Splenic Lesions



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Abbreviations

Three dimensional		
Acquired immunodeficiency syndrome		
Centimeter		
Computed tomography		
Diffusion-weighted imaging		
Fluorodeoxyglucose		
Gallium		
Human immunodeficiency virus infection		
Indium		
Millimeter		
Multiplanar reconstruction		
Magnetic resonance		
Magnetic resonance imaging		
Positron emission tomography		
Single-photon emission computed		
tomography		

STIR	Short TI inversion recovery		
SUV	Standardized uptake value		
Tc	Technetium		
WB-DWI	Whole body diffusion-weighted		
	imaging		

45.1 Introduction

The spleen is an organ that is commonly given less attention to than it deserves in oncologic imaging. One reason is that the spleen is rarely the site of primary malignancies. However, one should not underestimate the importance of the spleen, especially since certain splenic pathologies can have severe clinical presentations such as splenic rupture and even possibly hemoperitoneum [1].To better appreciate splenic pathology, we will first discuss the gross anatomy and microanatomy of the spleen. We will then discuss clinical and radiologic characteristics of benign and malignant splenic neoplasms, including mimickers of malignant lesions and common metastases, which are summarized in Tables 45.1 and 45.2.

Table 45.1 Imaging characteristics of benign splenic lesions

Table 45.1 Imaging characteristics of benign splenic lesions				
	US	CT	MR	
Cyst/pseudocyst	Posterior acoustic enhancement may contain echoes	Attenuation similar to water with a thin non-enhancing wall. Attenuation may be increased if infected or complex, and the wall may enhance if infected	Increased T2 signal intensity with variable T1 signal intensity and non-enhancing thin walls when not infected	
Gamna-Gandy bodies	Hyperechoic with or without shadowing		Hypointense T2 signal intensity, blooming on T1 in-phase images, and no enhancement	
Hemangioma	Solid echogenic or complex cystic appearance, and similar enhancement to background splenic parenchyma	Capillary – Iso- or hypoattenuating on non-contrast imaging, with homogeneous contrast enhancement. Cavernous – solid and cystic components with heterogeneous enhancement of the solid components following contrast administration	Hypo- to isointense to normal spleen parenchyma on T1-weighted images. Hyperintense to spleen on T2-weighted images. Variable enhancement	
Hamartoma	Echogenic, occasionally with cystic or calcified components. Early enhancement with rapid washout	Isoattenuating with background spleen and often only appreciated secondary to contour abnormalities. May enhance on postcontrast early arterial phase images	Isointense with background spleen on T1-weighted images, heterogeneous on T2-weighted images, and persistent enhancement on delayed imaging	
Lymphangioma	Similar appearance to cysts, except Doppler evaluation shows intrasplenic vascularity coursing about the periphery	Similar appearance to cysts	Multiloculated T2-hyperintense fluid with T2-hypointense septa. Variable T1 signal intensity without enhancement	
Littoral cell angioma	Mottled echotexture without discrete lesions or multiple solid lesions of variable echogenicity	Iso- or hypoattenuating lesions become isoattenuated or remain hypoattenuated following contrast administration	Usually hypointense on T1 and T2 images, but can have some increased T2 signal intensity depending on hemosiderin content	
Hemangiopericytoma	Hypoechoic with early arterial phase enhancement and decreased enhancement during portal and delayed phase imaging	Multilobular lesions with speckled calcifications as well as enhancing solid nodular components and/or septa	Hyperintense T2 signal intensity with corresponding hypointense T1 signal intensity	

Table 45.1 (continued)

	US	СТ	MR
Hemangioendothelioma	Hypoechoic masses, with anechoic areas and disorganized color flow in the setting of necrosis and tumoral angiogenesis	components (to a lesser degree	Heterogeneous T1 and T2 signal intensity with areas of decreased T1 and T2 signal intensity related to hemosiderin
Peliosis	Echogenic spleen with varying hypoechoic foci. Rapid central enhancement of the spleen with associated centripetal enhancement	Multiloculated blood-filled spaces with well-defined septa, occasionally with fluid-fluid levels. Significant enhancement of the lesions or enhancement of the dependent fluid	Increased T2 signal, occasionally hemorrhagic with increased T1 signal, occasionally hypervascular
Inflammatory pseudotumor	Hypoechoic with calcific components	Hypoattenuating mass with calcifications and heterogeneous enhancement	Hyperintense T2 signal intensity (occasionally hypointense), hypo- to isointense on T1-weighted imaging, and heterogeneous enhancement
Lipoma	Echogenic	Fat attenuation without enhancement	Increased T1 and T2 signal, which saturates on fat- suppressed images
Abscesses	Cystic lesions that progressively enlarge and may contain gas, possibly with subcapsular or extracapsular extension	Similar to ultrasound appearance	Hypointense T1 and hyperintense T2 signal intensity with minimal rim enhancement
Sarcoidosis	Variable hypoechoic to slightly hyperechoic inhomogeneous nodules without enhancement	Hypoattenuating nodules that do not enhance	Hypointense T1 and T2 signal without enhancement. Caseating granulomas are T2-hyperintense with peripheral hypointensity

Table 45.2 Imaging characteristics of malignant splenic lesions

	US	CT	MR
Angiosarcoma	Complex heterogeneous mass with foci of necrosis and/or hemorrhage. Hypervascularity on color Doppler interrogation in the solid portions	Similar to ultrasound appearance. May be hyperdense, or have punctate or large radial calcifications	Variable T1 and T2 signal related to necrosis, hemorrhage, and hemosiderin. Heterogeneous enhancement
Littoral cell angiosarcoma	Imaging characteristics of littoral cell angioma, with a more infiltrative or solid appearance	Imaging characteristic of littoral cell angioma, with a more infiltrative or solid appearance	Imaging characteristic of littoral cell angioma, with a more infiltrative or solid appearance
Pleomorphic undifferentiated sarcoma, fibrosarcoma, and leiomyosarcoma	Rare with no distinguishing characteristics. Can be cystic, solid, or complex masses	Rare with no distinguishing characteristics. Can be cystic, solid, or complex masses	Rare with no distinguishing characteristics. Can be cystic, solid, or complex masses

(continued)

	US	CT	MR
Kaposi sarcoma	Splenomegaly with multiple echogenic nodules. Heterogeneous echogenicity of the background parenchyma may be present	Spleen may be homogeneous or show hypoattenuating nodules that are iso- or hypoattenuating on delayed post-contrast images	Hyperintense on T2-weigted images
Lymphoma	Variable appearance from iso- to hypoechoic. May have increased through transmission. Hypoechoic with peripheral enhancement following contrast administration	Hypo- or isoattenuating without contrast enhancement	Iso- to hypointense on T1- and T2-weighted imaging. Lesions are better appreciated on post-contrast images as they are well circumscribed
Leukemia	No imaging abnormality. Occasional splenomegaly	No imaging abnormality. Occasional splenomegaly	No imaging abnormality. Occasional splenomegaly
Cystadenocarcinoma	Unilocular or multilocular large cysts	Unilocular or multilocular large cysts	Unilocular or multilocular large cysts
Metastases	Most commonly hypoechoic, but can be hyperechoic (e.g., colon). Inhomogeneous when necrosis is present	Decreased attenuation with peripheral enhancement. Calcification can be seen with primary cystadenocarcinomas	T2-hyperintense signal intensity and hypo- to isointense T1 signal intensity with variable enhancement

Table 45.2 (continued)

45.2 Anatomy

The spleen is a ductless glandular organ that is situated between the gastric fundus and the diaphragm (Fig. 45.1). The organ has diaphragmatic and visceral surfaces. The visceral surface can be further divided into gastric (anterior) and renal (posterior) portions. The gastric surface is in direct contact with the posterior wall of the stomach and tail of the pancreas. The medial aspect of the gastric surface is directed anteromedially and is termed the splenic hilum, where vessels and nerves enter and exit. The renal surface is anatomically associated with the superoanterior surface of the left kidney and sometimes the left adrenal gland. The colic surface sits upon the splenic flexure, phrenicocolic ligament, and usually the tail of the pancreas [2].

The spleen is held in position by two membranes – splenorenal ligament and gastrosplenic ligament [2], both of which are derived from the dorsal mesentery [3]. The splenorenal ligament and extends between the spleen and left kidney, through which the splenic vessels course. The gastrosplenic ligament extends between the spleen and stomach, through which the short gastric and left gastroepiploic branches of the splenic artery course [2].

The spleen functions as a blood-filtering organ that can be divided into three compartments – red pulp, marginal zone, and white pulp (Fig. 45.1). The red pulp is composed of splenic cords and rich plexus of venous sinuses. The splenic cords consist of reticular fibers, reticular cells, and macrophages. Various blood cell types are found in the spaces between the splenic cords. The splenic cords are also associated with lymphocytes and hematopoietic cells. The venous sinuses of the red pulp extend up to the marginal zone, where the systemic circulation is screened for antigens and pathogens. Some consider the marginal zone to be part of the white pulp, which is composed primarily of T and B lymphocytes [4, 5].

45.3 Benign Lesions of the Spleen

45.3.1 Splenic Cysts and Pseudocysts

Cysts of the spleen can be divided into true and false (or pseudo) cysts. True splenic cysts are rare entities [6], with primary cysts making up

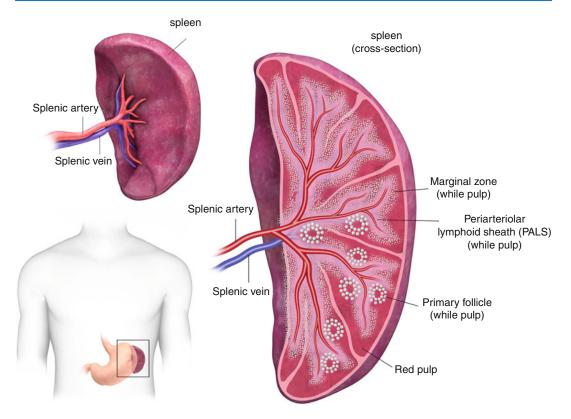


Fig. 45.1 Illustration demonstrates position of the spleen in the left upper quadrant of the abdomen, its gross anatomy including the splenic hilum, vasculature and anterior

only 20 % of splenic cysts [7]. Congenital or epithelial splenic cysts account for approximately 75 % of true splenic cysts. Generally speaking, congenital cysts are asymptomatic. However, cysts can enlarge and potentially hemorrhage in the setting of trauma. Additional potential complications include infection and rupture, possibly requiring partial or complete splenectomy [8].

True simple cysts are anechoic with posterior acoustic enhancement on ultrasound. Sometimes thin septa may be present. Internal echoes can be seen in the setting of hemorrhage. Calcifications, if present, are linear reflections with possible associated acoustic shadowing. Infected cysts may show acoustic enhancement of the thin wall and internal echoes (sediment). Ruptured cysts show discontinuity of the thin wall, with anechoic fluid extending beyond the normally spheroid shape of a cyst [8].

surfaces, and cross-sectional anatomy showing the components of the spleen including red pulp, white pulp, and marginal zone

On CT, simple cysts have internal attenuation similar to water with a thin, non-enhancing wall. However, the thin wall may enhance if infected. In addition, if the cyst is complex, septations and/ or calcifications may be seen. Hemorrhagic and infected cysts with sedimentation show higher attenuation than simple water [8].

On MRI, cysts have increased T2-weighted signal intensity with non-enhancing thin walls, and variable T1-weighted signal intensity, depending on the presence or absence of hemorrhagic products and/or sedimentation. Small foci of calcification are often difficult to identify due to lack of signal but can be seen as blooming on in-phase T1-weighted gradient-recalled images [8].

Pseudocysts make up about 75 % of nonparasitic splenic cysts. The main difference between these secondary cysts and primary cysts is that the wall is composed of fibrous tissue in secondary cysts (not an epithelial lining). Radiographically, these cannot be distinguished from primary cysts. True cysts generally do not need follow-up imaging, but it may be beneficial to follow-up pseudocysts if elicited by trauma to ensure their stability or involution [8].

45.3.2 Gamna-Gandy Bodies

Gamna-Gandy bodies represent hemorrhagic foci within the spleen, which result from portal hypertension [9, 10]. These lesions are composed of fibrous tissue associated with hemosiderin and calcium. Given these characteristics, on ultrasound, Gamna-Gandy bodies appear as hyperechoic foci in the background of normal splenic parenchyma, with or without posterior acoustic shadowing depending on quantity of calcifications. Sclerotic splenic veins associated with portal hypertension are seen as channels of spectral reflectors in the spleen. With MR imaging, the lesions are normally hypointense on T2-weighted imaging, bloom on in-phase T1-weighted gradient-recalled echo images, and do not enhance on post-contrast imaging (Fig. 45.2a–d). These findings in the setting of cirrhosis dismiss miliary tuberculosis, histoplasmosis, and disseminated *Pneumocystis carinii* infection [10].

45.3.3 Hemangiomas

Hemangiomas are the most common primary benign splenic neoplasms. Incidental hemangiomas generally measure <2 cm and commonly occur in the 30–50 years of age. Hemangiomas can be associated with angiomatosis syndromes such as Kasabach-Merritt syndrome, which consists of anemia, thrombocytopenia, and coagulopathy. A complication that can occur, particularly in large hemangiomas, is rupture. Additional complications include hypersplenism and malignant degeneration [1].

On radiographs, a hemangioma can manifest as a left upper quadrant mass or splenomegaly; however, these findings are not specific. Calcifications may or may not be present. On ultrasound, hemangiomas can be well defined or pedunculated and have a solid echogenic or complex cystic appearance. Echogenic foci with acoustic shadowing related to calcifications may be visualized if present [1]. With contrastenhanced sonography, hemangiomas usually enhance similar to and thus are isoechoic with background splenic parenchyma [11], though this can vary.

On non-contrast CT, capillary hemangiomas are hypoattenuating or isoattenuating lesions, which homogeneously enhance with intravenous contrast [1]. Enhancement of cavernous hemangiomas is associated with only the solid components and not the cystic components [1, 12]. On more delayed imaging, cavernous hemangiomas show discrete heterogeneous, mottled areas of enhancement [12]. Often the cystic components are associated with calcifications. With MR imaging, splenic hemangiomas are hypo- to isointense relative to normal spleen on T1-weighted imaging, whereas they are hyperintense on T2-weighted imaging (Fig. 45.3a, b) [1]. Note that hemangiomas of the liver can show T2-hyperintense signal on diffusion-weighted imaging due to T2 shinethrough effect, though there are cases that show restricted diffusion [13] which is also likely the case with splenic hemangiomas. There are three patterns of enhancement associated with hemangiomas: immediate/persistent homogeneous enhancement, early peripheral enhancement with homogeneous delayed enhancement, and peripheral, nodular enhancement with centripetal progression (Fig. 45.3c-f) [14]. If complications occur in large hemangiomas, such as hemorrhage or thrombosis, variable MR characteristics may be seen [1].

A case report has shown technetium (Tc-99m)labeled red blood cell scintigraphy with SPECT can be utilized for confirming presence of splenic hemangiomas [15]. By utilizing early and delayed planar and SPECT images, one can evaluate for focal blood pooling on delayed imaging, as has been shown with hepatic hemangiomas [16]. Alternatively, in Tc99m-labeled sulfur colloid studies, a photopenic defect can occur because the tracer accumulates only in the normal reticuloendothelial system tissue [17].

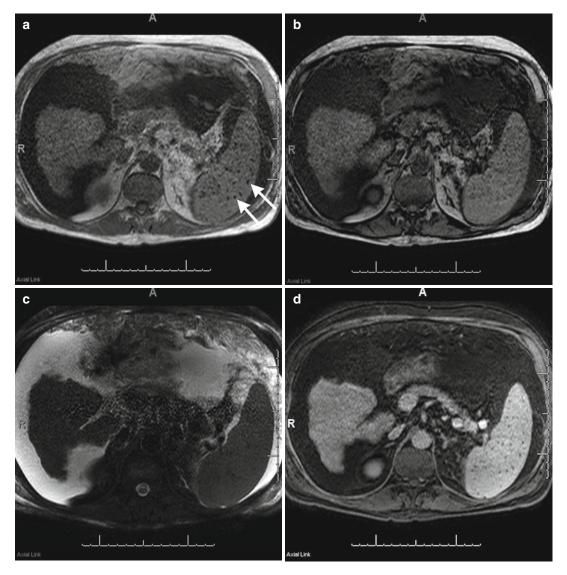


Fig. 45.2 A 59-year-old male with cirrhosis. There is blooming of iron-containing Gamna-Gandy bodies (*arrows*) on the in-phase T1-weighted images (**a**) when compared to out-of-phase T1-weighted images (**b**). In addi-

tion the Gamna-Gandy bodies have decreased signal intensity on fat-suppressed T2-weighted imaging (c), and no contrast enhancement on delayed 3D post-contrast fat-suppressed T1-weighted gradient-recalled echo imaging (d)

In general, no intervention or follow-up imaging is needed for hemangiomas, unless the patient is symptomatic. Often patients become symptomatic once hemangiomas have reached a certain size. If the patient has left upper quadrant pain or there is concern for rupture, partial or complete splenectomy may be considered for treatment and postoperative follow-up imaging may be obtained if there is concern for complications.

45.3.4 Hamartoma

Splenic hamartomas are rare benign lesions that can occur at any age, equally in men and women. Similar to hemangiomas, hamartomas are usually found incidentally but may be symptomatic if they are large, presenting as palpable masses, splenomegaly, or ruptured lesions. Hamartomas can be multiple and found

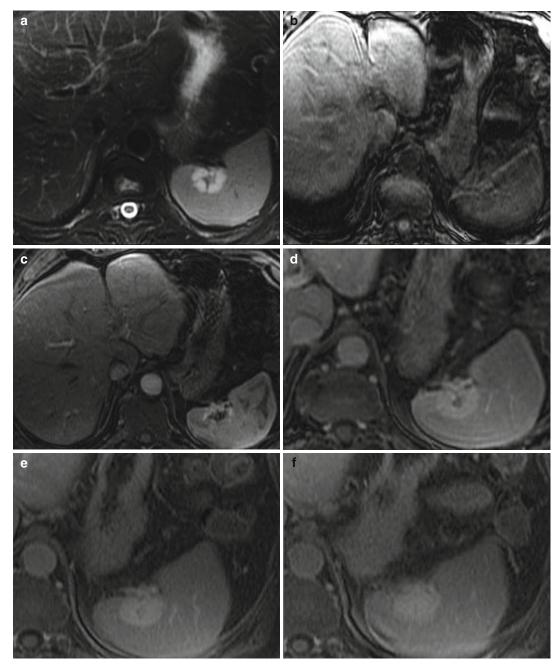


Fig. 45.3 Classic MR imaging characteristics of hemangiomas. Here we demonstrate that the splenic hemangioma is hyperintense on T2-weighted imaging (**a**) and hypointense on 3D precontrast fat-suppressed T1-weighted gradient-recalled echo imaging (**b**). During early post-con-

in extrasplenic areas and thus are associated with certain syndromes like tuberous sclerosis and Wiskott-Aldrich-like syndrome. Also similar to hemangiomas, thrombocytopenia and trast 3D post-contrast fat-suppressed T1-weighted gradientrecalled echo imaging (c), the hemangioma demonstrates discontinuous peripheral nodular enhancement, which fills in centripetally on delayed 3D post-contrast fat-suppressed T1-weighted gradient-recalled echo imaging (d-f)

anemia may be associated with splenic hamartomas [1].

Hamartomas tend to be solid masses. Thus, on ultrasound, hamartomas appear as echogenic

lesions, and sometimes may have cystic or calcific components. These lesions tend to have highly vascular components. In general, on postcontrast sonography, hamartomas demonstrate avid enhancement without early washout [18]. On CT, given the characteristics of the lesion, hamartomas tend to blend in with background splenic tissue, and often times the only defining characteristic is a subtle contour abnormality. In contrast, MR imaging has more defining characteristics. Though hamartomas are T1-isointense with the spleen, they show heterogeneous signal on T2-weighted imaging as well as post-contrast evaluation (Fig. 45.4). On delayed imaging, hamartomas show more uniform enhancement [1].

With Tc-99m radiolabeled isotopes, "hot spots" are also visualized for hamartomas [19]. It is important to note that given the vascularity of the lesions, they can mimic both benign lesions (such as hemangiomas) and metastatic lesions, and thus are difficult to diagnose with imaging alone.

45.3.5 Lymphangioma

Similar to other benign primary lesions of the spleen, patients with lymphangioma may be asymptomatic or have imaging that shows a large, multicentric mass that requires surgical intervention. Symptoms elicited from this lesion are the result of growth of the lesion that arose during childhood and now has mass effect upon surrounding organs. In addition, if the lesions are very large, bleeding, consumptive coagulopathy, hypersplenism, and portal hypertension may be elicited. When lymphangiomas occur in multiple organs, the process is called lymphangiomatosis [1].

With ultrasound and CT, these splenic lesions are most commonly incidentally detected. These lesions have the appearance of splenic cysts, with a wide range of sizes, and most commonly located in the subcapsular region. Commonly these lesions may have septa, internal debris, and calcifications. Doppler evaluation on ultrasound reveals intrasplenic vasculature coursing along the periphery of the cysts. With contrast-enhanced CT, no significant enhancement is detected [1]. Rarely, FDG uptake with PET/CT may be observed, possibly secondary to lymphatics present within the fibrous septa [20]. With MR imaging, lymphangiomas appear as T2-hyperintense multiloculated lymphatic fluid with intervening T2-hypointense septa. T1-weighted signal is variable in these lesions, depending on the absence or presence of hemorrhagic material and/or other debris (Fig. 45.5). On post-contrast imaging, if malignant degeneration has occurred, it is important to evaluate for soft tissue components [1].

Majority of lymphangiomas in the spleen are small and do not require surgery. However, larger ones, especially if symptomatic, may require partial or total splenectomy. Follow-up imaging can be considered if there is concern for postsurgical complications.

45.3.6 Littoral Cell Angioma

Littoral cell angioma is a rare vascular splenic lesion that can have benign and/or malignant components. Commonly, these are identified upon work-up of anemia and/or thrombocytopenia in symptomatic patients. Patients also can present with flu-like symptoms and pain. Splenomegaly is almost always associated with littoral cell angioma. The lesion has also been associated with other neoplastic processes including colorectal, renal, pancreatic adenocarcinoma, and meningioma [1].

Littoral cell angiomas usually present as multiple rather than solitary lesions. This pathology has widely variable appearance on imaging. On sonograms, the spleen may present with mottled echotexture without discrete lesions, whereas other cases present as individual lesions with variable echogenicity [1]. Usually with Tc-99m red blood cell scan, littoral cell angioma does not show radiotracer uptake [21]. On CT, these lesions appear as iso- or hypodensities [1, 22, 23] that become isoattenuated [1, 24] or remain hypoattenuated [22] with background spleen on post-contrast imaging. Like on sonography, coalescent lesions can be appreciated. MRI usually shows hemosiderinfilled T1- and T2-weighted hypointense signal given the cellular hematophagocytic capacity. However, some case studies have shown that if the lesions have less hemosiderin content, the lesions may have some T2-hyperintense signal [23, 24].

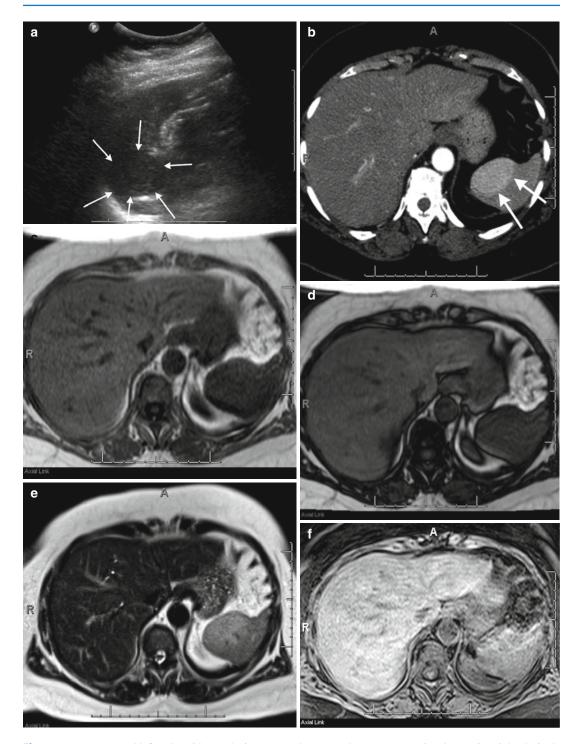


Fig. 45.4 A 71-year-old female with a splenic mass. Sagittal ultrasound of the spleen (**a**) demonstrates a solid mass arising from the upper pole of the spleen (*arrows*) with corresponding early arterial phase enhancement (*arrows*) on an axial-enhanced CT image (**b**). This mass shows isointense signal with splenic parenchyma on T1-weighted in-phase (**c**) and out-of-phase (**d**) imaging

but more heterogeneous signal on T2-weighted single shot fast spin echo (\mathbf{e}) imaging. The mass shows heterogeneous enhancement when comparing pre- (\mathbf{f}) and post (\mathbf{g})contrast-enhanced fat-suppressed T1-weighted gradient-recalled echo images. These findings are in keeping with a splenic hamartoma, which was pathologically confirmed



Fig. 45.4 (continued)

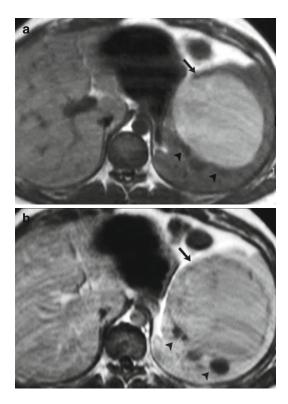


Fig. 45.5 A 27-year-old woman with splenic lymphangioma. A complex cystic mass was detected on a previous routine sonography study and confirmed later on enhanced CT. Axial unenhanced (a) and contrast-enhanced (b) gradient-echo T1-weighted images show a subcapsular multilocular mass with hypo- (*arrowheads*) and hyperintense (*arrow*) non-enhancing areas, revealing their cystic nature. Hyperintense areas were secondary to proteinaceous content. Diagnosis was confirmed after splenectomy. Artifact was the result of poor breath holding (With permission from Luna et al. [57])

Given that the symptoms are nonspecific, splenectomy is often performed for definitive diagnosis and treatment.

45.3.7 Hemangiopericytoma

Though hemangiopericytoma is considered a vascular benign lesion, it has high malignant potential. Only 25 % of these lesions arise within the abdomen, and when these do arise within the spleen, the lesions are usually asymptomatic or associated with splenomegaly [1].

These nodular lesions are hypoechoic on ultrasound. With contrast, lesions found in the liver show marked enhancement during arterial sonographic imaging and decreasing enhancement during portal and delayed imaging [25]. This may be similar in the spleen. On CT, the lesions appear as multilobular masses with smaller disseminated lesions throughout the spleen [1]. Speckled calcifications may be associated with the lesions and may show enhancing solid nodular components and/or septa on post-contrast CT images [12]. In some cases, the lesions may not be detected on CT or on Tc-99m sulfur colloid studies. On MR imaging, the lesions show T2-weighted hyperintense signal and T1-weighted hypointense signal [1].

Though these lesions are normally surgically excised, recurrence can occur in as many as 50 % of patients and are aggressive. Since recurrence has been reported even 20 years after initial therapy, close long-term surveillance is necessary [1].

45.3.8 Hemangioendothelioma

Patients with hemangioendotheliomas commonly present with left upper quadrant pain and palpable lesions. Patients also present with hypersplenism, hematologic abnormalities, and metastases. This condition occurs in children and young adult populations [1].

Hemangioendotheliomas are typically hypoechoic masses surrounded by normal splenic parenchyma. Anechoic areas may be present if tumor necrosis is present. On color Doppler images, disordered vascularization may be seen in the presence of tumoral neoangiogenesis. With CT, the lesions appear as hypoattenuated masses with enhancement of solid portions, which may appear hypovascular relative to background spleen. Necrotic and hemorrhagic areas do not show enhancement. These lesions may also have an infiltrative appearance. When the lesion occurs in the spleen, capsular retraction along surface lesions is not observed as is with liver lesions. On MR imaging, hemangioendotheliomas tend to be heterogeneous with hypointense T1- and T2-weighted signal due to presence of hemosiderin [1].

45.3.9 Peliosis

Peliosis is a disease consisting of blood-filled spaces within the spleen. It is rare for this process to occur as an isolated entity. The spectrum of causes of this disease is wide, but the most common association is anabolic steroids. The disease is also associated with aplastic anemia, tuberculosis, AIDS, and cancer. Like the majority of other benign splenic entities, this disease is commonly identified incidentally, unless a surface lesion ruptures, leading to intraperitoneal hemorrhage [1].

On ultrasound, the spleen is echogenic with varying hypoechoic foci related to the blood-filled spaces [1]. Contrast-enhanced ultrasound has been evaluated in hepatis peliosis, which results in rapid central enhancement with no centripetal enhancement [26]. With non-contrast CT, these blood-filled spaces may be multiloculated and contain well-defined septa. Some of these lesions may show fluid-fluid levels. With contrast, the lesions may significantly enhance and lose definition of septa and lobules, or show enhancement of the dependent fluid. If rupture is associated with these lesions, subcapsular splenic hematomas may be visualized [1].

The risks associated with biopsy of peliosis lesions are high, and thus this condition is commonly diagnosed upon splenectomy [1].

45.3.10 Inflammatory Pseudotumor

Inflammatory pseudotumor of the spleen is a very rare benign entity that is commonly misdiagnosed as other benign or malignant entities. The process usually occurs in middle-aged or older persons in both males and females. Though this entity can present as an incidental solitary lesion, patients can also present with left flank pain, fever, and/or splenomegaly. In others, anemia and leukocytosis may occur. In addition to the clinical manifestations of inflammatory pseudotumor, radiologic characteristics also mimic other processes. On ultrasound, this can present as hypoechoic masses with calcific components. On CT, this can appear as a hypodense mass with calcification on non-contrast studies but can then show heterogeneous enhancement with contrast administration. On MR imaging, characteristics correspond to fibrotic changes. Namely, the process is hyperintense on T2-weighted imaging (though in some cases this can show hypointense T2 signal), hypo- to isointense on T1-weighted imaging, and heterogeneous enhancement on delayed post-contrast imaging [27]. On PET/CT, these lesions show variable uptake, but sometimes intense FDG uptake can be visualized [28].

Given these characteristics, it is difficult to make the diagnosis on a presurgical basis. Further imaging is not necessary upon making this diagnosis.

45.3.11 Lipoma

Lipoma is another very rare nonvascular benign tumor of the spleen [8]. Lipomas are soft tissue masses composed entirely of fat. On ultrasound, lipomas are generally easy to detect as echogenic masses unless they are encapsulated. On CT, lipomas have the characteristic attenuation of fat and show no enhancement (Fig. 45.6). MR imaging normally shows high signal on T1-weighted imaging and T2 fast spin echo-weighted imaging and saturates on fat saturated sequences. If the lesions are simple lipomas, no enhancement is observed. Usually these masses do not require treatment unless the patient is symptomatic secondary to the lesion's mass effect, which can elicit pain.

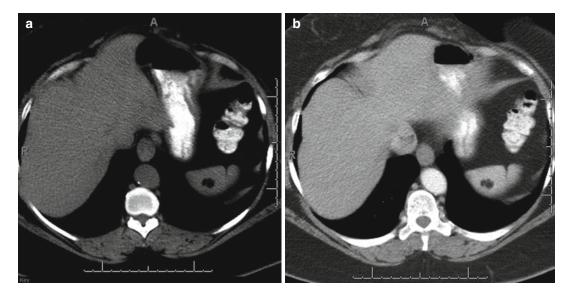


Fig. 45.6 Computed tomography shows a fat-attenuated lobulated lesion in the spleen on non-contrast axial CT imaging (**a**). Post-contrast axial imaging (**b**) illustrates characteristic lack of enhancement in this lipoma

45.3.12 Splenic Abscesses

Splenic abscesses are more commonly found in immunocompromised patients and can be solitary or multiple. For immunocompromised patients, the source of infection is often fungal (e.g., candidiasis). Clinical presentation often entails left upper quadrant pain, leukocytosis, and fever [5]. On sonography and CT, there is gas, progressive enlargement of the lesion, cystic components, and subcapsular/extracapsular extension [29]. Bacterial abscesses show no enhancement with ultrasound contrast material, but fungal/tuberculosis abscesses show enhancing rims and septae, which cannot be differentiated from tumors with central necrosis [30]. On MR imaging, abscesses commonly show T1-hypointense and T2-hyperintense signal with minimal peripheral enhancement of the capsules. Various nuclear medicine radioisotopes can be utilized to evaluate splenic abscesses. Gallium (Ga-67) citrate shows uptake in the rim of the abscess, whereas the central cavity is photon deficient. Indium (In-111) leukocyte and Tc-99m sulfur colloid scans show photon-deficient areas within the spleen [31].

45.3.13 Sarcoidosis

Splenic sarcoidosis does not usually clinically manifest itself. However, the disease is usually associated with systemic symptoms including fever, malaise, and weight loss. On physical examination, splenomegaly may be observed in 25–60 % of patients with splenic sarcoidosis. Concomitant lymphadenopathy in the abdomen is commonly observed [32].

Splenic sarcoidosis usually presents as diffuse, innumerable nodules measuring anywhere from 0.1 to 3.0 cm. Punctate calcifications are estimated in approximately 16 % of patients. Splenic sarcoidosis on ultrasound can vary from hypoechoic to slightly hyperechoic, inhomogeneous nodules resulting in a heterogeneous spleen (Fig. 45.7a) [32]. With contrast, splenic sarcoid foci do not enhance or are hypoenhancing relative to background spleen [30]. On contrastenhanced CT, splenic nodules are hypodense relative to the background spleen (Fig. 45.7b, c). On MRI, the lesions are hypointense on T1- and T2-weighted imaging and hypoenhance. If caseating granulomas are present, the lesions present as hyperintense lesions on T2-weighted imaging with peripheral hypointense signal. On delayed

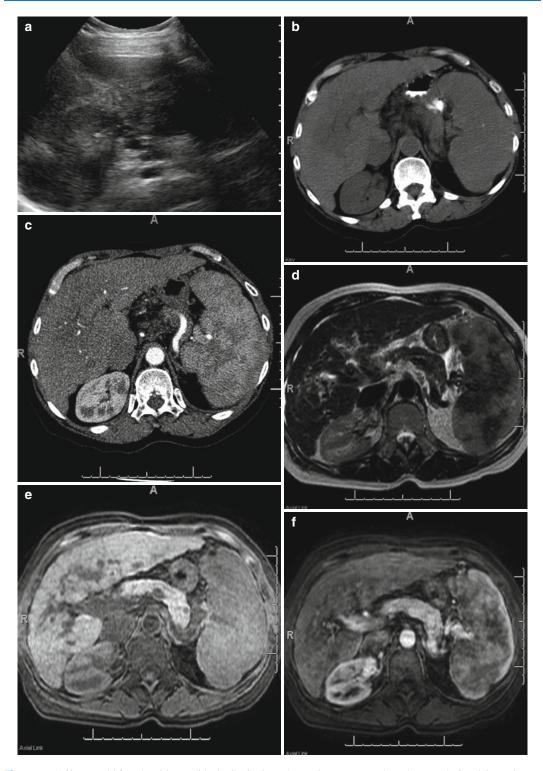
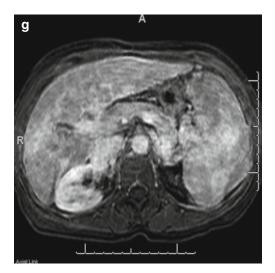
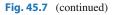


Fig. 45.7 A 68-year-old female with sarcoidosis. Sagittal ultrasound of the spleen (**a**) demonstrates an enlarged spleen with heterogeneous echogenicity. On non-enhanced (**b**) and enhanced (**c**) axial CT images, the echogenic areas on ultrasound correspond to areas of non-enhancement following contrast administration.

These also correspond to decreased signal intensity on T2-weighted single shot fast spin echo (d) and progressive contrast enhancement on 3D pre (e), post (f), and delayed (g) fat-suppressed T1-weighted gradient-recalled echo images, in keeping with splenic sarcoidosis





imaging, the lesions become less conspicuous (Figs. 45.7d–g and 45.8) [32].

45.4 Malignant Lesions of the Spleen

45.4.1 Angiosarcoma

Angiosarcoma is the most common nonhematolymphoid malignant tumor of the spleen. The tumor has no gender predilection and is more commonly found in elder patients. Some splenic angiosarcomas are associated with chemotherapy for lymphoma and radiation therapy for breast cancer. Symptoms include fever, fatigue, weight loss,

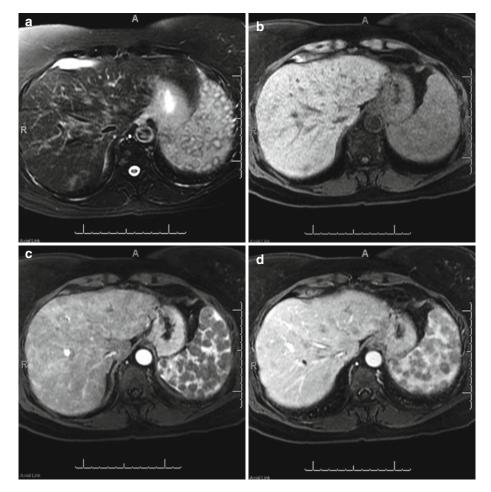


Fig. 45.8 A 68-year-old female with pathology-proven sarcoidosis. Axial fat-suppressed T2-weighted (**a**) image demonstrates multiple splenic target lesions with peripherally decreased T2-weighted signal intensity and central increased signal intensity corresponding to multiple case-

ating granulomas. These same lesions demonstrate progressive enhancement from pre (b) to early arterial (c), late arterial (d), and delayed (e) dynamic 3D fat-suppressed T1-weighted gradient-recalled echo images

heterogeneous enhancement may be observed (Fig. 45.9) [1].

Treatment of choice for splenic angiosarcoma is splenectomy. However, since metastases are often present at the time of diagnosis, angiosarcomas may be treated with chemotherapy and/or irradiation, sometimes in combination with surgical treatment. Follow-up imaging every 3 months during treatment is generally recommended given the aggressive nature of this tumor. However, majority of patients die within 1 year of diagnosis [1].

45.4.2 Littoral Cell Angiosarcoma

This rare malignant tumor has morphologic characteristics of littoral cell angioma but has an infiltrative or solid growth pattern reflective of angiosarcoma, making them difficult to distinguish from more classic angiosarcoma on imaging.

45.4.3 Pleomorphic Undifferentiated Sarcoma, Fibrosarcoma, and Leiomyosarcoma

Pleomorphic undifferentiated sarcoma, formerly called malignant fibrous histiocytoma [34], is the

Fig. 45.9 Delayed post-contrast 3D fat-suppressed T1-weighted gradient-recalled echo images demonstrates a large heterogeneously enhancing mass (*arrowheads*). The central hypointense area within the mass is reflective of necrosis with more peripheral solid enhancing tumor components. This mass is a pathology-proven angiosar-

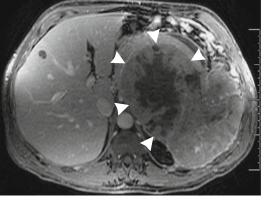
coma of the spleen

Fig. 45.8 (continued)

and abdominal pain as well as disorders such as anemia, thrombocytopenia, and other coagulative disorders. The abdominal pain may be associated with splenomegaly and left upper quadrant abdominal mass, which can rupture and cause hemoperitoneum. Metastasis to the liver, lungs, bone, bone marrow, and lymphatic system can occur [1].

At the time of imaging, angiosarcomas usually present as aggressive, irregular tumors with metastases. On ultrasound, the mass itself appears complex and heterogeneous, with possible foci of necrosis and/or hemorrhage. Hypervascularity may be noted with Doppler imaging in the more solid portions. Note that metastases at the time of presentation are also hypervascular. Similar characteristics are seen on CT, with more apparent evidence of hemoperitoneum if spontaneous rupture occurs. This can be seen as hyperdense products on non-contrast CT in the acute setting. The tumor can show punctate calcifications or large radial calcifications [1]. On PET/CT, angiosarcomas are FDG avid [33].

On MR imaging, the signal on both T1- and T2-weighted imaging can be mixed due to presence of necrosis and/or hemorrhage. Hypointense signal may also be due to presence of siderotic nodules (hemosiderin). In addition, due to presence of necrosis with solid tumor components,



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most common soft tissue malignant neoplasm in adulthood but is rarely found in the spleen. Less than 15 cases have been reported in the English medical literature as of 2012 [35]. Pleomorphic undifferentiated sarcoma is a polymorphic sarcoma that is very aggressive and results in splenomegaly [36]. Unfortunately, with imaging, there are no distinguishing characteristics to make this diagnosis prior to surgery. Surgical resection is the treatment of choice even in the setting of recurrence [35]. Two primary splenic malignant tumors that mimic pleomorphic undifferentiated sarcoma include fibrosarcoma and leiomyosarcoma. These primary malignant tumors all can present as cystic, solid, or complex masses [37].

45.4.4 Kaposi Sarcoma

Kaposi sarcoma is a spindle cell neoplasm that has a well-known association with human immunodeficiency virus (HIV) infection and acquired immunodeficiency syndrome (AIDS) [34]. AIDS-related Kaposi's sarcoma has a relatively common association with the liver, but not the spleen. Involvement of the spleen is usually undetected clinically [38].

With ultrasound imaging, splenomegaly is often detected with multiple echogenic nodules (5–12 mm), which are located along blood vessels and tend to coalesce [38]. Heterogeneous echogenicity may also be noted for the background parenchyma [39]. Interestingly, with CT imaging, the spleen may remain homogeneous [38]. In other cases, CT may show hypoattenuating nodules that are iso- or hypoattenuating on delayed post-contrast imaging [39]. Thus, Kaposi sarcoma enhancement can mimic that of hemangiomas.

In Tc-99m sulfur colloid liver-spleen imaging, reduced radiotracer uptake is reflective of Kaposi sarcoma [40].

45.4.5 Lymphoma

Lymphomatous involvement of the spleen is the most common splenic malignancy, presenting as

either a primary disease or more commonly as a metastatic systemic process. Less than 1 % [41] of lymphomas present as a primary splenic disease, with or without infiltration of lymph nodes within the splenic hilum [42, 43]. Primary splenic lymphoma is more prevalent in non-Hodgkin lymphomas [43] and in patients with AIDS-related lymphomas [37]. Secondary involvement of the spleen due to disseminated lymphoma occurs in up to one-third of Hodgkin lymphomas and slightly more often for non-Hodgkin lymphomas [37]. The most common presenting symptom of splenic involvement is left upper quadrant pain due to capsular distention, although constitutional symptoms such as fever or weight loss are also typical.

Grossly, lymphomatous involvement of the spleen can take many different forms and is dependent upon the cell type. Primary splenic lymphoma often presents as a bulky mass that may infiltrate through the splenic capsule and into adjacent structures [37, 41]. Presentations of secondary involvement include homogeneous splenomegaly without or with a focal mass or multiple masses, miliary pattern, or without any apparent change [37, 41, 43–45]. The pattern of involvement depends upon the cell type; large-cell lymphoma typically presents as a large, solitary mass [43, 46], while other non-Hodgkin lymphomas present diffusely as either multiple masses or miliary nodules [46].

With ultrasound, discrete lesions due to splenic lymphoma usually appear hypoechoic [45, 47, 48], but can also be isoechoic and therefore difficult to detect via this modality [11]. Occasionally, the lesions display increased With through transmission [41]. contrast administration, lymphomatous lesions are hypoechoic with irregular peripheral enhancement [45] and become more hypoechoic relative to normal splenic parenchyma as a function of time [11, 49]. The sensitivity and specificity of ultrasound for detecting and diagnosing splenic lymphoma are 54 and 100 %, respectively [48]. Ultrasound is limited in that it can detect larger lesions with a fairly high sensitivity but does not fare as well for diffuse splenic involvement.

Similarly, on CT imaging, focal lymphomatous lesions appear homogeneously hypoattenuated

relative to the surrounding parenchyma [43–45]. Infiltrative disease by primary splenic lymphoma is often diffusely hypo- or isoattenuated relative to the spleen [37]. Focal lesions typically do not enhance after contrast administration (Fig. 45.10a) [37, 43, 44]. Occasionally, wedge-shaped infarcts due to lymphoma are visualized.

On MRI, lymphomatous lesions are typically difficult to detect before contrast administration. Splenic lymphoma and normal parenchyma both have similar T1 and T2 relaxation times [37, 50],

and lesions therefore tend to appear isointense to hypointense on non-contrast imaging (Fig. 45.10b, c). With contrast, MRI becomes much more sensitive for detection of focal lymphomatous deposits, with lesions appearing well circumscribed and markedly hypointense relative to the surrounding spleen (Fig. 45.10d–g) [5, 37, 41, 43, 50]. Although these lesions are typically homogeneously hypointense, they can occasionally have small irregularities suggestive of necrosis, fibrosis, edema, or hemorrhage [43].

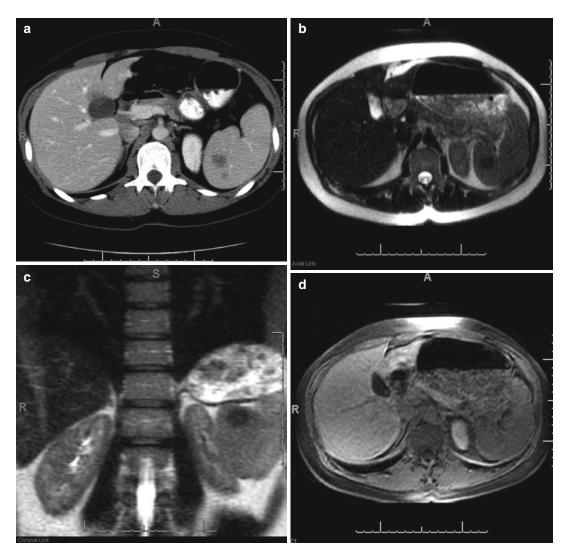


Fig. 45.10 A 29-year-old female with incidentally noted splenic lesions. Axial-enhanced CT (\mathbf{a}) image demonstrates non-enhancing splenic lesions with decreased signal intensity on axial (\mathbf{b}) and coronal (\mathbf{c}) T2-weighted signal shot fast spin

echo images with mild progressive enhancement from pre (**d**) to early arterial (**e**), late arterial (**f**), and delayed (**g**) 3D fatsuppressed T1-weighted gradient-recalled echo images. These lesions were pathologically proven to be lymphoma

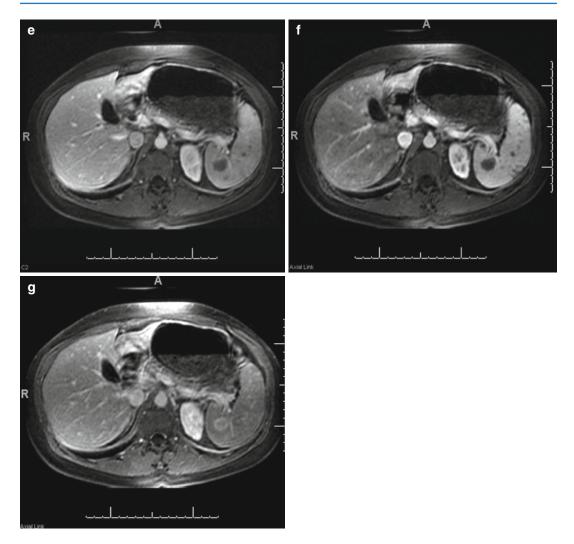


Fig. 45.10 (continued)

PET imaging, particularly with FDG, is especially useful in detection and staging of splenic lymphoma. Lesions tend to display avid FDG uptake. In addition, FDG uptake is avid in involved lymph nodes [42, 51, 52]. Tc-99m sulfur colloid scintigraphy studies will show photopenic defects, either focally or diffusely throughout the spleen depending on the degree of involvement. Photopenic defects can also represent a variety of other pathology, such as infarction, metastatic disease, and other masses such as hemangioma, abscess, or cyst [46].

45.4.6 Leukemia

Splenic involvement of leukemias most commonly present without any imaging abnormality [53]. When a splenic abnormality can be visualized, it is generally as homogeneous splenomegaly [5, 39, 41, 54] due to leukemic infiltration [55], especially for acute myelogenous leukemia subtypes [41]. When severe, this enlargement can result in splenic rupture [56].

Chloromas, most commonly associated with acute myelogenous leukemia, are rare. On MRI, they appear as multiple ill-defined



Fig. 45.11 A 5-year-old male with acute myeloid leukemia. MRI was performed for staging after positive bone marrow biopsy. Unenhanced (**a**) and immediate postcontrast (**b**) 3D fat-suppressed gradient-echo T1-weighted images show multiple hypovascular nodules (*arrows*) not seen on unenhanced MRI. The nodules

masses without enhancement on immediate post-contrast dynamic images [57] (Fig. 45.11).

Leukemia also has a propensity to damage and disrupt the splenic parenchyma and as such splenic infarction [37] is another common imaging abnormality. This parenchymal disruption also makes the spleen more susceptible to bacterial infection, and therefore splenic abscesses are also visualized more often in patients with leukemia.

45.4.7 Cystadenocarcinoma

Splenic cystadenocarcinoma is a very rare primary splenic malignancy, with only less than ten cases reported in the medical literature as of 2010. The most common complaint associated with this condition is upper abdominal pain and possible palpation of a left upper quadrant mass. Carcinoembryonic antigen and carbohydrate antigen 19-9 may be elevated [58]. As seen with cystadenocarcinomas of the pancreas, splenic cystadenocarcinomas present as cystic lesions that contain large cysts that are unilocular or multilocular cross-sectional on imaging. Diagnosis is confirmed upon definitive treatment with surgical resection of the lesion or possible splenectomy.

became isointense to spleen on delayed acquisition (not shown). The nodules represent chloromas, which tend to be more conspicuous, as in cases of lymphoma, on immediate post-contrast acquisition. In this case, the lesions disappeared after chemotherapy (With permission from Luna et al. [57])

45.4.8 Metastasis

Splenic metastasis occurs in approximately 7 % of oncologic patients, with hematogenous spread most commonly from the breast, lung, ovary, stomach, cutaneous melanoma (Fig. 45.12), and prostate gland [37].

On ultrasound, most metastases appear as hypoechoic lesions. Colonic carcinoma metastases appear as hyperechoic foci. If central necrosis has occurred in lesions, metastases may appear inhomogeneous [59]. On CT, metastases appear as decreased attenuation, whether they are cystic, solid, or infiltrative. Peripheral enhancement and enhancement of septa within discrete lesions may be observed. Calcifications may be present if the primary tumor is cystadenocarcinoma [37]. With MR imaging, splenic metastases have T2-weighted hyperintense signal and T1-weighted hypo- to isointense signal with variable enhancement characteristics depending on the type of metastasis [5]. In the setting of a known FDG avid neoplasm, FDG PET can accurately differentiate between benign and malignant lesions (Fig. 45.13). In addition, if an FDG avid lesion is identified on PET imaging with no known malignancy, further evaluation is necessary [51].

With ovarian carcinoma, gastrointestinal adenocarcinoma, and pancreatic cancer, peritoneal carcinomatosis can occur, causing splenic surface scalloping with associated cystic or solid implants on cross-sectional imaging. Direct invasion of the spleen by primary malignancies or metastases is rare, especially since the spleen is usually displaced by such pathology. However, possible primary malignancies that can invade the spleen include gastric, colonic, pancreatic, and left renal carcinomas as well as neuroblastomas, mesotheliomas, or retroperitoneal sarcomas (Fig. 45.14) [37].

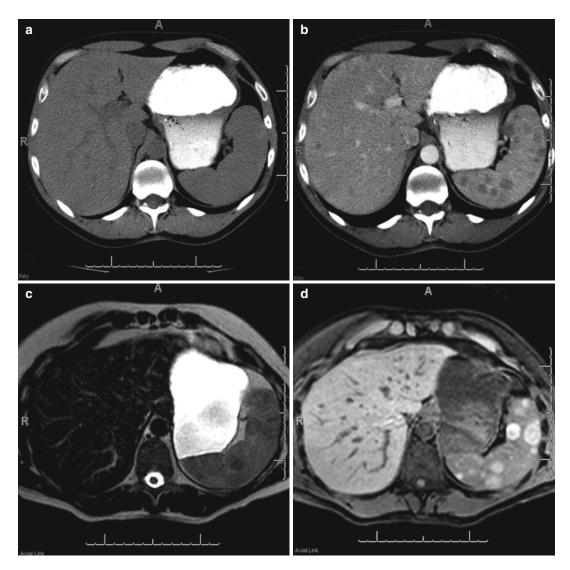


Fig. 45.12 A 42-year-old male with cutaneous melanoma. Axial unenhanced (**a**) and enhanced (**b**) CT images show mild enhancing splenic metastases. These metastases demonstrate decreased signal intensity on axial T2-weighted single shot fast spin echo (**c**) and increased signal intensity on axial fat-suppressed T1-weighted gradient-recalled echo (**d**) imaging related to melanin content. Following administration of

intravenous gadolinium, the lesions demonstrate mild internal enhancement, best appreciated on axial postcontrast subtraction imaging (e). Additionally, these metastases demonstrate increased signal intensity on diffusion-weighted imaging (f) related to reduced Brownian motion secondary to increased cellularity. Also noted on the diffusion-weighted image (f) are multiple subcentimeter hepatic metastases

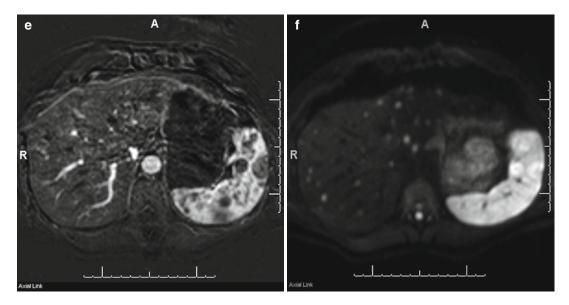


Fig. 45.12 (continued)

45.5 Role of Functional Imaging for Focal Splenic Lesions

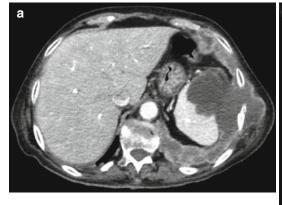
Though functional imaging of the spleen is a relatively young field, the potential impact is quite vast, particularly in differentiating benign and malignant lesions. We discussed earlier the characteristics of some splenic pathology with contrast-enhanced ultrasound, including lymphomatous lesions that initially show irregular peripheral enhancement and then increasingly are hypoechoic to background splenic parenchyma as a function of time [11, 49], whereas benign lesions including hemangiomas and hamartomas remain iso- or hyperechoic to background splenic parenchyma [11, 18].

Contrast enhancement on CT and, in particular, MR imaging also provide a distinctive role in characterizing splenic lesions. In addition to multiphasic post-contrast CT imaging characteristics of benign lesions, there are preliminary data that show CT perfusion of the spleen in the setting of lymphomatous involvement is lower than in control or cirrhotic patients [60]. FDG PET/CT activity provides a method for detection of splenic malignancy particularly in the setting of known malignancy (Fig. 45.15), but one needs to be aware of false positives that can occur such as with inflammatory pseudotumor. Post-contrast MR imaging allows for characterization of focal splenic benign lesions including cysts, hemangiomas, and hamartomas, while allowing for improved detection of primary splenic malignancies and metastatic disease [57]. Moreover, rapid dynamic contrast-enhanced MR imaging may further aid detection of malignant splenic involvement as has been shown in Hodgkin disease [61]. The role of MR diffusion-weighted imaging is still in question when it comes to splenic pathology due to the variable appearance of benign and malignant splenic pathology with this sequence (Fig. 45.16) and because physiologic diffusionweighted imaging of the spleen results in diffusion restriction.



Fig. 45.13 A 67-year-old female patient with history of prior surgery for stage T3N0M0 rectal carcinoma. Three years after intervention, the patient presented with elevated levels of carcinoembryonic antigen (CEA); PET-CT and WB-DWI MRI were performed to exclude metastatic disease. Coronal FDG PET shows no signs of local tumor recurrence although a focal area of increased metabolic activity in the presacral region is visualized (**a**, *arrowhead*). Note the presence of a small hypermetabolic focus (SUVmax 7.6) in the medial border of the spleen (**a**, *arrow*). Coronal WB MRI-STIR sequence (**b**, *arrow*)

reveals a nonspecific hypointense small lesion within the spleen, consistent with metastasis. Corresponding coronal MPR WB-DWI with inverted contrast grayscale (\mathbf{c} , *arrow*) demonstrates a small splenic lesion appearing as a white spot within the low signal spleen. Notice the absence of areas of restricted diffusion within the pelvis, excluding local recurrence. The native axial DWI-MR image (\mathbf{d} , *arrow*) with a *b* value of 700 s/mm² in the upper abdomen confirms the low signal lesion seen on DWI of the splenic lesion that corresponded to a splenic metastasis (Reprint with permission of Springer from Vilanova et al. [62])



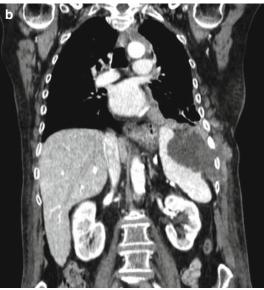


Fig. 45.14 A 70-year-old female with a malignant pleural mesothelioma receiving chemotherapy presented to the emergency room with severe left upper quadrant pain. During the physical examination, a painful lump was noted in her left abdominal lateral wall, at the level of the eighth and ninth ribs. Abdominal contrast-enhanced axial and coronal CT images (**a** and **b**, respectively) demonstrate diffuse nodular left pleural thickening

with heterogeneous enhancement in relation with the malignant pleural mesothelioma. The malignancy invades ribs, muscles, and the subcutaneous tissue of the chest wall and extends through the diaphragm to the abdominal cavity. The spleen is infiltrated by a well-defined, low-attenuated mass with peripheral enhancement, consistent with extensive central necrosis (With permission from Martin Noguerol et al. [63])

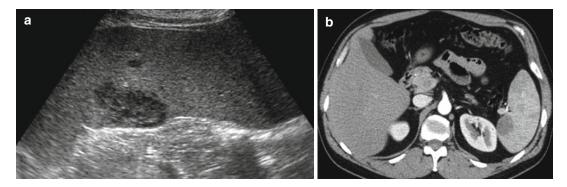


Fig. 45.15 A 44-year-old male underwent an ultrasound for elevation of hepatic enzymes, which demonstrated a normal-appearing liver and increasing splenomegaly of up to 17 cm over two consecutive years. Subsequently, a focal hypoechoic mass within the spleen was incidentally depicted (**a**). On multiphasic contrast-enhanced CT, the mass was homogeneously hypodense during arterial and portal phases (**b** and **c**, respectively). MRI was also performed for further characterization. The mass was hyperintense on T2-weighted sequence (**d**) and hypovascular during immediate (**e**) and 1-min post-contrast (**f**) phases of the dynamic series, with demonstration of delayed enhancement (**g**). Due to its nonspecific appearance, FDG PET-CT was also carried out, demonstrating hypermetabolism of the mass (**h**), with absence of other FDG avid lesions (**i**). Splenectomy was performed, which showed presence of a primary splenic high grade non-Hodgkin lymphoma

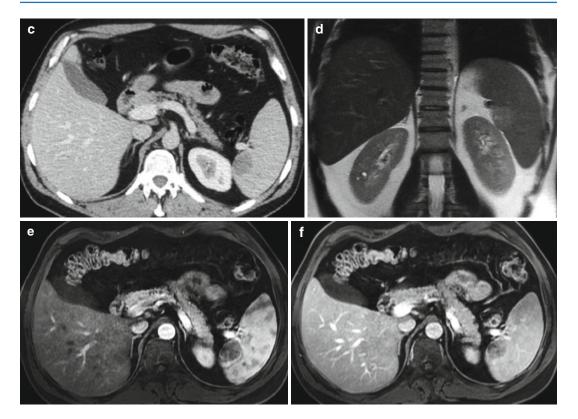
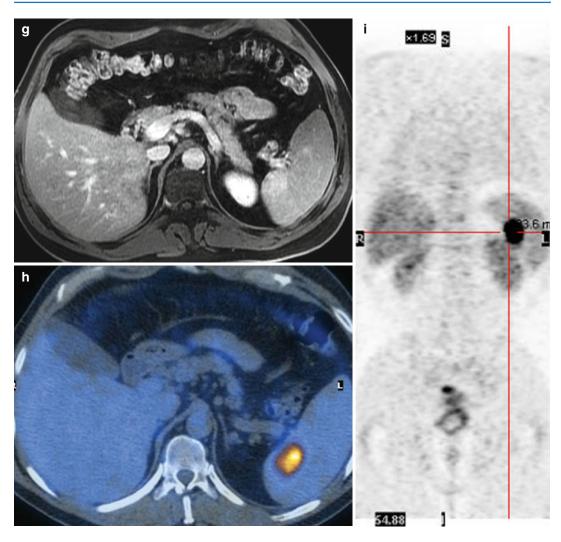


Fig. 45.15 (continued)





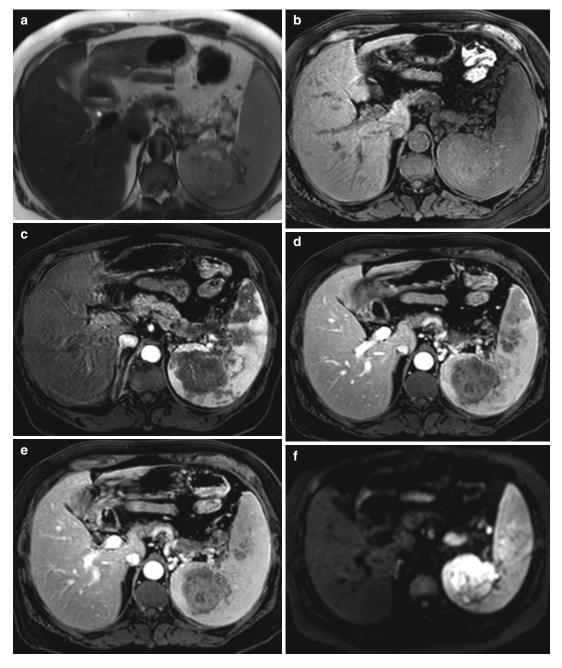


Fig. 45.16 Focal splenic lesion in patient with gastric carcinoma. Axial T2-weighted image demonstrates heterogeneous appearance of a splenic mass (\mathbf{a}) , which is also slightly hyperintense on precontrast fat-suppressed T1-weighted gradient-recalled echo image (\mathbf{b}) . On the dynamic series, the lesion is hypovascular on immediate

post-contrast acquisition (c) with progressive and heterogeneous enhancement on 1-min and delayed post-contrast acquisitions (d and e, respectively). On high *b* value diffusion-weighted image (f), the lesion is hyperintense with low signal on corresponding ADC map (g). All these features are indicative of splenic metastasis

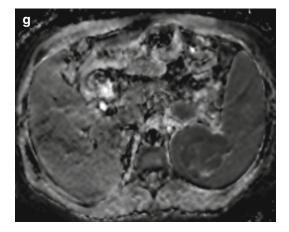


Fig. 45.16 (continued)

Conclusions

The spleen is not a topic of common discussion in radiology due to the rarity of primary malignancies arising from this visceral organ. However, it remains an important area of discussion due to the potential severe manifestations of pathologies that can occur. Functional imaging provides radiologists the necessary tools for accurate diagnosis of splenic pathology. Specifically, contrast-enhanced imaging plays a critical role. The use of CT perfusion and rapid dynamic contrast-enhanced MRI are areas that have been barely explored in understanding splenic pathology and will likely be of importance in the future. In addition, PET/ CT continues to play an important role in identifying FDG avid lesions in the setting of primary malignancies and metastases.

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