Revisiting the Spleen—An Imaging Review of the Common and Uncommon Splenic Pathology

Meshaal Nadeem[1] Hina Arif Tiwari[2] Kedar Jambhekar[3] Hemendra Shah[3] Roopa Ram[3]

[1] Department of Diagnostic Radiology, University of Arkansas for Medical Sciences, Little Rock, Arkansas, United States
[2] Division of Radiology, Department of Medical Imaging, University of Arizona Health sciences, Tuscon, Arizona, United States
[3] Department of Radiology, University of Arkansas for Medical Sciences, Little Rock, Arkansas, United States

Address for correspondence Meshaal Nadeem, DO, Department of Diagnostic Radiology, University of Arkansas for Medical Sciences, 4301, W. Markham Street, Little Rock, AR 72205, United States (e-mail: Mnadeem@uams.edu).

Abstract

Keywords
► sclerosing angiomatous nodular transformation
► spleen
► spontaneous rupture
► hydatid
► sarcoidosis
► lymphoma

The spleen is the largest lymphatic organ and is responsible for both hematological and immunological functions. Several common etiologies such as trauma, developmental variants, infectious/inflammatory conditions, and benign and malignant lesions can occur in the spleen. The role of imaging modalities such as ultrasound (US), computed tomography (CT), and magnetic resonance imaging (MRI) in diagnosing these conditions continues to evolve. The main objective of this review article is to illustrate the role of imaging in identifying the common and uncommon pathology of the spleen.

Introduction

The spleen is the largest lymphatic organ in the body and serves several important functions such as providing immunologic surveillance, red cell turnover, and hemodynamic support. A wide range of pathologic conditions affect the spleen and can be categorized into traumatic, infectious and inflammatory, vascular conditions, and benign and malignant tumors. Additionally, there are normal variants that also affect the spleen. These conditions can be imaged on multiple modalities such as ultrasound (US), computed tomography (CT), magnetic resonance imaging (MRI), and nuclear medicine (NM).

Anatomy

The normal spleen is intraperitoneal and appears semilunar in shape with an approximate length of 11 cm. The weight of the spleen averages around 150 g. Spleen length and volume, however, vary based on height and gender of a patient and can be calculated both on CT and US.[1] As measured on CT, a strong correlation has been shown between spleen volume and three dimensional coefficient calculated as maximum length × hilum thickness × vertical height.[2] As measured on US, the Eq. 0.524 × width × thickness × (maximum length + craniocaudal length)/2 has been shown to correlate closely with helical CT measurements.[3]

On macroscopic examination, the spleen has a diaphragmatic surface and a visceral surface with a central hilum from which the splenic artery and splenic vein emerge. On microscopic examination, the spleen consists of red pulp, which is composed of tortuous blood vessels and sinusoids, and interspersed cords of white pulp, which is composed of lymphatic tissue. On US, the spleen has a uniformly homogeneous echotexture with a mildly higher echogenicity compared with the normal liver (►**Fig. 1**).

On unenhanced CT, the spleen is homogeneous with Hounsfield units (HU) ranging from 40 to 60 HU. On arterial phase imaging, the spleen demonstrates diffuse heterogeneity with serpiginous cord like differentially enhanced areas, owing to variable rates of blood flow through the sinusoids in the splenic pulp. On portal venous phase, homogeneous enhancement is seen throughout the parenchyma (►**Fig. 2**).

On MRI, the spleen is hypointense to liver on T1-weighted images (T1WI) and hyperintense to the liver on T2-weighted images (T2WI). As with CT, heterogeneous arterial phase enhancement is seen and normalizes on the portal venous phase (►**Fig. 3**).

Variants of Size, Shape, and Location

Splenomegaly–As mentioned previously, several factors such as age, gender, and height influence the size of the spleen. The general criteria for splenomegaly include craniocaudal length of more than 13 cm, extension beyond the lower pole of the left kidney, and medial extension up to the aorta. Splenomegaly can be associated with a variety of pathophysiologic conditions such as passive congestion (liver cirrhosis, portal vein thrombosis and congestive heart failure), infiltrative disorders (glycogen storage diseases, myelodysplasias, sarcoidosis, hematologic malignancies and neoplasms), and immunologic conditions (infections, rheumatoid arthritis, extramedullary hematopoiesis) which are summarized in ►**Table 1**[4] (►**Fig. 4**).
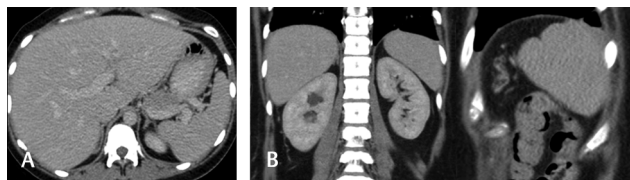

Fig. 1 Axial (**A**) and coronal (**B**) contrast-enhanced CT images of a patient with a normal spleen.
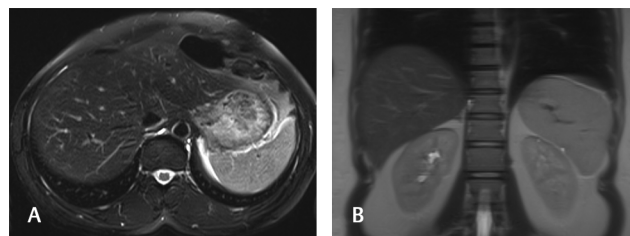

Fig. 2 Axial fat-suppressed T2-weighted image (**A**) and coronal T2-weighted Half Fourier-acquired single-shot turbo spin echo (HASTE) image (**B**) MRI of a patient with a normal spleen.
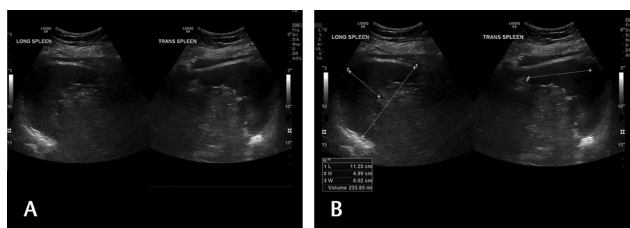

Fig. 3 Axial (**A**) and sagittal (**B**) ultrasound (US) images of a patient with a normal spleen.

Accessory spleen–Failure of fusion of splenic buds during embryogenesis can result in formation of sequestered splenic tissue, which is otherwise known as a splenule or supernumerary spleen. Their incidence is around 16%, and their most common location is at the splenic hilum or in the tail of the pancreas as well as around the gastrosplenic and pancreaticosplenic ligaments. They are incidentally seen nodules of variable size ranging from 1 cm to 4 cm and show a hilum. Accessory spleen can be misinterpreted as a pathologic peritoneal nodule or enlarged lymph node, many times leading to surgical excision.[5] Positive uptake on nuclear scintigraphy using Technetium-99 sulfur colloid or heat denatured tagged red blood cells help delineate the definitive splenic origin. In patients undergoing splenectomy for trauma, the accessory spleen may serve as the only residual salvageable splenic tissue[6] (►**Fig. 5**).

Polysplenia and asplenia–Numerous small foci of splenic tissue (polysplenia) or absent or very small splenic tissue (asplenia) are seen in heterotaxy syndromes, which refer to abnormal positioning of viscera in the chest and abdomen. While polysplenia is associated with bilateral left-sidedness (right-sided stomach, midline liver, intestinal malrotation, truncated pancreas, interrupted hepatic inferior vena cava with azygous continuation, bilateral bilobed lungs, complex cardiac anomalies), asplenia is associated with bilateral right-sidedness (bilateral trilobed lungs, intestinal malrotation, severe and often fatal complex cardiac anomalies)[7] (►**Fig. 6**).

Splenosis–Following traumatic or iatrogenic injury to the spleen, there may be heterotopic autologous transplantation of splenic tissue to unusual locations in the body, which is known as splenosis. Common sites include the left pleura, undersurface of diaphragm, greater omentum, peritoneum, and serosal surface of small bowel. Rare deposition along subcapsular liver and kidney and subcutaneous soft tissues

Table 1 Causes of splenomegaly

| |
|---|
| Hematologic–hemoglobinopathies, hemolytic anemias, thalassemias |
| Rheumatologic–rheumatoid arthritis, systemic lupus erythematosus, sarcoidosis |
| Infections–viral, mycobacterial, fungal, parasitic |
| Congestive–cirrhosis, venous thrombosis, congestive heart failure |
| Infiltrative–lymphoma, leukemia, metastasis, myeloproliferative disorders, glycogen storage disorders |
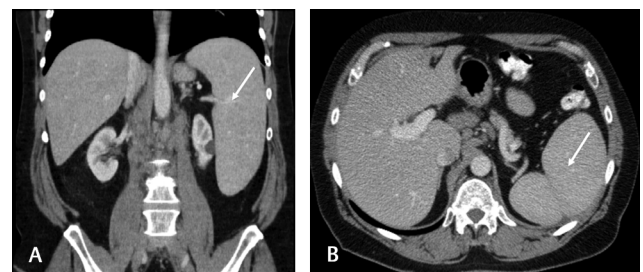
Fig. 4 Showing splenomegaly in a patient with nonHodgkin lymphoma (arrows in **A** and **B**).

of the abdominal wall at scar sites have also been reported. Splenosis deposits derive blood supply by parasitization from adjacent visceral feeding arteries.[8]

The significance of recognizing splenosis is its potential to be mistaken for a neoplasm or a pathological enlarged lymph node, particularly when the clinical history is unknown and the abdomen has not been imaged to detect an absent normal spleen. Splenosis has been mistakenly characterized as lung, adrenal, hepatocellular and gallbladder tumors.[9-12] Splenosis can also present with acute abdominal pain due to torsion or small/large bowel obstruction.[13,14] However, as splenosis shows presence of active splenic tissue, nuclear scintigraphy using heat denatured tagged red blood cell scan is sensitive in detecting ectopic splenic locations (►**Figs. 7, 8**).

Wandering spleen–Due to developmental laxity of the splenic ligament, the spleen can move about its pedicle, resulting in migration of the spleen from its normal position. This is often seen in children and young adults, more commonly in women. A wandering spleen can undergo torsion, infarction or rupture and can be treated by splenopexy or splenectomy.[15]

Trauma

As the spleen is a frequently involved organ in blunt abdominal injury, imaging trauma patients, particularly hemodynamically unstable patients with contrast-enhanced multidetector CT (MDCT), play an important role in triage. CT imaging should include arterial, portal venous, and delayed phases to detect parenchymal abnormalities such as contusion/laceration/rupture and vascular abnormalities such as pseudoaneurysm and extravasation. On CT, a splenic contusion appears as a poorly defined, nonlinear area of hypoattenuation, while a splenic laceration appears as a linear, irregular branching hypodensity.[16] Features of splenic rupture on CT include perisplenic hematoma and
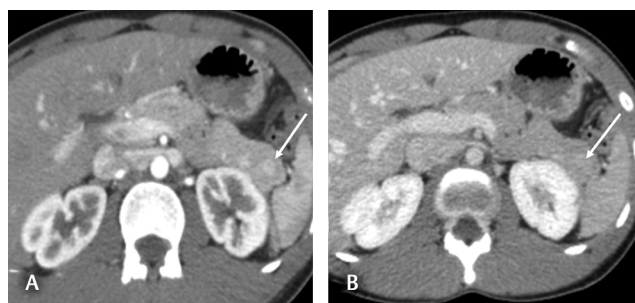

Fig. 5 Small enhancing nodule seen in the pancreatic tail on arterial phase (**A**) and portal venous phase (**B**) CT with enhancement pattern similar to the spleen. This is consistent with an accessory spleen in the pancreatic tail.
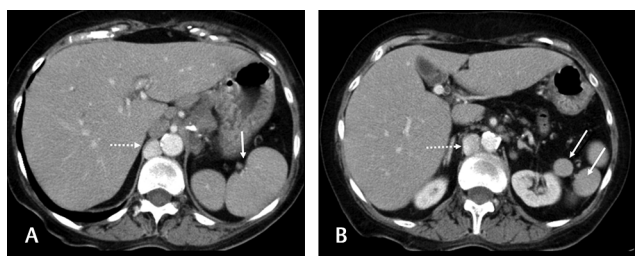

Fig. 6 Numerous variable size soft-tissue nodules (solid arrows in **A** and **B**) consistent with polysplenia in patient with heterotaxy. Note the absent intrahepatic inferior vena cava (IVC) and azygous continuation (dashed arrows in **A** and **B**).
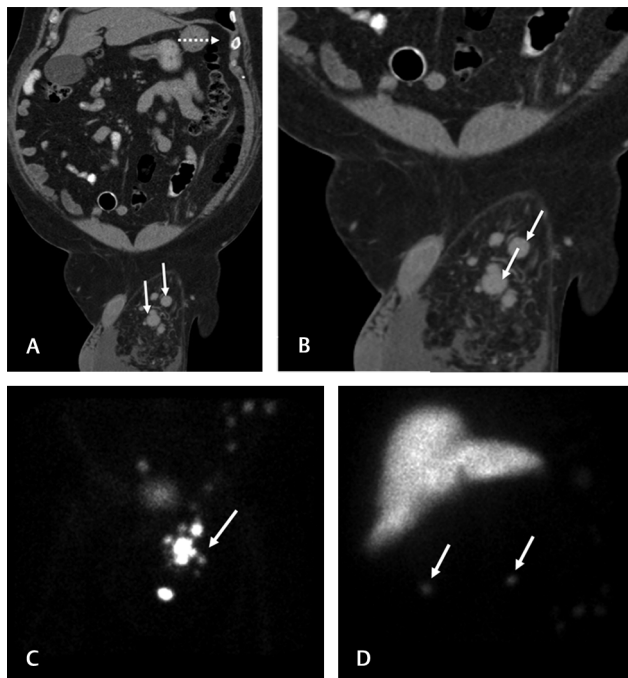
Fig. 8 Coronal CT images showing conglomerate of ovoid nodules in the left scrotum (arrows in **A** and **B**) in patient with history of prior trauma. Note the absence of spleen (dashed arrow in **a**). Technetium 99 tagged red blood cell scan shows that these nodules take up radiotracer are consistent with splenic tissue (arrows in **C** and **D**). Note the additional splenosis in the mid and left abdomen (arrows in **D**).

Fig. 7 Multiple ovoid nodules in the anterior midabdomen and right lower quadrant (arrows in **A**–**C**), which are seen on axial postcontrast CT in patient with history of prior trauma, with the largest measuring 5 cm and representing splenosis.

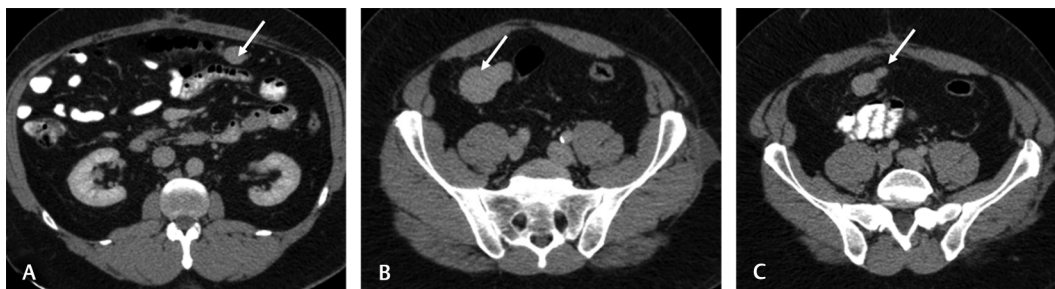hemoperitoneum. In hemodynamically stable patients with low energy trauma, there is an emerging role for contrast-enhanced ultrasound (CEUS) to screen and triage patients and particularly to minimize radiation in pediatric patients and women of reproductive age[17] (►Figs. 9–11).

The commonly used grading system for classifying splenic injuries is the American Association of Trauma Surgery (AAST) grading system,[18] which is summarized in ►Table 2. ►Table 2 shows the AAST grading system.

Management of patients with blunt splenic injury depends on hemodynamic status, presence of other organ injuries, as well as presence of significant CT findings. Nonoperative management is considered for hemodynamically

Fig. 9 Axial postcontrast CT images showing a 5.2 cm splenic laceration with extension into the hilum (solid arrows in **A**) and perisplenic hemorrhage (dashed arrow in **B**), which are consistent with grade 4 injury. No pseudoaneurysm or active extravasation was seen.

Fig. 10 Contrast-enhanced axial CT image showing active extravasation with a 1.4 cm pseudoaneurysm involving a lower pole branch of the splenic artery (arrow). Due to presence of vascular injury, this is consistent with grade 4 injury.

stable patients with less severe (lower than grade 3) injuries. Angioembolization is another treatment option for splenic salvage which can be considered in hemodynamically stable patients with higher grades of splenic injury. Splenectomy is an option reserved for patients with concomitant serious abdominal injuries, hemodynamic instability, or coagulopathy.[19]

Vascular

Splenic artery aneurysm and pseudoaneurysm–Splenic artery aneurysms and pseudoaneurysms are important vascular anomalies of the spleen, which can present with life-threatening hemorrhage and hence should be recognized on cross-sectional imaging for prompt treatment.

Splenic artery aneurysms are the most common nontraumatic cause of abdominal visceral aneurysms and occur in 0.1% of the population. The incidence is much more common in females with a 4:1 ratio, and most of them are discovered incidentally in asymptomatic patients imaged for other reasons. Most aneurysms are solitary, but up to 20% of them have been reported to be multiple.[20] Loss of normal elasticity and medial degeneration of the wall as a result of hypertension, hormonal factors, smoking, collagen vascular disease, pancreatitis and chronic liver disease have been proposed as etiology for development of splenic artery aneurysms. The
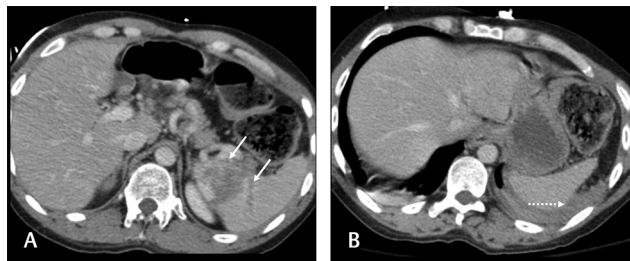
Table 2 Spleen injury scale

Grade	Imaging Criteria/CT Findings
I	Subcapsular hematoma < 10% surface area
	Parenchymal laceration < 1 cm depth
	Capsular tear
II	Subcapsular hematoma 10–50% surface area; Intraparenchymal hematoma < 5 cm
	Parenchymal laceration 1–3 cm depth
III	Subcapsular hematoma > 50% surface area; ruptured subcapsular or intraparenchymal hematoma ≥5 cm Parenchymal laceration > 3 cm depth
IV	Any injury in the presence of splenic vascular injury or active bleeding confined in splenic capsule Parenchymal laceration involving hilar or segmental vessels producing > 25% devascularization
V	Any injury in the presence of splenic vascular injury with active bleeding extending beyond the spleen into the peritoneum Shattered spleen

Fig. 11 Axial postcontrast arterial and portal venous phase images showing active contrast extravasation, which increases from early to later phases (solid arrows in **A** and **B**) with increase in size of perisplenic hematoma (dashed arrows in **A** and **B**) Coronal image shows a shattered spleen with irregular hypodensities and perisplenic hematoma which in association with vascular injury (arrows) is consistent with grade 5 injury.

most common site of involvement is the distal third of the artery, while proximal and mid segments involvement can be seen in the setting of pancreatitis. The aneurysm may have fusiform or saccular morphology, show partial thrombosis and peripheral calcification, and can spontaneously rupture in 2 to 3% with a mortality of 25%. Size > 2 cm is considered to be the size at which there is significant risk of rupture (►**Fig. 12**).

Due to their increase in size in pregnancy, splenic aneurysms are at high risk for rupture. Risk of rupture increases significantly to 20 to 50%, particularly with aneurysms larger than 2.5 cm during pregnancy, and can be associated with high maternal mortality of 75%. Due to this risk, aneurysms larger than 2 cm during pregnancy are treated with endovascular or surgical treatment.[21]

Pseudoaneurysms of the splenic artery are most commonly reported in the setting of pancreatitis, trauma, peptic ulcer disease, or postsurgery. They can be symptomatic and present with upper or lower gastrointestinal tract (GIT) hemorrhage and have a high mortality rate, with a reported 37% risk of rupture without treatment. On contrast-enhanced CT, focal outpouching of contrast is seen from the splenic artery or one of its branches surrounded by intrasplenic hematoma if there has been a recent bleed. Regardless of clinical presentation, splenic pseudoaneurysms are always treated due to high risk of rupture. Endovascular treatment using both proximal and distal coils (sandwich technique) or placement of stent graft across the pseudoaneurysm are commonly performed[22] (►**Fig. 13**).

Splenic vein thrombosis–Portal hypertension in patients with liver disease and perivenular inflammation as a sequelae of prior pancreatitis are the most common causes of splenic vein thrombosis. Other causes include involvement by pancreatic cancer and postsurgical and prothrombotic states. To shunt blood from the occluded splenic vein, short gastric collaterals open up, which maybe the cause of upper GI bleeding when they rupture.[23] On imaging, the splenic vein maybe expanded in the acute stage with filling defects within it, with the thrombus showing no vascularity on color Doppler or enhancement with postcontrast imaging.

Splenic infarction–Thromboembolism is the most common cause of splenic infarction. Both arterial or venous occlusion can result in splenic infarction.[24] Hematological disorders, autoimmune disorders, trauma, and surgery are the other causes.

The stage and size of the infarct determines the imaging appearance. Infarcts are seen as wedge-shaped areas with apex toward the hilum and broader base toward the capsule. During evolution, they can also have a round/mottled/irregular appearance and show no postcontrast enhancement. They may develop a fibrous central scar which appears echogenic on US.[25] The complications of an infarcted spleen are superimposed infection, hemorrhage, and splenic rupture (►**Figs. 14, 15**).

Spontaneous rupture of the spleen–Nontraumatic rupture of the spleen is a rare life-threatening abdominal emergency, with an incidence of 0.1 to 0.5%.[26] Common causes include infections such as malaria,[27] infective endocarditis,[28] primary systemic amyloidosis,[29] autosomal bleeding disorders,[30,31] and hematological malignancies. Unenhanced CT shows enlargement and irregular hyperdense areas within the spleen and precontrast T1WI and fat suppression best shows the hemorrhagic products on MRI.

Infection and Inflammation

Splenic abscess–Since the spleen itself serves a defense role, the incidence of splenic abscess is low and ranges from 0.14 to 0.7%
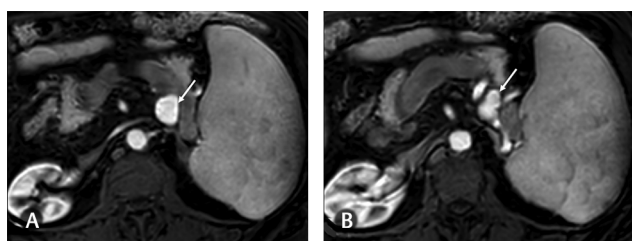

Fig. 12 Axial T1-weighted fat-suppressed postcontrast arterial phase images showing a rounded vascular lesion contiguous with the splenic artery, which is consistent with a splenic artery aneurysm (arrows in **A** and **B**).
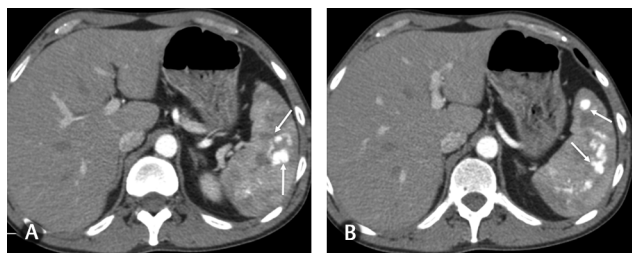

Fig. 13 Axial arterial phase CT images showing multiple lobulated foci of arterial enhancement with contrast density similar to the adjacent aorta (arrows in **A** and **B**) in patient with known staphylococcal bacteremia. These are consistent with mycotic aneurysms.
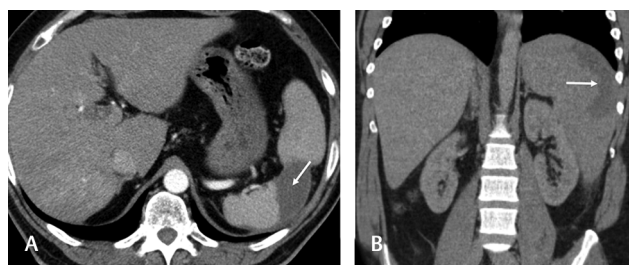

Fig. 14 Axial and coronal CT images showing large, peripheral wedge-shaped nonenhancing hypodensity representing splenic infarct seen involving the midspleen (arrows in **A** and **B**).
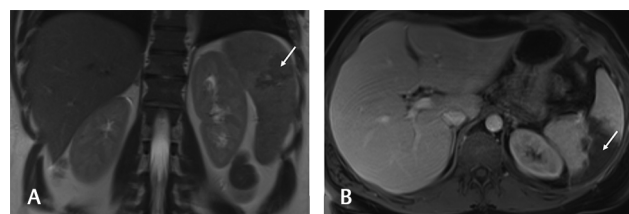

Fig. 15 Coronal T2-weighted image showing large peripheral wedge-shaped T2 hypointense area (arrow in **A**) and no associated post contrast enhancement (arrow in **B**).

in autopsy series.[32] Immunosuppressed patients and intravenous (IV) drug abusers are the most common population affected.[33] Contiguous spread from adjacent infected organs, direct inoculation from trauma, and underlying infarction are other causes. Fungal microabscesses in immunosuppressed patients can be caused by organisms such as Aspergillus, Cryptococcus, and Histoplasma. Tuberculosis is an additional cause of microabscesses. The clinical manifestations include fever, left-sided upper quadrant abdominal pain, and leukocytosis.

US shows an ill-defined, hypoechoic region in the splenic parenchyma with internal septations and debris.[34] Foci of air can also be seen at a later stage.[34] Associated findings include a left-sided pleural effusion. CT shows multifocal small hypodense lesions with peripheral rim of enhancement. MRI shows T1 hypointense and T2 hyperintense peripherally enhancing lesions.[1] Diffusion restriction is also seen due to presence of complex fluid. Ultrasound and CT also play a vital role in the image-guided percutaneous aspiration of these abscesses[35] (►Fig. 16).

Splenic hydatid cyst–Hydatid disease of the spleen is caused by Echinococcus species, with an incidence reported at around 2.5%.[36] Isolated splenic hydatidosis is less common.[37] The larval stage of Echinococcus granulosus is the causative agent, with human beings serving as accidental hosts. History of being from or travel to an endemic area should be sought in patients who may present with abdominal pain and splenomegaly. The World Health Organization (WHO) working group has proposed a classification for the sonographic appearance of hydatid cysts[38] (►Table 3).

A smooth-walled, anechoic cystic lesion is often seen in the early stage. Internal echoes from shifting of the brood capsules maybe seen and is referred to as the snowflake sign or hydatid sand. Gravity-dependent layering of the hydatid sand in the dependent portion of the cyst can be seen with positional imaging.[39] Multiple daughter cysts can be demonstrable within the mother cyst in the next stage. On further progression of the disease, free-floating membranes, which represent detached laminated membranes, can be seen as the water lily sign. Wall calcifications develop following death of the larva.

On CT, a unilocular or multilocular cystic lesion, with attenuation of cyst content being higher than simple fluid, was seen. The outer wall of the cyst known as pericyst may show calcifications, which is best seen on unenhanced CT as a hyperdense rim and is generated as a host response. When the membranes detach, on unenhanced CT, a wrinkled area of increased attenuation may be seen parallel to the pericyst as well. Daughter cysts may be present in clusters along the periphery of the mother cyst.

On MRI, the cyst contents show T1 hypointense and T2 hyperintense signal with the pericyst wall showing T2 hypointense signal due to the presence of dense collagen.[40] Incipient detachment of membranes is seen as curvilinear low signal within the cyst wall. Complications of hydatidosis include intraperitoneal rupture and anaphylactic shock, spread to other viscera, and fistulous communication with other structures[41] (►Fig. 17).

Sarcoidosis–Noncaseating granulomas associated with multisystem sarcoidosis can involve the spleen in up to 40% cases. While the liver is the most common abdominal organ to be involved along with the spleen, isolated splenic involvement has also been reported.[42] CT shows small size low-density lesions with no significant enhancement. On MRI, randomly distributed small T1 and T2 hypointense nodules with minimal delayed postcontrast enhancement may be seen.[43] The imaging differential diagnosis for splenic sarcoidosis includes lymphoma, fungal opportunistic infection, and amyloidosis (►Figs. 18, 19).

Table 3 WHO sonographic classification of hydatid cysts

| Cyst type	US characteristics	Cyst viability
Type 1	Unilocular, anechoic	Active
Type 2	Multiloculated, multiseptated anechoic with daughter cysts	Active
Type 3	Fewer daughter cysts, low level internal echoes, water lily sign	May be viable or inactive
Type 4	Heterogeneous, no daughter cysts	Inactive
Type 5	Thick calcified wall	Inactive

Abbreviation: US, ultrasound.
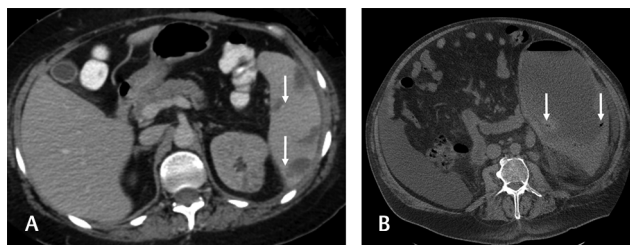
Fig. 16 Axial postcontrast CT images showing multiple hypodense lesions consistent with splenic abscesses (arrows in **A**) in a patient with history of infective endocarditis and cerebral abscesses. Some lesions show peripheral rim of postcontrast enhancement (bottom arrow in **A**). Large fluid collection with air fluid level within the spleen consistent with abscess (arrows in **B**) in another patient with splenomegaly from cirrhosis.
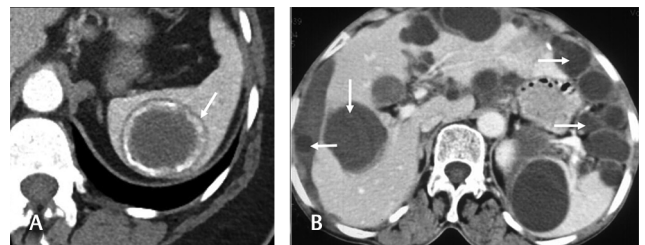

Fig. 17 Axial postcontrast CT images showing a well-circumscribed splenic cyst with thick peripheral rim of calcifications (arrow in **A**) in patient with isolated splenic hydatid disease. Another patient with disseminated hydatid disease showing cysts in liver, spleen, and peritoneum (arrows in **B**). Courtesy: Dr. Krishna Prasad Shanbhogue, M.D.

Benign and Malignant Masses of the Spleen

Splenic cysts–Cysts of the spleen are the most common benign splenic masses and are usually asymptomatic. Splenic cysts can be a) true cysts or b) pseudocysts; typically, indistinguishable from each other. True cysts are less common and account for only 20%. A true cyst wall is lined by epithelial cells. They can be congenital or form as a result of parasitic infection. Pseudocysts are more common than true cysts (80%), and they may form following splenic hematoma, infection or infarction; therefore, they may show mural calcifications (30–40% of cases) along their wall which is devoid of epithelial cells[44-47]

On CT, a unilocular, hypodense, nonenhancing lesion is seen which may show wall enhancement as a sequelae of prior trauma or infection (►Fig. 20).

Typical findings of a simple cyst are best seen on MRI, with homogenously hyperintense signal on T2WI, hypointense signal on T1WI and no internal enhancement on postgadolinium images. Hemorrhagic or proteinaceous contents within the cyst may show hyperintense signal on precontrast T1WI. Cyst wall enhancement can sometimes be seen similar to CT, based on the etiology of the cyst.[48]

Hemangioma–Splenic hemangiomas are the most common benign solid tumors of the spleen and are incidentally detected on imaging studies, more often in male patients. Frequency of hemangiomas is 0.03 to 14% on autopsy series. These benign vascular neoplasms are made of proliferation of vessels and range from capillaries to cavernous forms.
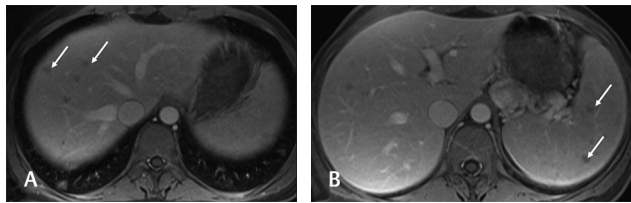

Fig. 18 Axial fat-suppressed postcontrast T1 weighted images in portal venous phase show multiple hypointense liver and spleen lesions (arrows in **A** and **B**) with mild splenomegaly in a patient with sarcoidosis.

Solitary hemangiomas are more common as compared with diffuse hemangiomatosis, which may be associated with syndromes (e.g. : Klippel–Tre´naunay–Weber, Turner, Kasabach–Merritt-like, and Beckwith-Wiedemann syndromes).[49-51]

Imaging appearance of splenic hemangioma is variable, based on its type: capillary or cavernous. Capillary hemangiomas are focal, usually round, and echogenic lesions on US. On contrast-enhanced CT, continuous peripheral arterial enhancement is seen with progressive centripetal fill-in on delayed phases. Unlike hepatic hemangiomas, interrupted peripheral nodular enhancement is not commonly seen with splenic hemangiomas. On MR imaging, hemangiomas appear hyperintense to splenic parenchyma on T2WI, and show progressive persistent uptake on dynamic postgadolinium images. Cavernous hemangiomas have nonvascular cystic components and therefore appear heterogeneous on T2WI, and also show heterogeneous enhancement on postcontrast CT or MR imaging[52-55] (►Fig. 21).

Hamartoma–Splenic hamartoma is an incidentally found, rare, congenital, benign splenic lesion which consists of nonneoplastic malformed red and white splenic pulp. As a hamartoma is made of spleen-like tissue, it may appear the same as splenic parenchyma on various imaging modalities, sometimes only seen as a contour bulge. Hamartomas appear mildly hypodense to splenic parenchyma on noncontrast CT, and have enhancement similar to adjacent splenic parenchyma on delayed postcontrast CT. They may be isointense to slightly hyperintense on T2WI and show homogenous delayed enhancement on postgadolinium MR imaging.[56,57]

Lymphangioma–Lymphangioma is a rare lymphovascular malformation with small cystic spaces filled with lymph, which is seen commonly in children. Similar to hemangioma, there are subtypes of lymphangiomas such as capillary and cavernous types. On US, lymphangiomas are typically subcapsular in location and classically appear as a well-defined, hypoechoic, loculated mass, which is avascular on color Doppler.

Hypodense cysts are seen on CT with peripheral wall enhancement or calcifications. On MR, lymphangiomas are

Fig. 19 Ultrasound (US) image of same patient showed ill-defined hypoechoic liver lesion which was biopsy proven sarcoid granuloma (calipers).

Fig. 20 Axial postcontrast CT image showing well-circumscribed rounded hypodense lesion in the posterior upper spleen consistent with simple cyst (arrow).
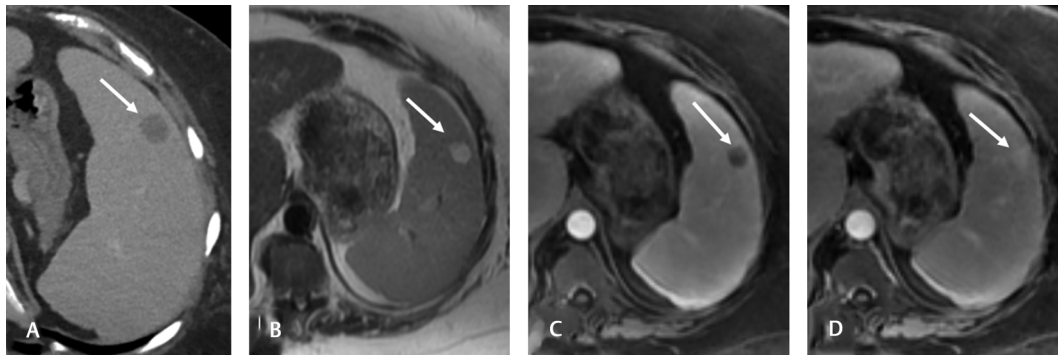
Fig. 21 Axial postcontrast image (arrow in **A**) showing hypodense indeterminate spleen lesion. Axial T2-weighted MRI (arrow in **B**) shows T2 hyperintense lesion corresponding to CT lesion. Axial arterial and delayed phase postcontrast T1 weighted images showing filling in of the lesion on the delayed image (arrows in **C** and **D**), consistent with hemangioma.

hyperintense on T2WI due to the cystic components, which may be multilocular with thin septations. If cysts contain blood or proteinaceous contents, hyperintense signal on precontrast T1WI is seen.[58-60]

Littoral cell angioma–Littoral cell angioma is an uncommon benign tumor of the spleen and is a relatively newly identified entity. This benign vascular neoplasm develops from the lining cells of the red-pulp sinuses, the so-called "littoral cells," and may contain hemorrhagic components.[61] Littoral cell angioma is usually multinodular and may involve the entire spleen, resulting in splenomegaly. On contrast-enhanced CT scan, lesions are hypodense to spleen on the portal venous phase and may become isodense on the delayed phase of postcontrast imaging, due to delayed filling in.[62] Littoral cell angiomas show hemangioma-like signal characteristics on T2-weighted images with inhomogeneous hyperintense appearance. Similar to CT findings, lesions may appear hypointense on unenhanced T1- weighted images, and show delayed contrast enhancement on dynamic post contrast imaging, following gadolinium administration with contrast pooling in the vascular channels. Hypointense signal on all MR sequences may be encountered as well due to due to hemosiderin accumulation within neoplastic littoral cells.[62]

Sclerosing angiomatoid nodular transformation (SANT)–SANT is a relatively newly described benign vascular condition of the spleen[63] and occurs due to an abnormal reaction of the red pulp to any vascular injury/inflammation. On US, these lesions can be heterogeneously hypoechoic with central linear echogenic foci which can have acoustic shadowing.[64] On contrast-enhanced US, a radiating spoke wheel pattern of enhancement has been described.[65] CT scan shows an isodense or hypodense mass, which can have central calcification, and can also show a radiating spoke wheel pattern of postcontrast enhancement, which histologically corresponds to stellate fibrous septations separating the vascular angiomatoid nodules. On MRI, central T1-hyperintense signal consistent with hemorrhage has been reported as well as a T2-hypointense stellate scar.[66] Progressive radiating centripetal enhancement is also described on late phase postcontrast images.[66] While MRI can help in narrowing the

differential diagnosis, tissue sampling is needed to confirm the diagnosis.

Lymphoma–Spleen can be commonly involved by Hodgkin's and nonHodgkin's lymphoma. Splenic involvement by lymphoma can be classified as primary and secondary. Primary splenic lymphoma is a rare entity. It comprises less than 1% of all lymphomas, which is usually nonHodgkin's lymphoma, diffuse large B cell type. There may or may not be associated splenic hilar lymphadenopathy.

Secondary splenic lymphoma refers to involvement of the spleen and lymph nodes, other than those in the splenic hilum.[67-69]

The following four classic patterns of imaging are observed with lymphomatous involvement of the spleen:

1. Splenomegaly secondary to diffuse infiltration without focal mass.
2. Small, miliary lesions.
3. Multiple focal nodules/masses.
4. Solitary mass.

Diffuse infiltration of the splenic parenchyma by lymphoma may be seen as normally enhancing spleen. Very small lymphomatous lesions may be difficult to appreciate. Splenic nodules may appear hypodense on contrast-enhanced CT. On MR imaging, nodules signal may be isointense on T1- and T2-weighted sequences, with delayed homogenous uptake of gadolinium on postcontrast sequences. Calcification due to posttreatment effects can be appreciated on CT examinations (►**Fig. 22**).

Angiosarcoma–Angiosarcoma is the most common primary nonhematopoietic malignant tumor of the spleen. It is a very rare and aggressive tumor comprising less than 2% of all soft-tissue sarcomas with poor prognosis, and usually with diffuse metastasis to the liver, lungs, bones and lymphatic structures or splenic rupture at presentation. It has been associated with radioactive contrast, thorotrast, and chemotherapy for lymphoma and breast cancer.[70-72] Multiple complex heterogeneously enhancing masses are seen on contrast-enhanced CT with areas of internal hemorrhage and necrosis accounting for this appearance. On MR imaging, variable high and low signals on T1- and T2-weighted images
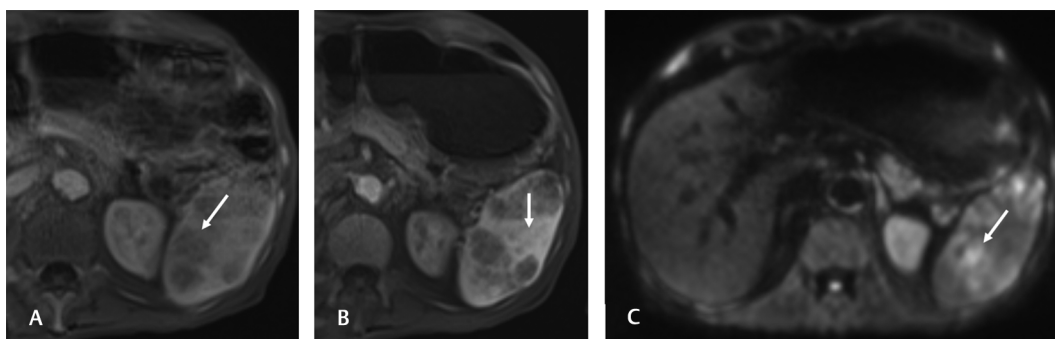
Fig. 22 Axial fat-suppressed postcontrast T1-weighted images of the abdomen showing enlarged spleen with multiple hypointense nodules (arrows in **A**), which show heterogeneous postcontrast enhancement (arrows in **B**) and diffusion restriction (arrow in **C**) in biopsy proven lymphoma.
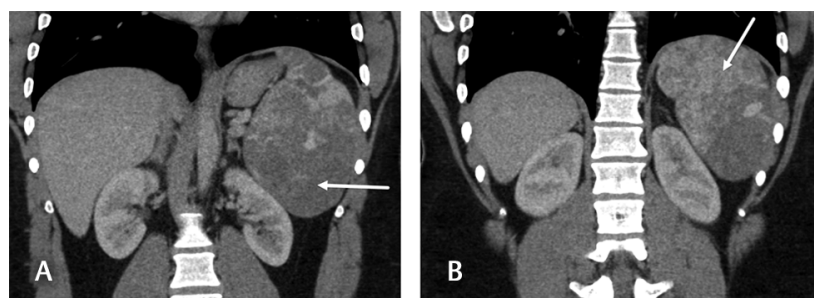

Fig. 23 Coronal postcontrast images showing an enlarged spleen with heterogeneous areas of internal necrosis (arrow in **A**) and enhancement (arrow in **B**). Courtesy: Dr. Victoria Chernyak, M.D.
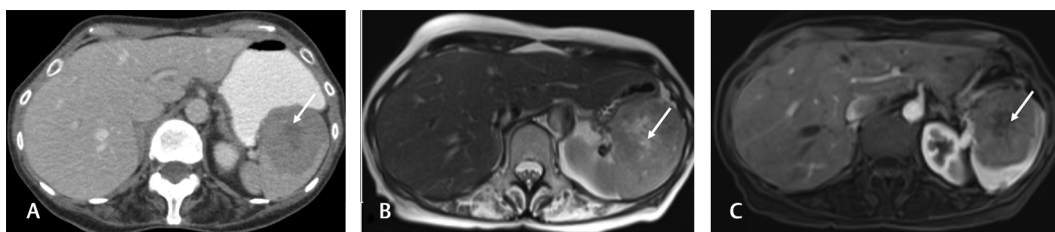

Fig. 24 Axial postcontrast CT (arrow in **A**) showing a heterogeneously enhancing lesion in the upper pole of spleen. Axial T2-weighted image and axial T1 fat-suppressed postcontrast image (arrows in **B** and **C**) showing the same lesion; biopsy showed metastatic ovarian adenocarcinoma.

are seen with heterogeneous postcontrast enhancement within solid components of the mass on postgadolinium enhanced images. Internal blood products of different ages, siderotic nodules, and necrosis may also be seen (►**Fig. 23**).

Metastasis–Spleen is an uncommon site for metastasis, likely due to lack of afferent lymphatics and plethora of lymphatic tissue within the gland. Splenic involvement is seen in diffuse metastatic disease spreading through hematogenous route. Primary malignancies metastasizing to the spleen include melanoma, breast, lung, ovary, colon, stomach, and pancreas.[73-75]

Splenic metastasis can be solitary or multiple. On portal venous phase of contrast-enhanced CT, they appear as hypodense lesions which may show "target appearance" secondary to rim enhancement. At MR imaging, metastases may be hyperintense on T2-weighted images due to necrosis or cystic change. They may be hypo- to isointense on T1-weighted images and difficult to see, but on postgadolinium sequences, heterogeneous contrast enhancement with

peripheral ring-like pattern similar to CT can be observed. Melanoma metastasis may show intrinsic precontrast T1 signal secondary to intrinsic paramagnetic properties of melanin and/or blood products (►**Fig. 24**).

Miscellaneous Conditions

Peliosis–Peliosis is an unusual condition characterized by formation of variable-sized haphazardly organized blood-filled spaces in the spleen, which when located along the splenic surface may result in hemorrhage due to rupture. The rupture maybe posttraumatic or iatrogenic when these lesions get inadvertently biopsied. The dusky and purple color of the spleen on macroscopic examination contributes to its name. Peliosis has been reported in patients with use of anabolic steroids, hematologic conditions such as myeloma and aplastic anemia, and medical debilitating diseases such as tuberculosis, acquired immuno deficiency disorder (AIDS) and cancer.[76] On US, lesions are ill-defined and hypoechoic
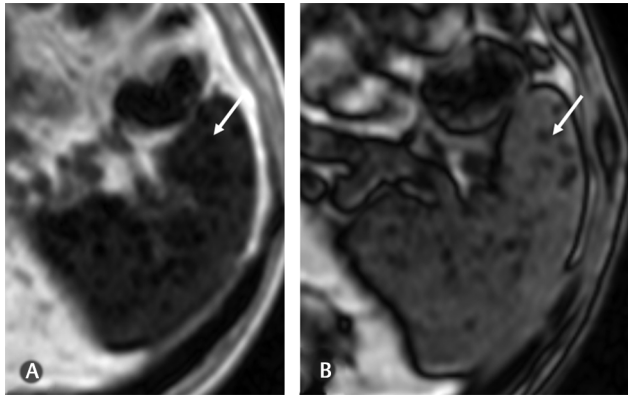
Fig. 25 Axial T1-weighted MR images of the spleen showing small T1 hypointense nodules, which show loss of signal on the in phase image (arrow in **A**) compared with out of phase (arrow in **B**), consistent with siderotic nodules in patient with known cirrhosis.

with nonspecific imaging appearance. Variable patterns are described on CT such as well-defined multiloculated hypoattenuating lesions with septations on noncontrast CT. Septal enhancement can be seen on postcontrast imaging. Hypoattenuating lesions with fluid hematocrit levels can also be seen. Subcapsular hemorrhage can result in calcifications on CT. On MRI, lesions appear as complex cystic spaces and have variable T1 and T2 signals, based on the stage of hemorrhage within them. Heterogeneous postcontrast enhancement can also be seen.

Gamna Gandy Bodies

Gamna Gandy bodies, also known as siderotic nodules, are formed from episodes of microbleeding into the splenic parenchyma with organization of iron, fibrous tissue, and multinucleated giant cells around sites of hemorrhage. Conditions predisposing to increased splenic circulatory pressure and resulting in hemorrhages such as portal hypertension, hematologic causes (such as sickle cell disease), and malignancies are the most common causes.[77] On US, these nodules appear as small echogenic foci in an enlarged spleen. On CT, they may calcify and appear as hyperdense foci. On MRI, they appear as small hypointense rounded foci on T2WI, which show drop out of signal on in phase T1WI and blooming artifact on gradient echo (GRE)-based sequences due to presence of hemosiderin (►**Fig. 25**).

Conclusion

The spleen serves a wide range of hematological and immunological functions and can be involved in several common conditions that affect other solid abdominal organs. Knowledge of varied radiological appearances of splenic pathology on various cross-sectional imaging modalities is crucial to accurately diagnose these conditions, some of which may have surgical implications.

Conflict of Interest
None declared.

References

1   Elsayes KM, Narra VR, Mukundan G. Lewis JS Jr, Menias CO, Heiken JP. MR imaging of the spleen: spectrum of abnormalities. Radiographics 2005;25(4):967–982

2   Kucybała I, Ciuk S, Tęczar J. Spleen enlargement assessment using computed tomography: which coefficient correlates the strongest with the real volume of the spleen? Abdom Radiol (NY) 2018;43(9):2455–2461

3   Yetter EM, Acosta KB, Olson MC, Blundell K. Estimating splenic volume: sonographic measurements correlated with helical CT determination. AJR Am J Roentgenol 2003;181(6):1615–1620

4   Sjoberg BP, Menias CO, Lubner MG, Mellnick VM, Pickhardt PJ. Splenomegaly: a combined clinical and radiologic approach to the differential diagnosis. Gastroenterol Clin North Am 2018;47(3):643–666

5   Shan GD, Chen WG, Hu FL, et al. A spontaneous hematoma arising within an intrapancreatic accessory spleen: A case report and literature review. Medicine (Baltimore) 2017; 96(41):e8092

6   Yildiz AE, Ariyurek MO, Karcaaltincaba M. Splenic anomalies of shape, size, and location: pictorial essay. Scientific World Journal 2013;2013:321810

7   Applegate KE, Goske MJ, Pierce G, Murphy D. Situs revisited: imaging of the heterotaxy syndrome. Radiographics 1999;19(4):837–852, discussion 853–854

8   Lake ST, Johnson PT, Kawamoto S, Hruban RH, Fishman EK. CT of splenosis: patterns and pitfalls. AJR Am J Roentgenol 2012; 199(6):W686–93

9   Remtulla M, Drury NE, Kaushal NA, Trotter SE, Kalkat MS. Thoracic splenosis masquerading as advanced lung cancer. Thorax 2017;72(2):189–190

10 Valli M, Arese P, Gallo G, Flecchia D. Right retroperitoneal splenosis presenting as an adrenal mass. Eur J Surg 1999;165(12): 1197–1198

11 Toh WS, Chong RC, Tan CH, Ding C, Vishalkumar S. Intrahepatic splenosis mimicking hepatocellular carcinoma in a patient with non-alcoholic fatty liver disease: a case report. HPB (Oxford) 2019;21:S854

12 Tsai LL, Kaliannan K, Mortele KJ. Gallbladder splenosis: a hereto unreported mimicker of a gallbladder neoplasm. Clin Imaging 2015;39(2):318–320

13 Carman KA, Walters BS, Kennedy AP. Abdominal splenosis with acute torsion of an omental splenule. J Pediatr Surg Case Rep 2018;32:53–54

14 El-Kheir A, Abdelnour M, Boutros JG. Simultaneous small bowel and colon obstruction due to splenosis. A case report and review of literature. Int J Surg Case Rep 2019;58:63–66

15 Desai DC, Hebra A, Davidoff AM, Schnaufer L. Wandering spleen: a challenging diagnosis. South Med J 1997;90(4):439–443

16 Karlo CA, Stolzmann P, Do RK, Alkadhi H. Computed tomography of the spleen: how to interpret the hypodense lesion. Insights Imaging 2013;4(1):65–76

17 Piccolo CL, Trinci M, Pinto A, Brunese L, Miele V. Role of contrast-enhanced ultrasound (CEUS) in the diagnosis and management of traumatic splenic injuries. J Ultrasound 2018;21(4):315–327

18 Kozar RA, Crandall M, Shanmuganathan K, et al; AAST Patient Assessment Committee. Organ injury scaling 2018 update: Spleen, liver, and kidney. J Trauma Acute Care Surg 2018;85(6): 1119–1122

19 Jabbour G, Al-Hassani A, El-Menyar A, et al. Clinical and radiological presentations and management of blunt splenic

trauma: a single tertiary hospital experience. Med Sci Monit 2017;23:3383–3392

20 Madoff DC, Denys A, Wallace MJ, et al. Splenic arterial interventions: anatomy, indications, technical considerations, and potential complications. Radiographics 2005;25(Suppl 1): S191–S211

21 Sadat U, Dar O, Walsh S, Varty K. Splenic artery aneurysms in pregnancy–a systematic review. Int J Surg 2008;6(3): 261–265

22 Jesinger RA, Thoreson AA, Lamba R. Abdominal and pelvic aneurysms and pseudoaneurysms: imaging review with clinical, radiologic, and treatment correlation. Radiographics 2013;33(3):E71–E96

23 Weber SM, Rikkers LF. Splenic vein thrombosis and gastrointestinal bleeding in chronic pancreatitis. World J Surg 2003;27(11):1271–1274

24 Goerg C, Schwerk WB. Splenic infarction: sonographic patterns, diagnosis, follow-up, and complications. Radiology 1990;174(3 Pt 1) :803–807

25 Llewellyn ME, Jeffrey RB, DiMaio MA, Olcott EW. The sonographic "bright band sign" of splenic infarction. J Ultrasound Med 2014;33(6):929–938

26 Hyun BH, Varga CF, Rubin RJ. Spontaneous and pathologic rupture of the spleen. Arch Surg 1972;104(5):652–657

27 Jiménez BC, Navarro M, Huerga H, López-Vélez R. Spontaneous splenic rupture due to Plasmodium vivax in a traveler: case report and review. J Travel Med 2007;14(3):188–191

28 Dimitrakakis G, Von Oppell U, Zilidis G, Srivastava A. Splenic rupture complicating aortic valve replacement for bacterial endocarditis. Interact Cardiovasc Thorac Surg 2008;7(1):138–140

29 Fernández de Larrea C, Cibeira MT, Rovira M, Rosiñol L, Esteve J, Bladé J. Spontaneous rupture of the spleen as immediate complication in autologous transplantation for primary systemic amyloidosis. Eur J Haematol 2008;80(2):182–184

30 Khalife H, Muwakkit S, Al-Moussawi H, et al. Spontaneous splenic rupture in a patient with factor XIII deficiency and a novel mutation. Pediatr Blood Cancer 2008;50(1):113–114

31 Bhan P, Al-Hilli S. Spontaneous rupture of the spleen in Factor XIII deficiency: a report of two cases. Pol J Radiol 2010;75(1):81–83

32 de Bree E, Tsiftsis D, Christodoulakis M, Harocopos G, Schoretsanitis G, Melissas J. Splenic abscess: a diagnostic and therapeutic challenge. Acta Chir Belg 1998;98(5):199–202

33 Chang KC, Chuah SK, Changchien CS, et al. Clinical characteristics and prognostic factors of splenic abscess: a review of 67 cases in a single medical center of Taiwan. World J Gastroenterol 2006;12(3):460–464

34 Ralls PW, Quinn MF, Coletti P, Lapin SA, Halls J. Sonography of pyogenic splenic abscess. AJR 1982;138(3):523–525

35 Fotiadis C, Lavranos G, Patapis P, Karatzas G. Abscesses of the spleen: report of three cases. World J Gastroenterol 2008;14(19):3088–3091

36 Ahmad QA, Ahmed MS. Splenic hydatid, a rare presentation of hydatid disease. Annals 2010;16:129–131

37 Dilli A, Tatar IG, Ayaz UY, Hekimoglu B. Isolated splenic hydatid disease. Case Rep Med 2011;2011:763895

38 WHO Informal Working Group. International classification of ultrasound images in cystic echinococcosis for application in clinical and field epidemiological settings. Acta Trop 2003;85(2):253–261

39 Lewall DB, McCorkell SJ. Hepatic echinococcal cysts: sonographic appearance and classification. Radiology 1985; 155(3):773–775

40 Kamaya A, Weinstein S, Desser TS. Multiple lesions of the spleen: differential diagnosis of cystic and solid lesions. Semin Ultrasound CT MR 2006;27(5):389–403

41 Uriarte C, Pomares N, Martin M, Conde A, Alonso N, Bueno MG. Splenic hydatidosis. Am J Trop Med Hyg 1991;44(4): 420–423

42 Jhaveri K, Vakil A, Surani SR. Sarcoidosis and its splenic wonder: a rare case of isolated splenic sarcoidosis. Case Rep Med 2018;2018:4628439

43 Ganeshan D, Menias CO, Lubner MG, Pickhardt PJ, Sandrasegaran K, Bhalla S. Sarcoidosis from head to toe: what the radiologist needs to know. Radiographics 2018;38(4): 1180–1200

44 Labruzzo C, Haritopoulos KN, El Tayar AR, Hakim NS. Posttraumatic cyst of the spleen: a case report and review of the literature. Int Surg 2002;87(3):152–156

45 Urrutia M, Mergo PJ, Ros LH, Torres GM, Ros PR. Cystic masses of the spleen: radiologic-pathologic correlation. Radiographics 1996;16(1):107–129

46 Thut D, Smolinski S, Morrow M, et al. A diagnostic approach to splenic lesions. Applies Radiology 2017;46(2):7–22

47 Webb WR, Brant WE, Major NM, Fundamentals of body CT. Philadelphia: Saunders; 2014

48 Palas J, Matos AP, Ramalho M. The spleen revisited: an overview on magnetic resonance imaging. Radiol Res Pract 2013;2013:219297

49 Peene P, Wilms G, Stockx L, Rigauts H, Vanhoenacker P, Baert AL. Splenic hemangiomatosis: CT and MR features. J Comput Assist Tomogr 1991;15(6):1070–1072

50 Pui MH, Ellis JV. Magnetic resonance imaging of splenic hemangiomatosis. Can Assoc Radiol J 1994;45(3):225–227

51 Kaza RK, Azar S, Al-Hawary MM, Francis IR. Primary and secondary neoplasms of the spleen. Cancer Imaging 2010; 10(1):173–182

52 Ramani M, Reinhold C, Semelka RC, et al. Splenic hemangiomas and hamartomas: MR imaging characteristics of 28 lesions. Radiology 1997;202(1):166–172

53 Vilanova JC, Capdevila A, Aldomà J, Delgado E. Splenic epithelioid hemangioma: MR findings. AJR Am J Roentgenol 1994;163(3):747–748

54 Disler DG, Chew FS. Splenic hemangioma. AJR Am J Roentgenol 1991;157(1):44

55 Thompson SE, Walsh EA, Cramer BC, et al. Radiological features of a symptomatic splenic hamartoma. Pediatr Radiol 1996; 26(9):657–660

56 Ohtomo K, Fukuda H, Mori K, Minami M, Itai Y, Inoue Y. CT and MR appearances of splenic hamartoma. J Comput Assist Tomogr 1992;16(3):425–428

57 Yu RS, Zhang SZ, Hua JM. Imaging findings of splenic hamartoma. World J Gastroenterol 2004;10(17):2613–2615

58 Wadsworth DT, Newman B, Abramson SJ, Carpenter BL, Lorenzo RL. Splenic lymphangiomatosis in children. Radiology 1997;202(1):173–176

59 Morgenstern L, Bello JM, Fisher BL, Verham RP. The clinical spectrum of lymphangiomas and lymphangiomatosis of the spleen. Am Surg 1992;58(10):599–604

60 Abbott RM, Levy AD, Aguilera NS, Gorospe L, Thompson WM. From the archives of the AFIP: primary vascular neoplasms of the spleen: radiologic-pathologic correlation. Radiographics 2004;24(4):1137–1163

61 Levy AD, Abbott RM, Abbondanzo SL. Littoral cell angioma of the spleen: CT features with clinicopathologic comparison. Radiology 2004;230(2):485–490

62 Bhatt S, Huang J, Dogra V. Littoral cell angioma of the spleen. AJR Am J Roentgenol 2007;188(5):1365–1366

63 Martel M, Cheuk W, Lombardi L, Lifschitz-Mercer B, Chan JK, Rosai J. Sclerosing angiomatoid nodular transformation (SANT): report of 25 cases of a distinctive benign splenic lesion. Am J Surg Pathol 2004;28(10):1268–1279

64 Kim HJ, Kim KW, Yu ES, et al. Sclerosing angiomatoid nodular transformation of the spleen: clinical and radiologic characteristics. Acta Radiol 2012;53(7):701–706

65 Wang TB, Hu BG, Liu DW, Gao ZH, Shi HP, Dong WG. Sclerosing angiomatoid nodular transformation of the spleen: a case report and literature review. Oncol Lett 2016;12(2):928–932

66 Lewis RB, Lattin GE Jr, Nandedkar M, Aguilera NS. Sclerosing angiomatoid nodular transformation of the spleen: CT and MRI features with pathologic correlation. AJR Am J Roentgenol 2013;200(4):W353–60

67 Bhatia K, Sahdev A, Reznek RH. Lymphoma of the spleen. Semin Ultrasound CT MR 2007;28(1):12–20

68 Ahmann DL, Kiely JM, Harrison EG Jr, Payne WS. Malignant lymphoma of the spleen. A review of 49 cases in which the diagnosis was made at splenectomy. Cancer 1966;19(4):461–469

69 Rabushka LS, Kawashima A, Fishman EK. Imaging of the spleen: CT with supplemental MR examination. Radiographics 1994;14(2):307–332

70 dos Santos Silva I, Malveiro F, Jones ME, Swerdlow AJ. Mortality after radiological investigation with radioactive Thorotrast: a follow-up study of up to fifty years in Portugal. Radiat Res 2003;159(4):521–534

71 Falk S, Krishnan J, Meis JM. Primary angiosarcoma of the spleen. A clinicopathologic study of 40 cases. Am J Surg Pathol 1993;17(10):959–970

72 Thompson WM, Levy AD, Aguilera NS, Gorospe L, Abbott RM. Angiosarcoma of the spleen: imaging characteristics in 12 patients. Radiology 2005;235(1):106–115

73 Schön CA, Görg C, Ramaswamy A, Barth PJ. Splenic metastases in a large unselected autopsy series. Pathol Res Pract 2006;202(5):351–356

74 Lam KY, Tang V. Metastatic tumors to the spleen: a 25-year clinicopathologic study. Arch Pathol Lab Med 2000;124(4):526–530

75 Hahn PF, Weissleder R, Stark DD, Saini S, Elizondo G, Ferrucci JT. MR imaging of focal splenic tumors. AJR Am J Roentgenol 1988;150(4):823–827

76 Davidson J, Tung K. Splenic peliosis: an unusual entity. Br J Radiol 2010;83(990):e126–e128

77 Halefoglu AM. Case series: Gamna-Gandy bodies of the spleen: a supportive finding for portal hypertension. Indian J Radiol Imaging 2007;17:186–187