The cystic spaces can be small (capillary) but are most commonly large (cavernous) and distended with blood cells and proteinaceous fluid. The intervening septa are thin and fibrous; when abundant sclerosis is present, the lumina may be obliterated. Immunohistochemical analysis shows reactivity for vascular-associated markers (ERG, Wilms tumor protein 1 [WT1], CD31, CD34, factor VIII) but negativity for CD8, allowing distinction of hemangioma from splenic hamartoma (19).

Imaging Features.—Splenic hemangioma can show the well-described imaging characteristics of typical hepatic hemangioma: *(a)* well-defined hyperechoic mass without posterior acoustic shadowing at US; *(b)* well-circumscribed mass with peripheral nodular discontinuous early enhancement and homogeneous progressive enhancement at CT and MRI; and *(c)* homogeneous T2 hyperintensity at MRI (Fig E3). However, splenic hemangioma complicated by fibrosis, hemorrhage, or cystic degeneration can have a variable multimodality imaging appearance. Hemangioma rarely shows flow at color or power Doppler US. Although the contrast-enhanced US enhancement pattern has been studied, no defining pattern has been reliably identified (20).

As such, splenic hemangioma is best evaluated with dynamic contrast-enhanced CT or MRI (20). It can have variable dynamic enhancement patterns, including immediate homogeneous enhancement that persists in the delayed phase, early peripheral enhancement with homogeneous uniform delayed enhancement, or peripheral enhancement with centripetal progression (Fig 8) (21). Areas of cystic degeneration will not enhance, and areas of fibrosis can show progressive enhancement. Fluorodeoxyglucose (FDG) PET/CT can be performed for lesions with atypical features or for patients at high risk for malignancy or metastatic disease, as hemangiomas will not show metabolic activity.

Splenic Hamartoma

Clinical Features.—Splenic hamartoma is a rare (prevalence <1%) benign nodular malformation composed exclusively of disorganized red pulp without intervening white pulp, of uncertain pathogenesis; most consider it a congenital malformation, although true neoplasm of red pulp origin or reactive proliferative response to prior trauma are also possibilities (19). Also referred to as splenoma or focal nodular hyperplasia of the

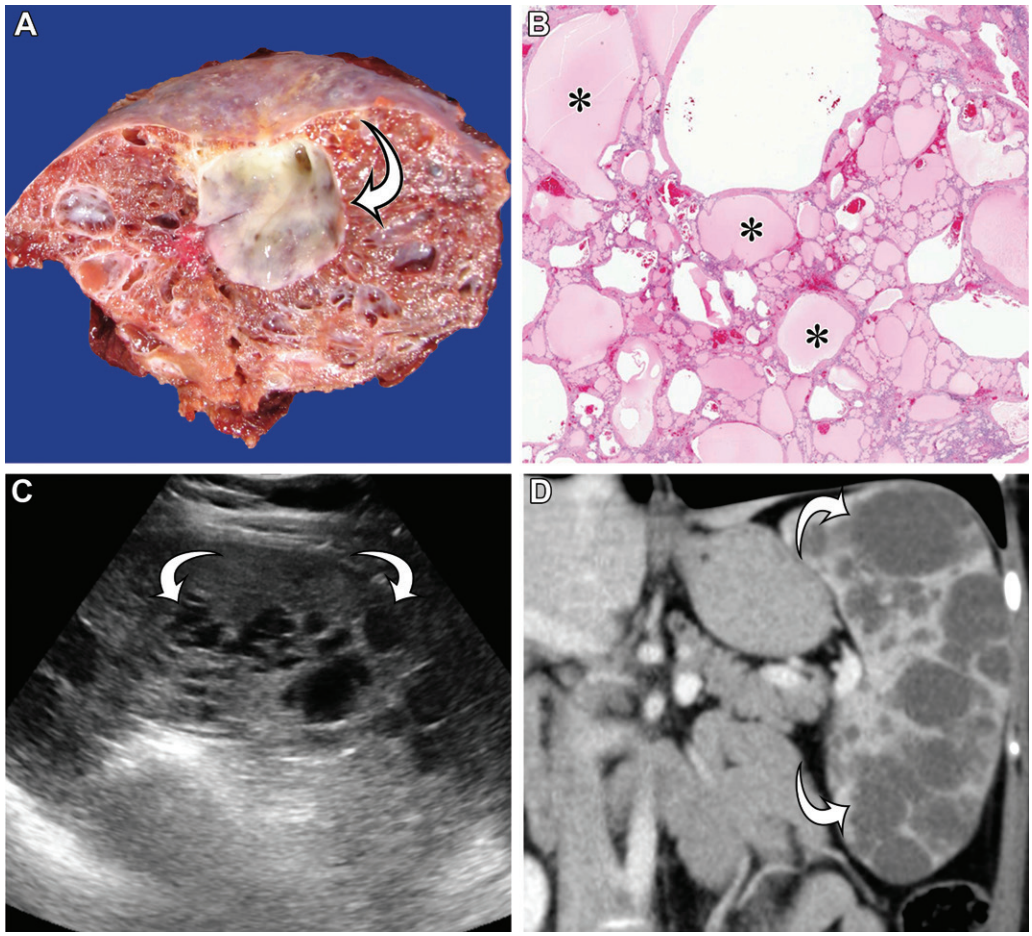

Figure 6. Lymphangioma in a 20-year-old woman with Klippel-Trénaunay-Weber syndrome and left upper quadrant abdominal pain. **(A)** Photograph of the cut gross specimen shows innumerable cystic spaces of various sizes throughout the spleen, including a larger cyst (arrow) with a thin fibrous wall. **(B)** Photomicrograph shows multiple cystic spaces lined by flat endothelial cells and containing acellular proteinaceous fluid (*). (H-E stain; original magnification, ×40.) **(C)** US image along the long axis of the spleen shows multiple unilocular or multilocular anechoic or hypoechoic spaces (arrows) of various sizes. **(D)** Coronal contrast-enhanced CT image shows enlargement of the spleen with numerous well-circumscribed, thin-walled, unilocular or multilocular hypoattenuating lesions (arrows) of various sizes without enhancement.

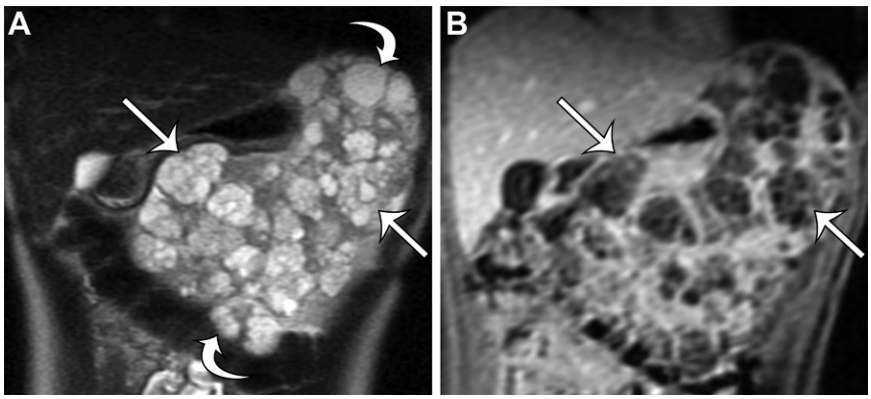

Figure 7. Lymphangioma in a 22-year-old woman with a history of Klippel-Trénaunay-Weber syndrome. **(A)** Coronal T2-weighted image shows numerous thin-walled unilocular or multilocular hyperintense cystic lesions (curved arrows) of variable sizes throughout the enlarged spleen, with hypointense septa seen within multiloculated lesions (straight arrows). **(B)** Coronal contrast-enhanced MR image during the equilibrium phase shows progressive enhancement of the septa within the multiloculated lesions (arrows).

spleen, hamartoma has no age or gender predilection and is most commonly found incidentally in asymptomatic patients.

Women are more likely to present with symptomatic larger lesions, suggesting a role for hormonal stimulation (19). Larger hamartomas can manifest with splenomegaly. Rarely, hematologic disorders including pancytopenia, anemia, or thrombocytopenia can occur from sequestration of hematopoietic cells. Splenic hamartoma has been associated with other hamartomatous entities, including tuberous sclerosis (19).

Table 2: Solid Splenic Lesions—Key Radiologic and Pathologic Features, by Number

Lesion	Nature	Key Imaging Features	Key Pathologic Features
Solitary solid vascular lesions			
Hemangioma	Benign	Usually solitary; rarely diffuse Typical: similar to in liver (hyperechoic and T2 hyperintense to spleen) Doppler US: flow rare; *CT and MRI: immediate homogeneous or progressive enhancement, with persistent delayed enhancement Large (cavernous): central myxoid nonenhancement Sclerosed: progressive enhancement of fibrosis *FDG PET/CT: no metabolic activity	Unencapsulated Variably sized vascular spaces Thin fibrous septa If abundant sclerosis, lumina may be obliterated CD8 negative (different from hamartoma)
Hamartoma	Benign	Rare, almost always solitary *Splenic focal nodular hyperplasia Well-circumscribed, usually large (bulging contour) US: hypoechoic with increased vascularity (different from hemangioma) Early arterial enhancement, progressively matching spleen	Unencapsulated Well demarcated Disorganized red pulp Variably sized vascular spaces CD8 positive (different from hemangioma)
SANT	Benign	Solitary well-circumscribed solid mass Nodular texture with fibrous rim and radiating bands *Characteristic spoke-wheel enhancement (early rim enhancement with progressive centripetal radial enhancement of fibrous bands and nodules *MRI: T2-hypointense radiating fibrous bands, T1 susceptibility artifact reflects hemosiderin	Red pulp entrapped by exaggerated stromal proliferation Confluent and discrete vascular nodules Dense fibrous rim, internodular myxoid-to-fibrous stroma coalesce to form central stellate scar
Angiosarcoma	Malignant	Discrete angiosarcoma: solitary to few well-circumscribed masses Diffuse angiosarcoma: splenomegaly, heterogeneous enhancement of irregular or poorly defined nodular vascular masses Hemorrhage ± necrosis more common than cystic appearance	Arises from endothelial cells spread throughout spleen: well-circumscribed solitary nodules to diffusely infiltrating masses Markedly atypical cells High mitotic index CD31 positive
Solitary solid nonvascular lesions			
Lymphoma	Malignant	*Most common malignant tumor involving spleen, usually secondary (primary splenic lymphoma is rare) Variable appearance depending on subtype: splenomegaly, tiny diffuse miliary nodules, small nodules, or large solitary mass Homogeneous; hypoechoic, hypoattenuating, and hypoenhancing to spleen *Restricted diffusion; avid activity at FDG PET/CT	Variably sized neoplastic lymphocytes Nodules in white pulp to diffuse sheetlike infiltration Immunohistochemistry crucial to determine specific lineage
Inflammatory pseudotumor (IPT)	Benign	Well-circumscribed mass Inflammatory infiltrate: hypoechoic, hypoattenuating, and hypoenhancing to spleen *Central fibrous scar: T2 hypointense and progressively enhancing	Mixed inflammatory cells and spindle cell proliferation Interspersed fibrous stroma Necrosis, hemorrhage, sclerosis
Multiple solid lesions			
Inflammatory (sarcoidosis)	Benign	Numerous tiny hypoechoic, hypoattenuating, and hypoenhancing solid nodules *Hypointense with all MRI sequences; best seen on T2-weighted fat-suppressed and postcontrast images	Multiple well-defined noncaseating granulomas
Littoral cell angioma (LCA)	Benign	*Numerous progressively enhancing nodules Variable echogenicity Variable T2 SI (increased for hypercellular LCA, decreased when hemosiderin present)	Unencapsulated, multiple Anastomosing vascular channels ± papillary fronds CD8 negative (different from normal littoral cells)
Metastases	Malignant	Uncommon, *found almost exclusively in patients with known or widespread cancer Variable appearance depending on primary tumor	Variable: dependent on primary tumor

Note.—FDG = fluorodeoxyglucose, SI = signal intensity, ± = with or without, * = key findings for each entity.

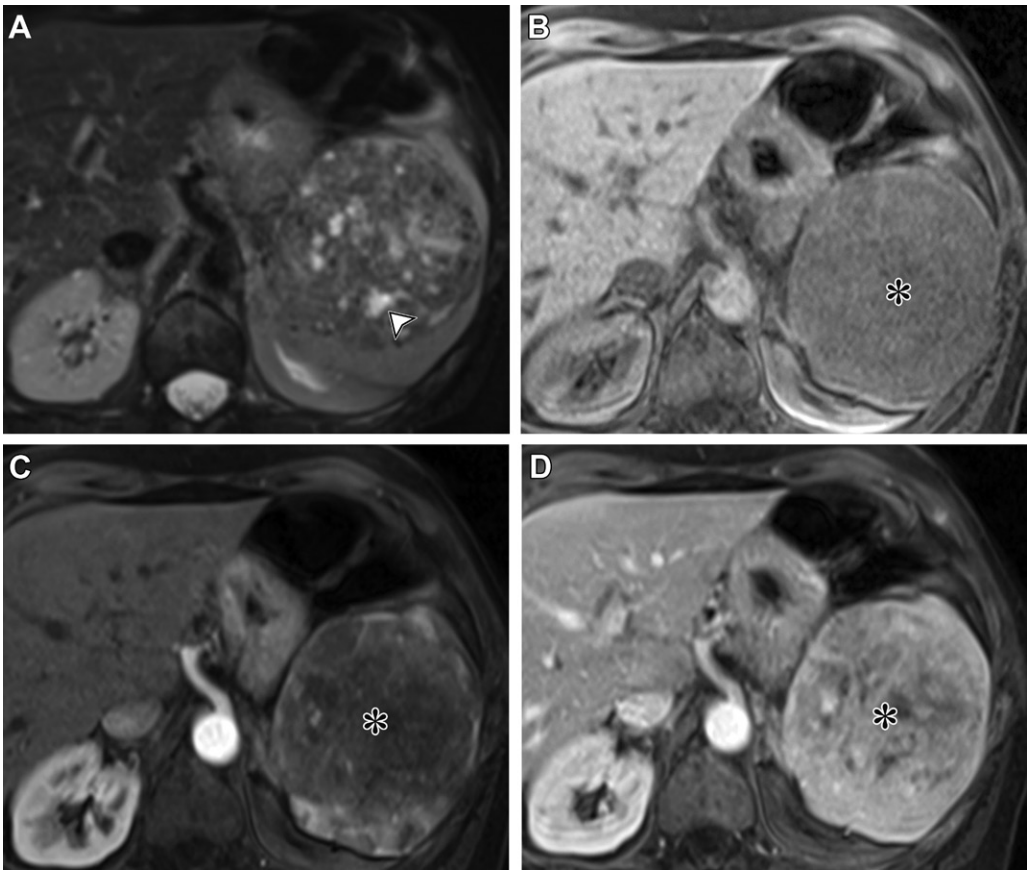

Figure 8. Hemangioma in a 58-year-old woman with progressive sporadic vomiting and sharp left upper quadrant pain. **(A)** Axial fat-saturated T2-weighted image shows a well-circumscribed solid mass with areas of hyperintense signal (arrowhead). **(B)** Axial noncontrast T1-weighted image shows that the mass (*) is isointense. **(C)** Axial arterial phase image shows early peripheral enhancement of the mass (*). **(D)** Axial portal venous phase image shows progressive homogeneous enhancement of the mass (*).

Pathologic Features.—Hamartoma is almost always solitary and usually 4–5 cm (range, 1–20 cm). Grossly, it is a well-circumscribed, unencapsulated, bulging dark-red lesion well demarcated from the uninvolved spleen (Fig 9). Histologically, hamartoma is composed of red pulp without normal white pulp elements. There are numerous disorganized sinuses and cords lined by plump endothelial cells. Histologic variations have been described on the basis of dominant expression of one or more components of the red pulp (19,22).

Occasionally, large and bizarre stromal cells are present (1). Variable inflammatory cells are present without well-organized lymphoid follicles (in contrast to normal spleen and hemangioma) (22). Fibrosis, hemorrhage, and calcifications may be present, particularly in long-standing lesions. Extramedullary hematopoiesis may be present. Positive immunohistochemical staining of the endothelial lining of the vascular channels with CD8 is a key distinguishing feature from hemangioma (1,19).

Imaging Features.—Reflecting their pathologic appearance, most hamartomas are well-circumscribed solid vascular masses similar to the splenic red pulp at multimodality imaging. At US, they are typically homogeneous well-demarcated masses, hyperechoic to the normal background spleen when not complicated by hemorrhage or cystic change (Fig 9). Difficult to identify at noncontrast CT, as they are isoattenuating to the adjacent splenic parenchyma, larger lesions may show contour abnormality as the only noticeable finding (Fig 9).

At MRI, hamartoma is T1 isointense and mildly T2 hyperintense to the spleen (Fig 9) (23). The hypervascular red pulp component manifests as increased blood flow at color Doppler US (Fig 9) (23,24). Dynamic contrast-enhanced CT and MRI show early arterial and persistent delayed enhancement, equilibrating with the background spleen (Fig 9) (23). Calcifications and cystic changes can be present (21,23).

Sclerosing Angiomatoid Nodular Transformation

Clinical Features.—SANT is a morphologically distinctive benign vascular lesion. Although the term was first coined in 2004, similar lesions

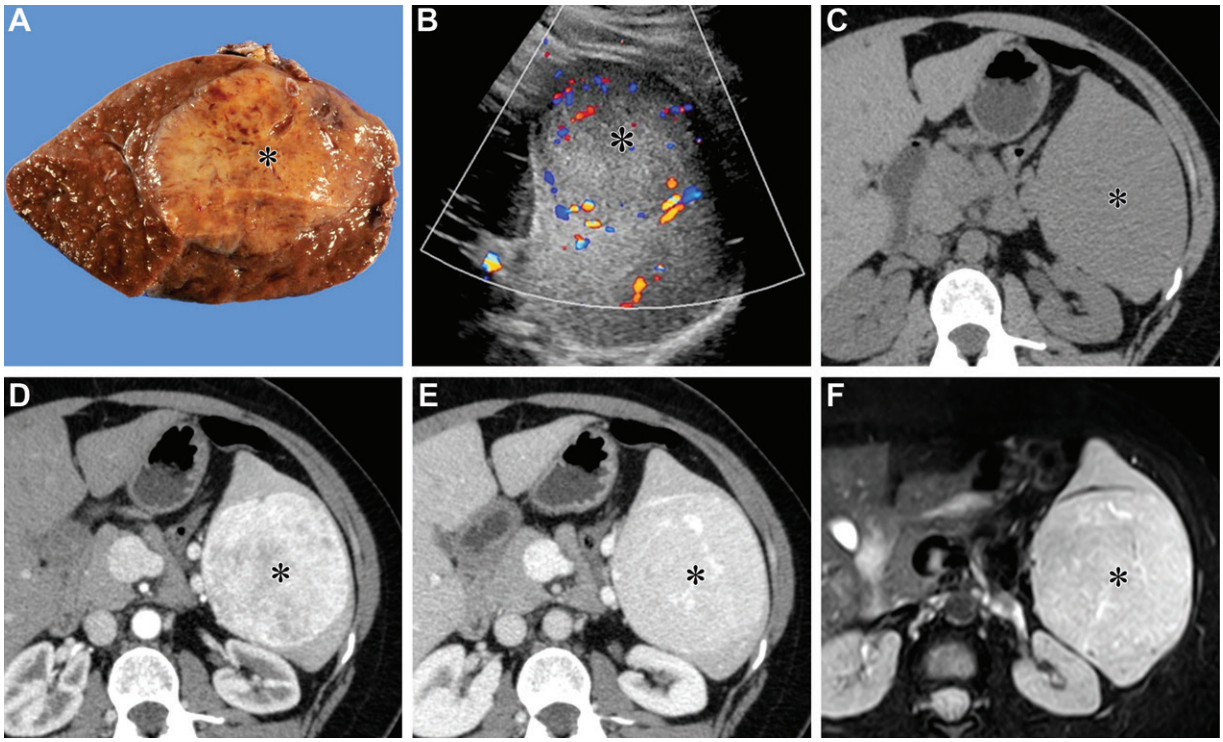

Figure 9. Pathologic and radiologic features of hamartoma. **(A)** Photograph of the cut gross specimen shows a well-circumscribed nonencapsulated mass (*) bulging above the cut surface of the spleen. **(B)** Color Doppler image shows a well-circumscribed homogeneous mass (*) with intrinsic flow. **(C)** Axial nonenhanced CT image shows only slight contour abnormality of the mass (*). **(D)** Axial arterial phase CT image shows diffuse early hyperenhancement of the mass (*). **(E)** Axial portal venous phase CT image shows persistent delayed enhancement of the mass (*), equilibrating with that of the spleen. **(F)** Axial T2-weighted image shows that the mass (*) is mildly hyperintense to the spleen. The MRI dynamic enhancement pattern is similar to that of CT.

were described earlier as capillary cord hemangioma, multinodular hemangioma, and splenic hemangioendothelioma (1,17,19,25). The pathogenesis remains unclear; however, on the basis of histopathologic features, it is thought to reflect altered red pulp entrapped by an exaggerated stromal proliferation due to inflammation, hemorrhage, or traumatic injury. Primarily discovered incidentally in asymptomatic patients, SANT has been reported concurrently with other malignancy, although most consider this coincidental during routine surveillance imaging (26). Patients can rarely present with anemia, splenomegaly, or abdominal pain (25,27).

Pathologic Features.—Grossly, SANT is a solitary, well-circumscribed, unencapsulated mass of individual and coalescing red-brown nodules embedded in a dense fibrous stroma, often with a central tan-white stellate scar (Fig 10) (19,25–27). Histologically, the angiomatoid nodules are composed of red pulp, with all three immunohistochemically distinct types of blood vessels (veins, capillaries, and sinusoids) seen in the normal red pulp, a distinguishing characteristic from hemangioma, hamartoma, and littoral cell angioma (LCA), which contain only one type of vascular channel (25,26). The internodular stroma is composed of myxoid-to-dense fibrous stroma, with sclerosis or fibrinoid acellular material, and scattered myofibroblasts and inflammatory cells (Fig 10) (1).

Imaging Features.—The multimodality imaging features of SANT have been described in the literature in several case reports and generally parallel the gross appearance as a solitary well-defined mass (26,28–31). A characteristic appearance at contrast-enhanced US, CT, and MRI is early peripheral rim enhancement with radiating bands of progressive centripetal enhancement, described as a spoke-wheel appearance (Fig 10) (26,28–30). At MRI, radiating T2-hypointense bands extending toward the center of the mass are thought to reflect the fibrous stroma (Fig 10) (26,31). Susceptibility artifact can indicate the presence of hemosiderin (26).

Angiosarcoma

Clinical Features.—Although it is the most common primary nonhematolymphoid malignant splenic tumor, angiosarcoma is rare (19). Patients present almost exclusively after 40 years of age with abdominal pain and other constitutional symptoms of malignancy, such as weight loss, fever, and fatigue. Up to 30% of patients

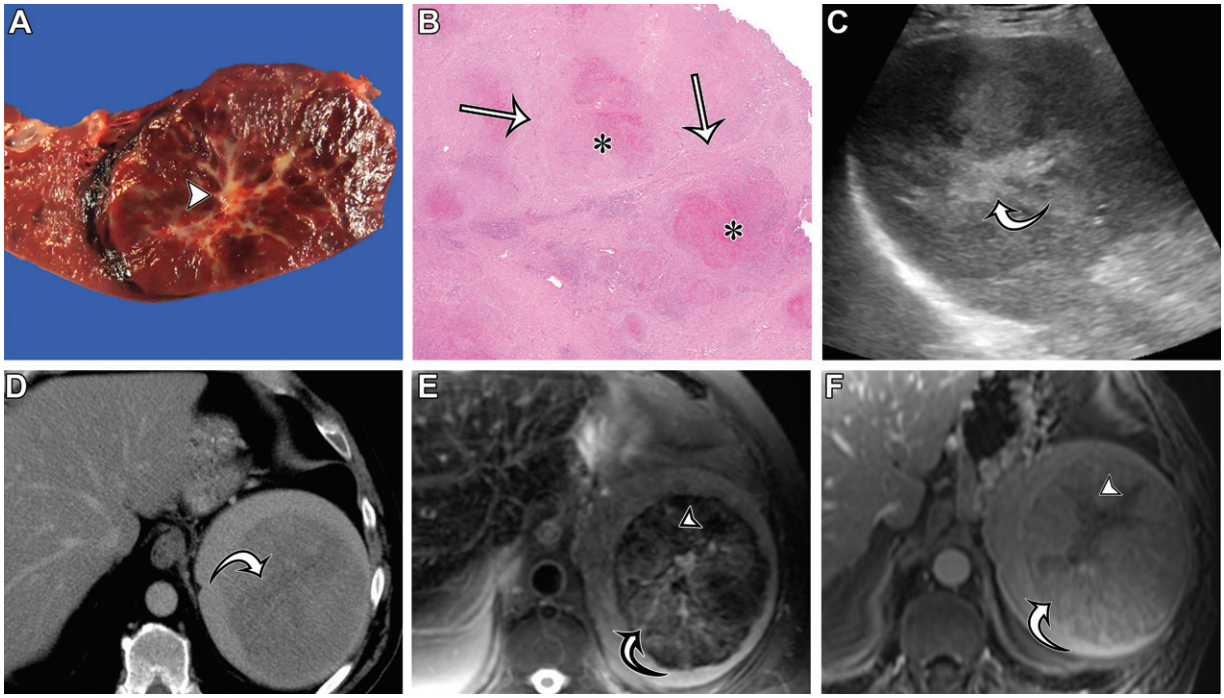

Figure 10. Pathologic and radiologic features of SANT. **(A)** Photograph of the cut gross specimen shows a well-circumscribed firm multinodular mass composed of red-brown nodules, with a central stellate tan-white scar (arrowhead). **(B)** Photomicrograph shows angiomatoid nodules similar to red pulp (*) with internodular bandlike fibrous stroma (arrows). (H-E stain; original magnification, ×10.) **(C)** On a US image, the central stellate scar (arrow) is echogenic. **(D)** Axial contrast-enhanced CT image shows a predominantly hypoenhancing mass with a hypoattenuating linear stellate scar (arrow). **(E)** Axial fat-suppressed T2-weighted image shows the mass to be composed of hypointense angiomatoid nodules (arrowhead) with intervening radiating linear hyperintense bands (arrow), which correlate with internodular fibrous stroma. **(F)** Axial contrast-enhanced MR image shows enhancement of the angiomatoid components around the periphery of the mass (arrow), similar to the background red pulp, and mild progressive enhancement along the radiating fibrous bands (arrowhead). Progressive central enhancement is thought to reflect progressive enhancement of the angiomatoid nodules combined with progressive enhancement of the fibrous bands.

can present with spontaneous splenic rupture (32). Anemia, thrombocytopenia, and leukocytosis can be seen (1,32). Often disseminated at diagnosis, with metastases to the liver, lung, lymph nodes, and bone at initial presentation, splenic origin can be difficult to prove (1,32).

Pathologic Features.—Primary splenic angiosarcoma arises from endothelial cells, which are abundant throughout the red pulp; as such, these neoplasms can manifest grossly as solitary well-circumscribed firm nodules to diffuse poorly defined purple-red necrotic and hemorrhagic masses (Fig 11) (19,32,33). The spleen is usually enlarged. Microscopically, there are disorganized, irregular, anastomosing vascular channels with atypical endothelial cells (bland or spindle shaped), high mitotic index, and hyperchromatic nuclei.

Papillary projections covered by malignant endothelial cells may protrude into the vascular cystic spaces. At immunostaining, angiosarcoma stains positive for endothelial markers, including ERG, Wilms tumor protein 1 (WT1), and CD31 (CD31 is considered the most sensitive marker) (33). Fibrosis, hemorrhage, hemosiderin, and calcifications may be present.

Imaging Features.—Concomitant with the variable pathologic appearance of angiosarcoma, imaging features are heterogeneous and nonspecific. The spleen is often enlarged, and when infiltrated by diffuse angiosarcoma, will enhance heterogeneously. Discrete angiosarcomas appear as a large dominant mass or multiple masses.

As hemorrhage and cystic necrosis are common, angiosarcoma can appear cystic. Increased flow in the bizarre and dilated vascular channels can be shown at color Doppler US (34). At CT and MRI, angiosarcomas can be solitary or multiple, poorly defined, heterogeneous nodular masses—due to the presence of both solid components and areas of hemorrhage and necrosis—with heterogeneous nodular enhancement of solid components and vascular channels (Fig 11) (35–37).

Lymphoma

Clinical Features.—Lymphoma is the most common malignant tumor involving the spleen. Primary splenic lymphoma—defined as lymphoma confined to the spleen and perisplenic lymph nodes—is rare, comprising less than 1% of all lymphomas (1). Most patients present with

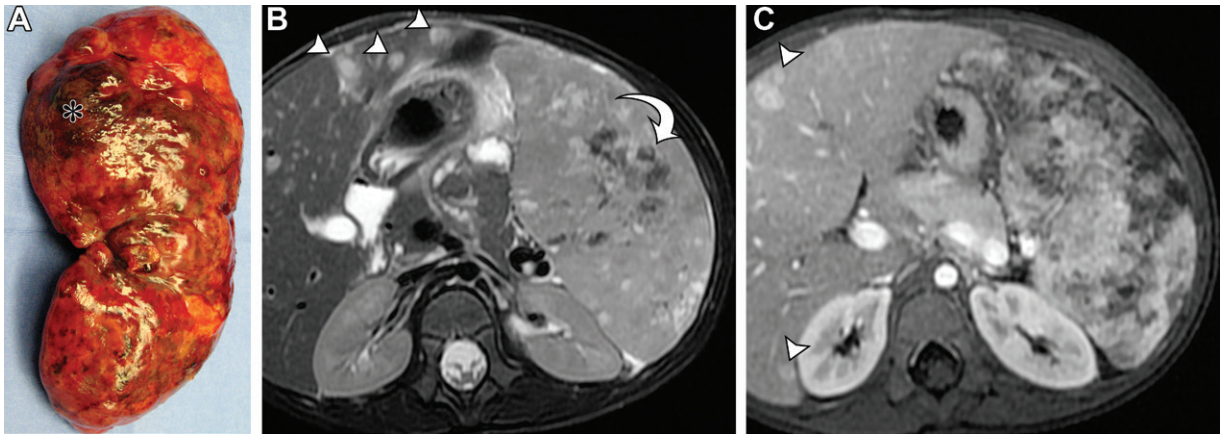

Figure 11. Angiosarcoma in a 72-year-old man with intermittent fever, unintentional weight loss, and persistent dull left upper quadrant pain after a fall. **(A)** Photograph of the gross specimen shows an enlarged nodular spleen with multiple purple-red masses (*). **(B, C)** Axial T2-weighted **(B)** and contrast-enhanced T1-weighted **(C)** images show multiple masses with heterogeneous signal intensity throughout the enlarged spleen, with central T2-hypointense areas of hemorrhage (arrow in **B**) and heterogeneous bizarre nodular enhancement of the solid components and vascular channels. Multiple T2-hyperintense and enhancing masses are also seen in the liver (arrowheads).

classic systemic symptoms of lymphoma including weight loss, fatigue, and fever. Patients with splenic involvement can present with left upper quadrant pain and splenomegaly.

Pathologic Features.—The wide spectrum of lymphomas (B- and T-cell lymphomas, Hodgkin disease) can lead to myriad gross appearances, including splenomegaly without discrete masses, tiny diffuse miliary nodules, small nodular masses, or a large solitary mass (Fig 12) (1). Microscopically, variably sized neoplastic lymphocytes localize as nodules in the white pulp or as diffuse sheetlike infiltration. Immunohistochemistry is crucial to determine the specific lineage of the tumor.

Imaging Features.—The imaging features of splenic lymphoma reflect the pathologic appearance and can manifest as splenomegaly, diffuse infiltration, multiple small or large nodules, or a solitary mass. As elsewhere, lymphomatous nodules are usually homogeneous and hypovascular. At US, lymphomas are hypoechoic to the adjacent spleen and show scattered penetrating vascularity at color Doppler US (Fig E4) (38,39). At CT, they are low-attenuation solid lesions, hypoenhancing to the adjacent splenic parenchyma (Fig 12).

At fluorodeoxyglucose (FDG) PET/CT, splenic lymphoma is FDG avid (standardized uptake value [SUV] = 6.9 ± 7.9) (40,41). At MRI, lymphoma is usually T1 hypointense and has variable T2 signal intensity (Fig E4). Lymphoma demonstrates increased restricted diffusion compared with adjacent normal splenic parenchyma. Postcontrast images may show mild enhancement, although the lesion is typically hypoenhancing to normal spleen (42).

Inflammatory Pseudotumor

Clinical Features.—The term *inflammatory pseudotumor* (IPT) has been used to describe a reactive tumorlike lesion found throughout the body, rarely in the spleen. Originally described in 1984, splenic IPT is most often seen in middle-aged or older patients (43,44). Splenic IPT is strongly associated with Epstein-Barr virus (EBV) infection. IPT-like follicular dendritic cell (FDC) tumor is the most frequent subgroup of EBV-associated IPT (45).

Patients can have a wide range of clinical presentations, from asymptomatic to left upper quadrant or epigastric pain, fever, weight loss, and splenomegaly. Anemia, thrombocytopenia, hypergammaglobulinemia, and elevated levels of inflammatory markers have also been described (46,47).

Pathologic Features.—Grossly, splenic IPT is a firm well-circumscribed yellow-white or tan mass. Areas of necrosis, hemorrhage, or sclerosis can be grossly visible (Fig 13) (45). Histopathologically, it is composed of a mixture of inflammatory cells and spindle cell proliferation, with interspersed collagenous stroma (43). The spindle cells are positive for EBV (1).

Imaging Features.—At US, IPT can appear as a solitary, well-circumscribed, hypoechoic, solid masslike lesion (48). Lesions are isoattenuating at noncontrast CT but are better seen after contrast material administration as a large well-circumscribed hypoenhancing solitary mass, with progressive enhancement of the fibrous component (Fig 13) (44). Calcifications may be present (Fig 13). At MRI, the lesion is T1 isointense to hypointense with heterogeneous signal intensity on T2-weighted images, frequently

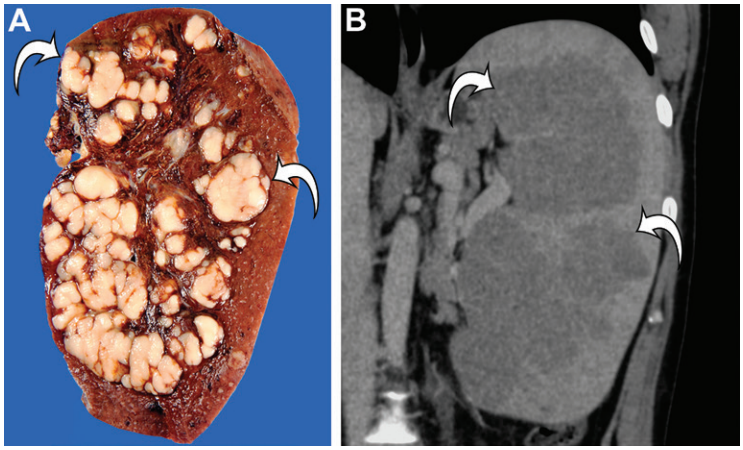

Figure 12. Lymphoma in a 37-year-old man with fatigue, weight loss, early satiety, and massive splenomegaly. **(A)** Photograph of the sectioned resected enlarged spleen shows near-complete replacement of the splenic parenchyma with innumerable well-defined, solid, tan nodules (arrows). **(B)** Coronal contrast-enhanced CT image best shows splenomegaly and innumerable homogeneously hypoattenuating nodules (arrows), which are hypoenhancing to the residual normal spleen.

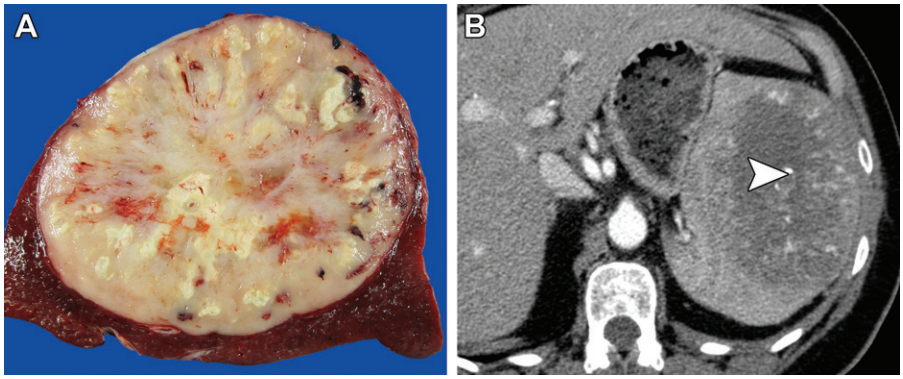

Figure 13. IPT in a 59-year-old woman with chest pain. **(A)** Photograph of the sectioned gross specimen shows a well-circumscribed yellow-white mass. **(B)** Axial contrast-enhanced CT image during the portal venous phase shows a well-defined solitary hypoenhancing mass with central punctate calcifications (arrowhead).

with a central stellate low-signal-intensity scar. A consistently reported MRI feature is delayed enhancement, presumably due to abundant fibrous stroma (48,49).

Multiple Solid Masses

Rarely are multiple solid splenic masses identified incidentally; most patients will have symptoms or splenic disease will be expected clinically. The most frequent source is diffuse inflammatory disease (sarcoidosis) or infectious disease (microabscesses, discussed earlier as multiple cystic masses). Other considerations for multiple solid masses include lymphoma, LCA, and metastases. Although LCA is typically incidentally found in asymptomatic patients, lymphoma and metastases are associated with symptoms or widespread primary malignancy (Table 2).

Inflammatory Lesions

Clinical Features.—Sarcoidosis, a systemic granulomatous disease of unclear cause, is an inflammatory process involving the spleen. Splenic involvement is often asymptomatic but can manifest as splenomegaly. More frequent sites of involvement include the lung and mediastinal and hilar lymph nodes.

Pathologic Features.—Grossly, sarcoidosis appears as multiple solid nodules, which can coalesce to form larger masses (Fig 14). Histologic features include well-defined granulomas with prominent epithelioid macrophages and intervening red pulp (Fig E5). A few small foci of necrosis may be present. Older granulomas can show fibrosis and hyaline change. The presence of giant cell inclusions—Schaumann bodies (with basophilic spherules composed of lipomucoglycoproteins) and asteroid bodies (stellate eosinophilic inclusions)—is suggestive of sarcoidosis, although it can be seen in other granulomatous diseases.

Imaging Features.—Multimodality imaging may show splenomegaly or multiple nodules (50). At US, discrete nodules are hypoechoic; when nodules are ill defined and multiple, the spleen may appear diffusely heterogeneous (Fig E5). Although difficult to delineate at noncontrast CT, splenic sarcoid nodules are hypoenhancing to adjacent normal splenic parenchyma at contrast-enhanced CT, best seen in earlier phases (Fig 14) (50). At MRI, lesions are hypointense with all sequences and hypoenhancing but are most conspicuous on T2-weighted fat-suppressed and postcontrast images (Figs 14, E5) (4,51).

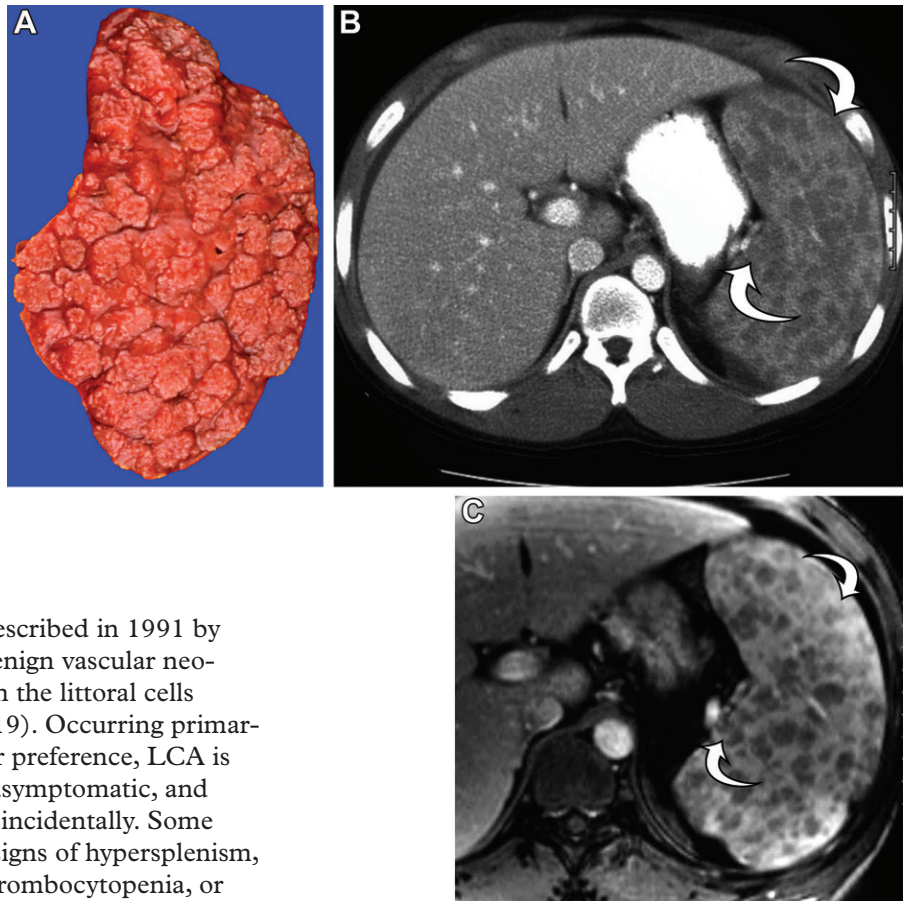

Figure 14. Sarcoidosis in a 38-year-old woman with pulmonary manifestations. **(A)** Photograph of the cut resected spleen shows near-complete replacement of the splenic parenchyma with variably sized solid discrete or confluent nodules. **(B, C)** Axial contrast-enhanced CT **(B)** and T1-weighted **(C)** images show innumerable hypoenhancing solid nodules (arrows) throughout the spleen.

Littoral Cell Angioma

Clinical Features.—First described in 1991 by Falk et al (52), LCA is a benign vascular neoplasm thought to arise from the littoral cells lining the splenic sinuses (19). Occurring primarily in adults without gender preference, LCA is benign. Most patients are asymptomatic, and most tumors are identified incidentally. Some patients may present with signs of hypersplenism, including splenomegaly, thrombocytopenia, or anemia (19,53).

Pathologic Features.—LCAs appear as multiple spongy and cystic nodules well delineated from the background splenic parenchyma, ranging from red to black (Fig E6). Histologically, LCAs have ill-defined boundaries, without a defined capsule, and are composed of multiple irregular cystic vascular channels lined by plump cuboidal endothelial cells (Fig E6). The vascular channels can have irregular lumina with papillary projections into the vascular space. Abundant detached littoral cells and desquamated macrophages from the lining endothelium may be present in the vascular spaces (19). At immunostaining, the cells are positive for endothelial markers (CD31, factor VIII–related antigen) but consistently negative for CD8 (unlike normal littoral cells).

Imaging Features.—The most consistent imaging manifestation of LCA is splenomegaly with numerous progressively enhancing nodules (54). Variable US characteristics have been reported, including diffusely mottled echotexture without discrete lesions as well as multiple hypoechoic, isoechoic, and hyperechoic nodules (55,56). At CT, the most commonly reported appearance is multiple hypoattenuating lesions, initially hypoenhancing in the early and portal venous phases, with homogeneous isoenhancement to background spleen on delayed phase images (Fig 15) (54,57). Small case reports have described the MRI features as consistently T1 hypointense with progressive enhancement (Fig 15). Reported T2 imaging features are more variable, with some authors describing increased signal intensity—likely reflecting cellularity—and others reporting decreased signal intensity, presumed to be due to the presence of hemosiderin (56,58).

Metastasis

Clinical Features.—Splenic metastases are uncommon and usually seen in the context of widespread disease. Metastases can be cystic or solid and solitary or multiple. The route of spread is thought to be hematogenous, and the most common tumors to metastasize to the spleen are breast cancer, lung cancer, ovarian cancer, melanoma, and colon cancer (3).

Pathologic Features.—Grossly, the spleen is usually normal in size with single or multiple foci of metastatic lesions. The histologic appearance of cystic metastases will mimic that of the original tumor (3). Immunohistochemistry is often used

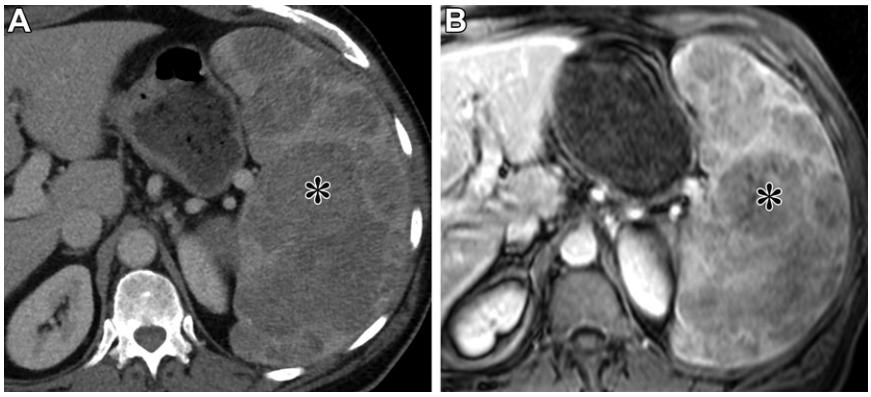

Figure 15. LCA in a 65-year-old man with chronic thrombocytopenia. Axial contrast-enhanced CT **(A)** and T1-weighted **(B)** images in the portal venous phase show splenomegaly with multiple hypoenhancing lesions (*).

to phenotype the metastatic tumor, especially with poorly differentiated metastases. Cytokeratins typically highlight carcinomas, and SOX10, S100, BRAF, human melanoma black–45 (HMB-45), and microphthalmia transcription factor (MITF) will stain positive for melanoma.

***Imaging Features.*—**At US, splenic metastases are predominantly hypoechoic but can be of mixed echogenicity or hyperechoic (3). At CT, splenic metastases appear as hypoattenuating or cystic lesions with variable enhancement (3). At MRI, metastases are typically T2 hyperintense and T1 hypointense, with variable enhancement depending on the primary tumor type (4).

Treatment and Prognosis

For asymptomatic patients with splenic lesions that can be characterized as benign at imaging—such as nonparasitic cyst, pseudocyst, lymphangioma, hemangioma, hamartoma, and SANT—no treatment is necessary and clinical observation, possibly with imaging follow-up, is sufficient (59). However, larger cysts, pseudocysts, and lymphangioma can cause symptoms or become complicated by intracystic hemorrhage or rupture, while hemangioma, hamartoma, and SANT can lead to hypersplenism or increase the risk for splenic hemorrhage or rupture, requiring surgical intervention (14,19,59).

To preserve splenic function in patients with symptomatic cysts or pseudocysts, minimally invasive surgical techniques—including marsupialization, fenestration, or partial cystectomy—have been performed with good outcomes (59,60). For larger or symptomatic cysts and lymphangioma, conventional treatment is total or partial splenectomy. Conservative treatment methods such as aspiration, drainage, and sclerosis are not recommended for lymphangioma owing to high risk of recurrence (14). For symptomatic or complicated hemangioma, hamartoma, and SANT, partial or total splenectomy is a surgical option; partial splenectomy may be appropriate for patients with single lesions (61–63).

Patients with infectious diseases of the spleen—including bacterial, fungal, and parasitic abscesses—are often symptomatic; the diagnosis can be established with clinical, laboratory or serologic, and imaging features. Percutaneous aspiration can help confirm the diagnosis and direct therapy. Untreated abscesses have a poor prognosis; the mortality rate is significantly improved with treatment, which consists of combined medical and surgical options (64). Image-guided percutaneous drainage has been successfully employed as spleen-sparing treatment of abscess (65). Splenectomy may be necessary when targeted medical therapy fails.

For high-surgical-risk patients with hydatid disease, percutaneous aspiration and injection of scolicidal agents is an option, although with increased risk of potential spillage and peritoneal dissemination (64). Retained laminated membranes increase risk for recurrent or secondary infection (59,64). Spleen-sparing surgical options include hydatid cyst enucleation or unroofing with omentoplasty; however, unroofing leaves the pericyst layer, increasing risk of postoperative infection. Total splenectomy is curative (66).

When the diagnosis cannot be established confidently with imaging alone, tissue sampling is required, which can be performed with total or partial splenectomy, historically the mainstay of diagnosis and treatment (19). At many institutions, image-guided percutaneous splenic biopsy has been avoided owing to concern about hemorrhage and the misperception of low diagnostic accuracy (67). However, a systematic review and meta-analysis of the complications and accuracy of percutaneous image-guided biopsy using smaller needles (18-gauge maximum) showed a low major complication rate (2.2%), comparable with published complication rates for liver and kidney biopsy, with high diagnostic accuracy (sensitivity = 87%, specificity = 96%) (67).

Core needle biopsy can be helpful for benign entities for which prospective imaging diagnosis is difficult, such as SANT, LCA, and IPT, preventing unnecessary splenectomy (45,47,62,63,67–70). When necessary, partial splenectomy can be performed for definitive diagnosis and curative treatment of localized tumors (71). IPT-like follicular dendritic cell (FDC) tumor is considered a low-grade malignant lesion, requiring splenectomy and postresection surveillance (45,47,68).

Angiosarcoma is a highly aggressive tumor with a poor prognosis; most patients die within 12 months of diagnosis irrespective of treatment (32,33). Treatment options for lymphoma are stage dependent but usually include chemotherapy. Splenic metastases typically occur in widespread metastatic disease, where surgery may not be an option. In rare cases of isolated splenic metastases, splenectomy is the treatment of choice (72,73).

Conclusion

Splenic lesions are often unexpected findings at imaging and can present a diagnostic challenge. Using a stepwise approach by first determining if they are cystic or solid, then further characterizing them as solitary or multiple lesions, can help in diagnosis or providing a proper differential diagnosis (Fig 1).

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