Primary vascular neoplasms of the spleen constitute the majority of non-hematolymphoid splenic tumors. The benign primary vascular tumors include hemangioma, hamartoma, and lymphangioma, whereas those of variable or uncertain biologic behavior include littoral cell angioma, hemangioendothelioma, and hemangiopericytoma. The primary malignant vascular neoplasm of the spleen is angiosarcoma. Peliosis is a rare lesion of unknown cause that is usually found incidentally in asymptomatic patients but may be associated with hematologic or metastatic disease. Although these vascular neoplasms of the spleen are uncommon, their importance lies in that they must be differentiated from the more common neoplastic disorders of the spleen, such as lymphoma and metastasis. The most common echogenic solid or complex cystic mass in an asymptomatic patient is splenic hemangioma. However, the imaging appearance of splenic hemangiomas may be complex, and differentiation of these lesions from malignant disease may not be possible. The diagnosis of splenic hamartoma may be suggested when findings of increased blood flow on color Doppler images are seen in association with a homogeneous solid echogenic mass. A large subcapsular solitary cystic abnormality discovered incidentally in a child in association with internal septations and tiny mural nodules favors the diagnosis of lymphangioma. Any invasion of the surrounding splenic parenchyma by a splenic lesion should indicate a more aggressive or malignant process. Evaluation of a focal splenic abnormality identified on sonograms should be followed up with computed tomography or magnetic resonance imaging with and without contrast material enhancement. Splenectomy may be required for definitive evaluation of a splenic mass with atypical features.

Abbreviations: AIDS = acquired immunodeficiency syndrome, H-E = hematoxylin-eosin

Index terms: Angioma, 775.3194 • Hamartoma, 775.314 • Hemangioendothelioma, 775.3193 • Hemangiopericytoma, 775.3199 • Lymphangioma, 775.3194 • Sarcoma, 775.322 • Spleen, neoplasms, 775.314, 775.3193, 775.3194, 775.3199, 775.322

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Introduction

Primary splenic tumors are uncommon and are classified as lymphoid tumors, nonhematolymphoid tumors, and tumorlike lesions. Vascular neoplasms are the most common primary nonhematolymphoid tumors and arise from the vascular elements that compose splenic red pulp. In contrast, splenic white pulp is composed of lymphatic tissue and gives rise to lymphoid neoplasms.

Clinically, there is a wide range of presenting signs and symptoms that may be encountered in patients with primary vascular neoplasms of the spleen. These patients are frequently asymptomatic, and the neoplasms are discovered incidentally during an imaging evaluation conducted for other reasons. On the other hand, the clinical presentation may be dramatic because these tumors may cause spontaneous splenic rupture and massive hemoperitoneum.

The biologic behavior of primary vascular splenic neoplasms ranges from benign to malignant. Hemangioma is the most common benign primary neoplasm of the spleen. Other less common benign vascular neoplasms include hemangioendothelioma, lymphangioma, and littoral cell angioma. Although rare, angiosarcoma is the most common malignant primary vascular neoplasm of the spleen. Hemangioendothelioma is considered by some authors to represent an intermediate entity between hemangioma and angiosarcoma. Hemangiopericytoma is most often encountered as a soft-tissue primary tumor, but occasionally it originates in the spleen. Hemangiopericytoma is often characterized as having variable biologic behavior, but it is regarded as a tumor that may have very high malignant potential. Finally, peliosis is rare nonneoplastic tumorlike disorder that is included in the discussion of primary vascular neoplasms because its imaging and pathologic appearances mimic those of neoplasms.

This article summarizes the current literature and our experience with primary vascular neoplasms of the spleen. Knowledge of the clinical, pathologic, and radiologic spectrum of these neoplasms is important because of the wide spectrum of clinical and radiologic manifestations. Herein, we review the clinical, pathologic, and radiologic features of these entities, with emphasis on those characteristics that may allow a more specific diagnosis.

Normal Spleen

The spleen is an intraperitoneal organ that is normally located in the left upper quadrant of the abdomen and supported by the gastrospenic and
The spleen contains two distinct tissues: red pulp and white pulp (Fig 3). The red pulp is composed of slender and nonanastomosing arterial vessels, thin-walled venous vessels called splenic sinuses, plates of cells called splenic cords that lie between sinusoids, and red pulp veins that drain the sinusoids (Fig 3) (1). The white pulp is composed of lymphatic tissue. The organization of lymphoid cells within the white pulp is similar to that found in the cortex of a lymph node. T cells are usually found in the periairteriolar sheath (Fig 3a), and B cells are found in primary and secondary follicles (Fig 4). Lymphoid follicles (malpighian corpuscles) have a central artery that is surrounded by a germinal center, mantle zone, and marginal zone (Fig 4). The marginal zone is the transition between the white and red pulp.
On cross-sectional images, the normal spleen is typically located in the left upper quadrant, although its position and orientation may vary considerably from person to person. On sonograms, the spleen has homogeneous mid- to low-level echotexture (Fig 5). On computed tomographic (CT) scans, the normal spleen enhances in a mottled pattern during the arterial (peak aortic enhancement) and early portal venous phases of intravenous contrast material enhancement. The mottled enhancement pattern is caused by variable flow rates through the cords and sinuses of the splenic red pulp (3). The splenic parenchyma becomes homogeneous in the middle to late portal venous phases of contrast enhancement. The equilibrium phase of contrast enhancement typically shows washout of contrast material (Fig 6).

The normal splenic parenchyma has low signal intensity on T1-weighted magnetic resonance (MR) images and high signal intensity on T2-weighted MR images. MR images obtained after dynamic administration of a gadolinium contrast agent may show a homogeneous or mottled pattern of enhancement during the arterial phase (4). The mottled pattern is similar to that observed on arterial phase contrast-enhanced CT scans and is secondary to variable flow rates through the cords and sinuses of the red pulp (Fig 7).

Figures 5, 6. (5) Transverse sonogram of the normal spleen shows a diffusely homogeneous mid-level echotexture throughout the spleen. (6) Contrast-enhanced CT images of the normal spleen show a mottled pattern of enhancement in the early portal venous phase (a), homogeneous enhancement during the portal venous phase (b), and washout of contrast material during the equilibrium phase of contrast enhancement (c).
Hemangioma

Clinical Features
Hemangioma, although rare, is the most common benign primary neoplasm of the spleen. Its prevalence at autopsy ranges from 0.3% to 14%, and hemangiomas are found most often in adults from mid-30s to mid-50s years of age (5–7). Some studies indicate that hemangioma occurs with equal frequency among men and women, but others indicate that there is a slight predominance among men (8). Most hemangiomas are small lesions that are found incidentally and patients usually have no symptoms. The natural course of hemangiomas is slow growth, and symptoms or complications, when present, occur late. Hemangiomas can become large and manifest as a palpable nontender mass in the left upper quadrant. Generalized splenomegaly may be present, but results of laboratory evaluation are often normal.

Splenic hemangiomas may occur as part of generalized angiomatosis as seen in Klippel-Trénaunay syndrome. Complications include rupture, hypersplenism, and malignant degeneration (7,9). Spontaneous rupture has been reported as the most common complication, occurring in 25% of patients (9). Kasabach-Merritt syndrome (anemia, thrombocytopenia, and coagulopathy) has been reported in patients with large hemangiomas (10). Splenectomy is curative for patients with symptoms.

Pathologic Features
Splenic hemangiomas are thought to be congenital in origin, arising from sinusoidal epithelium. Histopathologic evaluation reveals a nonencapsulated proliferation of vascular channels of variable size, ranging from capillary to cavernous, which are lined with a single layer of endothelium filled...
Figure 8. Splenic hemangioma. (a) Photomicrograph (original magnification, ×4; H-E stain) shows a solitary, nonencapsulated splenic hemangioma (arrows) that is well demarcated from the adjacent normal spleen. The hemangioma is composed of multiple blood-filled spaces. (b) Photomicrograph (original magnification, ×40; H-E stain) shows the blood-filled spaces of the hemangioma (H) lined by flat endothelial cells (arrows).

Figure 9. Splenic hemangiomatosis in a 9-year-old boy with Klippel-Trénaunay syndrome and worsening left upper quadrant pain. (a) Longitudinal sonogram of the spleen shows splenomegaly and multiple echogenic masses (arrows). (b) Axial T1-weighted MR image shows multiple subtle hypointense masses (arrow) throughout the spleen. (c) Axial T2-weighted MR image shows several high-signal-intensity masses (arrow). (d) Photograph of the cut surface of the resected spleen shows multiple spongy masses (arrow). Central fibrosis (arrowhead) is present in several of the hemangiomas.
with red blood cells (Fig 8) (11). These blood-filled spaces are separated by thin fibrous septa or splenic pulp tissue. Splenic hemangiomas are most frequently cavernous and may be single or multiple (12). In diffuse angiomatosis, neoplastic vascular channels may replace the whole spleen. Gross examination will reveal solitary or multiple blue-red spongy nodules in the spleen (Fig 9). Size is variable, but most lesions are less than 2 cm in diameter. These lesions may be intrasplenic or protruding and can be solid or cystic. Smaller hemangiomas, both capillary and cavernous, tend to be solid, whereas larger cavernous lesions can develop thrombosis, infarction, fibrosis, and pseudocystic degeneration caused by necrosis.

Solid hemangiomas are firm and darker in color than normal splenic red pulp. Completely solid hemangiomas may contain both vascular channels and areas of fibrosis, but they do not have cystic spaces. Fibrotic changes, which appear light-gray to white, result in a relative decrease in the overall proportion of the remaining blood-filled vascular channels. Cystic hemangiomas are composed of cystic spaces of various sizes that are filled with serous or hemorrhagic fluid due to necrosis. Calcium deposits may be present in firm fibrotic areas of the mass or in the surrounding intratumoral cystic spaces (5).

**Radiologic Features**

The radiologic appearance of hemangioma ranges from solid to cystic, depending on gross morphology, but predominantly a hemangioma appears as a solid mass with cystic spaces (5,13). On radiographs, the lesion may manifest as a mass in the left upper quadrant or as splenomegaly. Calcification, when present, appears as multiple small punctate calcifications or peripheral curvilinear calcifications.

On sonograms, a hemangioma may manifest as a well-defined intrasplenic or pedunculated echogenic solid or complex cystic mass (Fig 9). Echogenic calcifications with acoustic shadowing may be present (Fig 10).

On unenhanced CT scans, capillary hemangiomas appear as hypoattenuating or isodense, well-marginated masses. Homogeneous and marked contrast enhancement occurs during intravenous administration of contrast material.
Cavernous hemangiomas have a combination of solid and cystic components. The solid component appears isoattenuating or hypoattenuating relative to normal spleen, with enhancement of only the solid tissue (Fig 12). Ferrozzi et al (14) have described late- or delayed-phase contrast enhancement of cavernous hemangiomas as discrete mottled areas of heterogeneous attenuation rather than centripetal enhancement as seen in the liver. This finding likely reflects the fact that the cystic spaces, often central in location, do not contain blood-filled vascular channels. Curvilinear or eggshell calcification is most often seen in cystic hemangiomas. Mottled central calcification is more common in solid lesions, whereas coarse calcification may be found in areas of necrosis or long-standing thrombosis (Fig 10b) (5,14).

The MR imaging appearance of splenic hemangiomas has been described as being similar to that of hepatic hemangiomas (8,15). Splenic hemangiomas are hypointense to isointense, compared with normal spleen, on T1-weighted images and hyperintense on T2-weighted images (Fig 9). Dynamic MR imaging after administration of gadopentetate dimeglumine has shown that splenic hemangiomas have three patterns of enhancement: (a) immediate homogeneous enhancement that persists, (b) early peripheral enhancement with uniform delayed enhancement, and (c) peripheral enhancement with centripetal progression but persistent enhancement of a central fibrous scar (16). Others have described central persistent low signal intensity on contrast-enhanced T1-weighted images in those lesions with a central scar (8). However, larger hemangiomas may have a variable MR imaging appearance be-
cause of complicating features such as hemorrhage, infarction, and thrombosis (Fig 13). As a result, differentiation of a splenic hemangioma from malignant disease may not be possible solely on the basis of MR imaging findings.

Ramani et al (8) reported that both splenic hemangiomas and hepatic hemangiomas demonstrate peripheral enhancement with centripetal progression. However, splenic hemangiomas do not have well-defined peripheral nodules that coalesce over time. This characteristic is thought to reflect the differences in vascular supply to the background organ rather than inherent differences between splenic and hepatic hemangiomas (8). This hypothesis suggests that enhancing peripheral nodules (seen in hepatic hemangiomas on both contrast-enhanced CT and MR images) may be less conspicuous in splenic hemangiomas during the arterial phase because splenic enhancement obscures them. In addition, hemangiomas in the spleen may be less conspicuous compared with those in the liver because of the longer T2 values of the splenic parenchyma on T2-weighted images.

The contrast enhancement patterns of splenic hemangiomas as seen on CT and MR images and described by Ferrozzi et al (14) and Ramani et al (8), respectively, appear to differ with regard to progressive centripetal enhancement. In the Ramani et al series, 19 of 22 lesions demonstrated a progressive centripetal pattern of enhancement with dynamic contrast enhancement techniques. In addition, two small splenic hemangiomas in their series demonstrated early uniform enhancement that persisted on delayed T1-weighted images. These findings correlate with the pattern of enhancement of capillary splenic hemangiomas on CT scans as reported by Ferrozzi et al. (Note that Ramani et al did not classify the majority of the lesions into capillary or cavernous splenic hemangiomas, unlike Ferrozzi et al, who did.) To our knowledge, there is no additional evidence to support the finding that the contrast enhancement patterns of splenic hemangiomas would differ significantly between current CT and MR imaging techniques. Moreover, a consistent finding on both CT and MR contrast-enhanced images is that larger splenic hemangiomas appear to lack well-defined enhancing peripheral nodules.

Hamartoma

Clinical Features

Hamartomas, which are also known as splenomas, splenadenomas, or nodular hyperplasia of the spleen, are rare benign lesions first described by Rokitansky in 1861 (17). Approximately 120 cases of splenic hamartoma have been reported in the English-language literature. Review of autopsy series has shown that the incidence of splenic hamartoma ranges from 0.024% to 0.13% (18). Hamartomas may occur at any age with equal gender predilection. Most patients have no symptoms, and the discovery of a splenic hamartoma is an incidental finding. Larger lesions may manifest with a palpable mass, splenomegaly, or rupture. Thrombocytopenia and anemia may occur from sequestration of hematopoietic cells. Signs and symptoms usually associated with larger lesions are more common in female patients (19).

Hamartomas of the spleen have been associated with hamartomas elsewhere in the body and have been reported in cases of tuberous sclerosis and Wiskott-Aldrich–like syndrome (17,20–22). The association of splenic hamartoma with tuberous sclerosis lends support to the hamartomatous
nature of the latter condition. In addition, an association of splenic hamartoma with malignancy has been suggested. In a study by Lam et al, three of six splenic hamartomas were associated with neoplastic diseases (18,23).

Pathologic Features
Splenic hamartoma is a malformation composed of an anomalous mixture of normal splenic red pulp elements. The hamartoma is thought to be congenital in origin, reflecting a focal developmental disturbance in the spleen. Some consider a splenic hamartoma to be a neoplasm (a form of hemangioma or lymphangioma) or possibly a post-traumatic lesion (17,24). Still others believe that splenic hamartoma might arise from an acquired proliferative process, a theory that supports the association of hamartoma with malignancy (23).

Hamartomas are usually well-circumscribed, solid, bulging nodular lesions that tend to compress the adjacent parenchyma. They are most often solitary but may manifest as multiple nodules. Their gross appearance is typically dark red to grayish-white, and lesions up to 19 cm in size have been reported (18). Despite their well-defined appearance at gross examination, hamartomas do not appear well defined at microscopic analysis. Their expansile growth compresses the surrounding red pulp, which can be demonstrated with a reticulum stain. They lack fibrous trabeculae, but focal sclerosis as well as minute calcifications may be present (1). At histopathologic examination, they contain a mixture of unorganized vascular channels lined by endothelial cells and surrounded by fibrotic cords of predominant splenic red pulp with or without (lymphoid) white pulp (Fig 14) (17). No organized lymphoid follicles (malpighian corpuscles) are present. Additional findings include plasmacytosis; extramedullary hematopoiesis; and increased numbers of macrophages, eosinophils, and mast cells (17,25).

Pathologic differentiation of hamartoma from hemangioma may be difficult. It has been suggested that hamartomas are sclerosed angiomas (6). When immunohistochemical techniques are used, the endothelial cells of splenic hamartoma are CD8 positive (Fig 14c), whereas those of hemangioma are not (26). Hamartomas contain sinus and pulp cordlike elements, whereas hemangiomas encompass well-organized lymphoid tissue. When sclerosis is a predominant feature, a
diagnosis of a sclerosed hemangioma rather than hamartoma is suggested (1). In patients with Hodgkin lymphoma, the presence of a hamartoma may be confused with lymphomatous involvement of the spleen (17). However, the absence of Reed-Sternberg cells and the compression (rather than invasion) of the surrounding parenchyma allow differentiation of splenic hamartoma from lymphoma.

**Radiologic Features**

The radiologic features of splenic hamartoma have been previously described (14,16,27–32). Sonography appears to be more sensitive than CT (30). On sonograms, splenic hamartomas are typically solid homogeneous masses, but some may be heterogeneous with cystic changes or, in rare cases, some contain coarse calcification secondary to ischemia or hemorrhage (33,34). Most hamartomas are hyperechoic relative to the adjacent normal splenic parenchyma. On color Doppler images, these lesions often demonstrate increased blood flow (Fig 15) (27). On angiograms, a splenic hamartoma appears as a hypervascular mass consisting of tumor vessels with aneurysmal dilatation, arteriovenous shunts, vascular lakes, and tumor blush resembling a typical malignant vascular pattern (35). The typical hypervascularity seen with both color Doppler imaging and angiography is thought to reflect the hypervascularity of the red pulp within the hamartoma. However, a single case of an avascular hamartoma with associated marked calcification has been described (36).

On CT scans, hamartomas often appear nearly isoattenuating relative to normal spleen before and after intravenous administration of contrast material and, therefore, can be difficult to detect; however, they can also appear heterogeneous with contrast enhancement (Fig 16). A contour abnormality may be the only finding present (30,33,34,37).
Ramani et al (8) have shown that splenic hamartomas are isointense relative to normal splenic parenchyma on T1-weighted MR images, heterogeneously hyperintense on T2-weighted MR images, and heterogeneously diffusely enhanced on T1-weighted MR images obtained immediately after gadolinium injection (Fig 17). On delayed MR images, hamartomas were shown to have a more uniform homogeneous pattern of enhancement (8). When visible at CT and MR imaging, hamartomas are well-defined masses with smooth borders and no infiltration of the surrounding splenic parenchyma.

The importance of imaging splenic hamartomas lies in the need to differentiate them from malignant lesions of the spleen such as lymphoma and metastasis. Hamartoma should be included in the differential diagnosis of a focal splenic mass that demonstrates substantial increased blood flow at color Doppler imaging. Although the radiologic features seen with splenic hamartoma may be suggestive of the diagnosis, a definitive preoperative diagnosis can rarely be made on the basis of imaging findings alone.

**Lymphangioma**

**Clinical Features**

Splenic lymphangioma is a relatively rare benign tumor with clinical manifestations that range from an asymptomatic incidental finding to a large multicentric, symptomatic mass requiring surgical intervention. Most splenic lymphangiomas occur in children, with adult cases being reported less frequently. Splenic lymphangiomas usually initially grow without causing significant clinical effects; when the lesions cause symptoms, they are related to compression of adjacent structures. There is a close relationship between the occurrence of symptoms and splenic size (38). Abdominal symptoms related to the size of the spleen include left upper quadrant pain, nausea, and abdominal distention. The complications associated with more extensive or larger lymphangiomas of the spleen include bleeding, consumptive coagulopathy, hypersplenism, and portal hypertension (39,40).

Lymphangiomatosis is a syndrome in which multiple organs are involved. Frequent anatomic sites of involvement include the mediastinum, retroperitoneum, axilla, and neck. Case reports in
which cystic hygroma of the neck was a synchronous or metachronous finding have been described (41–43). In children with lymphangiectasis, Wadsworth et al (44) reported simultaneous involvement in the liver, pericardium, mediastinum, lung, and bone. These findings suggest a possible relationship between the occurrence of multiorgan involvement and patient age. In a young patient with splenic lymphangioma, the diagnostic evaluation should be extended to include extrasplenic organs (44).

Treatment of incidental or small lesions typically does not require surgery. However, symptomatic larger lesions have generally been treated with splenectomy or partial splenectomy. Although percutaneous aspiration techniques are safe, they appear to have little value in long-term control of splenic lymphangiomas (45).

Pathologic Features

The pathologic appearance of splenic lymphangioma covers a broad spectrum, which includes solitary nodules, multiple nodules, and diffuse lymphangiomatosis. Lymphangiomas of the spleen are usually microcystic or solid and may show central scarring (42,46). On the basis of size and location of the vascular channels, lymphangiomas can be divided into three types: capillary, cavernous, and cystic (41). Unlike the random localization seen with hemangiomas, lymphangioma often involves the capsule and trabeculae of the spleen, where lymphatics are normally concentrated (1).

Of the solitary focal lesions, the subcapsular lymphangioma is most common. Similar-appearing satellite lesions may surround a larger lesion. A solitary focal lymphangioma may also be intraparenchymal. With larger multifocal lesions, the tumors are separated by distinct residual splenic tissue and they cause the spleen to be nodular and enlarged.

Lymphangioma may also manifest as a large solitary cyst or may be part of diffuse lymphangiomatosis involving the spleen. With lymphangiomatosis, the spleen may be diffusely replaced by expanding lymphangiomas that leave little remaining splenic parenchyma (47,48). Conversely, the spleen may not be extensively involved in lymphangiomatosis, but vasoformative malformations or tumors may be found in other organs. The liver is the most frequent secondary organ involved (43).

As seen at histologic analysis, capillary, cavernous, and cystic lymphangioma each consists of a single layer of flattened endothelium-lined spaces, which are filled with eosinophilic proteinaceous material instead of blood as seen in hemangiomas (Fig 18). When the histologic characteristics are not clear, the endothelial origin of the cyst may be established with immunohistochemical techniques that demonstrate reactivity for factor VIII, a finding that confirms the diagnosis of lymphangioma (49). At gross examination, these cysts have thick fibrous walls with an internal morphology characterized by fibrous trabeculae. Hyalinization and calcification of the fibrous connective tissue may be present (50).

Both splenic lymphangioma and hemangioma are vasoformative tumors and they are closely related, but splenic lymphangioma is less common. There is no firm consensus as to the exact origin of splenic lymphangioma, which may represent a hamartomatous rather than a neoplastic lesion (1,51). Others have proposed a unified concept of lymphangioma and cystic hygroma as being a congenital developmental defect (52). There have been very few reports of a splenic lymphangioma developing into malignant lymphangiosarcoma (53). When atypical morphology of the endothelial cells is present, careful long-term follow-up of the affected patients is recommended to determine whether these features indicate malignancy or simply an unusual expression of the endothelial cells (1).

Radiologic Features

Lymphangioma of the spleen is often found incidentally at sonographic and CT examinations performed for another reason. The spleen may be of normal size or splenomegaly may be present. Sonography and CT typically reveal splenic cysts
of various sizes, ranging from a few millimeters to several centimeters in diameter. On sonograms, these cystic lesions appear as well-defined hypoechoic masses with occasional internal septations and intralocular echogenic debris (13). Tiny echogenic calcifications that correspond to histologic findings may be identified. Color Doppler sonography can demonstrate the vasculature of the mass, including the intrasplenic arteries and veins along the cyst walls. With a large mass, color Doppler sonography can also help determine the organ of origin by demonstrating the vessels at the splenic hilum (38). Angiography typically reveals an avascular mass.

On CT scans, lymphangiomas appear as single or multiple thin-walled low-attenuation masses with sharp margins that are typically subcapsular in location (Fig 19). No significant contrast enhancement is typically seen. The presence of curvilinear peripheral mural calcifications suggests the diagnosis of cystic lymphangioma (Fig 20) (54,55).

On T1-weighted MR images, the cystic lesions appear hypointense relative to the surrounding visceras. However, high T1 signal intensity may result from internal bleeding or the presence of large amounts of intracystic proteinaceous content. T2-weighted images demonstrate multiloculated hyperintense areas that correspond to the dilated lymphatic spaces. The intervening septa appear as hypointense bands, corresponding to the presence of fibrous connective tissue (50).
The mural calcifications best seen at CT are difficult to identify at MR imaging. However, because of high contrast resolution, MR imaging may prove useful in the detection of solid elements within the cystic lumen in the very rare case in which malignant degeneration may be present (53).

**Littoral Cell Angioma**

**Clinical Features**

Littoral cell angioma of the spleen is a rare vascular tumor that was first described in 1991 (56). Although these tumors were originally thought to be benign, their biologic behavior has not been firmly established because there have been several reports of littoral cell angioma with malignant features (57,58). Littoral cell angiomas may occur at any age and have no gender predilection. Typically, patients with littoral cell angioma are found to have a splenic abnormality when they are being evaluated for laboratory evidence of anemia or thrombocytopenia (56, 59,60). Other systemic symptoms such as fever, chills, weakness, fatigue, and pain have been reported (61). Splenomegaly is almost always present. An association between littoral cell angioma and other malignancies, including colorectal, renal, and pancreatic adenocarcinoma and meningioma, has been described (62). In most patients, because symptomatic hematologic problems are present and because the imaging findings are nonspecific, splenectomy is typically performed for definitive evaluation and treatment.

**Pathologic Features**

Littoral cell angioma is a neoplasm with characteristic morphologic and immunophenotypic features that distinguish it from other vascular splenic tumors. The gross examination of the spleen containing littoral cell angioma typically reveals splenomegaly. On the cut surface of the spleen, multiple focal nodules of similar size corresponding to spongolike vascular spaces are usually present. Nodules range in color from red to black, an appearance that reflects the presence of blood products of variable age. These nodules are typically well delineated from surrounding splenic tissue but do not have a surrounding capsule. Tiny cystic spaces associated with the nodular lesions, and corresponding to the vascular spaces seen histologically, may be seen as well.

Littoral cell angioma arises from littoral cells that originate from the splenic red pulp sinuses and that have features intermediate between those of endothelial cells and macrophages. Littoral cell angioma is characterized histologically by anastomosing vascular channels lined by tall or flat endothelial cells, which may anastomose with normal splenic sinuses at the periphery. The vascular channels may have a pseudopapillary aspect or may appear as dilated cavernous vascular spaces.
Plump exfoliated cells are often found within the lumina of the vascular channels. Foci of extramedullary hematopoiesis, hemosiderin pigment (Fig 21c), and calcification have been reported at histopathologic examination (59). The pathologic diagnosis can be confirmed with immunohistochemical staining, which reveals that the lining cells are positive for endothelial marker factor VIII-related antigen and histiocytic markers CD68 and lysozyme (1, 59,61). Differentiation of littoral cell angioma from angiosarcoma is determined by the absence of irregular anastomosing vascular channels and the presence of cellular atypia, mitoses, or invasion of surrounding organs.

**Radiologic Features**

Littoral cell angioma should be considered in the differential diagnosis of multiple splenic lesions in patients with evidence of hypersplenism. The appearance of littoral cell angioma on sonograms, CT scans, and MR images has been previously reported (59,61,63,64). All modalities usually demonstrate splenomegaly and multiple lesions of similar size and appearance.

The sonographic appearance of littoral cell angioma is variable and includes reports of mottled echotexture without discrete lesions, as well as findings of isoechoic, hypoechoic, and hyperechoic lesions (Fig 22) (60,61,63,65). When the nodular lesions of littoral cell angioma are hyperechoic, the differential diagnosis is narrower and includes hemangiomatosis, hamartoma, and Kaposi sarcoma in patients with acquired immunodeficiency syndrome (AIDS) (66).

On abdominal CT scans obtained with or without contrast material, littoral cell angioma typically manifests as multiple hypovascular lesions. Lesions with this appearance have a broad differential diagnosis, including other primary vascular tumors of the spleen, other neoplastic entities, infection, and systemic diseases such as sarcoidosis. However, on delayed contrast-enhanced images, littoral cell angiomas homogeneously enhance and become isoattenuating relative to the remaining splenic parenchyma, a finding that may help limit the differential diagnosis (Fig 23) (65).

Levy et al (59) described a remarkable uniformity to the clinical characteristics and CT features in eight cases of littoral cell angioma. The unifying CT feature was identified as innumerable splenic masses (Fig 24). Lesions up to 6 cm in diameter were reported, with shapes ranging...
**Figure 22.** Sonographic features of littoral cell angioma. (a) Longitudinal sonogram of the spleen in a 50-year-old woman with asymptomatic splenomegaly shows a heterogeneous splenic echotexture with multiple hyperechoic masses. (b) Transverse sonogram of the spleen in a 75-year-old man with asymptomatic splenomegaly shows a focal hyperechoic mass (arrow).

**Figure 23.** Littoral cell angioma in a 55-year-old woman who was found to have splenomegaly when she was evaluated for leg swelling. (a) Contrast-enhanced CT scan obtained during the early portal venous phase shows multiple, partially confluent hypoattenuating masses in the spleen. (b) Contrast-enhanced CT scan obtained during the late portal venous phase shows that the masses homogeneously enhance and become imperceptible.

**Figure 24.** Littoral cell angioma in a 39-year-old woman who was found to have splenomegaly when she was being evaluated for peptic ulcer disease. (a) Contrast-enhanced CT scan shows the enlarged spleen, which contains innumerable hypoattenuating masses. (b) Photograph of the cut surface of the resected spleen shows multiple blood-filled spaces (arrow).
from round to geographic. Most lesions had distinct margins, but ill-defined margins and coalesced lesions were also identified (59). No capsular calcification or tiny cystic spaces associated with the nodular lesions seen at histologic examination are identified at CT. Significant abdominal adenopathy, which is typically seen in patients with splenic metastasis and lymphoma, is not present in patients with littoral cell angioma.

On MR images, the nodular lesions of littoral cell angioma typically appear markedly hypointense with both T1- and T2-weighted pulse sequences, a finding that reflects the presence of hemosiderin in the lesions due to the hemophagocytic capacity of the neoplastic cells (Fig 25).

**Peliosis**

**Clinical Features**

Peliosis is a rare disease, characterized by multiple blood-filled spaces in the spleen, that usually occurs in conjunction with peliosis hepatitis (67). Approximately 40 cases of isolated splenic peliosis have been reported in the literature (68). Most cases have an association with use of anabolic steroids; hematologic disorders such as aplastic anemia; and wasting diseases such as tuberculosis, AIDS, and cancer (69,70). Peliosis is often discovered incidentally, but its clinical significance lies in the potential of peliotic lesions on the surface of the spleen to rupture and cause life-threatening intraperitoneal hemorrhage. Several fatal cases have been reported (71,72). Rupture of the lesions can occur spontaneously or secondary to minimal trauma. Careful consideration must be given before needle biopsy of a suspected peliotic lesion is performed (70). As a result, the definitive diagnosis and treatment of peliosis of the spleen is made with splenectomy.

**Pathologic Features**

Pathologic examination of splenic peliosis reveals dark red cavities of various sizes within the splenic parenchyma. Histopathologic analysis reveals many different sized, blood- or thrombi-filled lakes without an endothelial lining (Fig 26). Peliosis differs from splenic hemangioma in that the blood-filled spaces are haphazardly scattered in the red pulp, and there is preferential involvement of the parafollicular areas of the spleen (1,67).
Radiologic Features
On sonograms, peliosis of the spleen appears as an echogenic mass in the left upper quadrant with numerous poorly defined foci of varying hypoechochogenicity. The same mass on unenhanced CT scans appears as a hypoattenuating, multiloculated lesion with well-defined septa. On contrast-enhanced CT images, the lesion demonstrates significant enhancement with loss of definition of the lobules and septa (72). Others have reported multiple hypoattenuating nodules with fluid-fluid levels, which are thought to reflect a hematocrit effect, that show enhancement in their dependent portions (70). Most commonly, peliosis manifests as multiple small, well-defined hypoattenuating lesions on CT scans (Fig 27). Calcification or extracapsular extension is not seen. However, if lesions rupture, subcapsular hematoma and intraperitoneal hemorrhage may be evident at CT.

The radiologic differential diagnosis of peliosis includes hemangiomatosis, lymphangioma, and angiosarcoma. Other entities would be considered less likely based on both the imaging and clinical findings.

Hemangiopericytoma
Clinical Features
Hemangiopericytoma is characterized as a vascular tumor with variable biologic behavior and is known to have relatively high malignant potential. This tumor, first described by Stout and Murray...
in 1942, is a rare vascular lesion that appears to arise from Zimmerman pericytes (73). Hemangiopericytoma rarely originates in the spleen as a primary tumor and was first reported by Guadajara Jurado et al (74) in 1989. Almost 50% of these tumors arise in the lower extremities and soft tissues, and 25% have an abdominal origin. When present in the spleen, hemangiopericytoma is typically asymptomatic or results in splenomegaly (74,75).

The treatment of choice for splenic hemangiopericytoma is wide surgical excision. The prognosis in these patients is uncertain, with recurrence rates as high as 50% (74). Hemangiopericytoma originating in the abdomen, excluding the stomach and uterus, behaves aggressively (76). The most common site of recurrence is local, followed by lung and bone for distant metastasis (77). Careful long-term follow-up is required to confirm ongoing stability, because recurrence has been shown to occur 20 years after initial treatment (78).

Pathologic Features
At gross examination, hemangiopericytoma has been described as well-defined, slightly gray intrasplenic nodules (79), with or without associated hemorrhage and necrosis. At histologic analysis, splenic hemangiopericytoma appears identical to the same tumor that occurs most frequently in the muscles of the lower extremities and subcutaneous soft tissues. Hemangiopericytoma, which appears microscopically well defined and nonencapsulated, consists of pericytes proliferating around vascular channels lined with endothelium and may occur wherever there are capillaries. A reticulum cell dye technique is used to demonstrate that the tumor cells are external to the basement membrane of the vascular channels (80). Immunohistochemical stain for factor VII shows no reactivity for tumor cells and reactivity only for endothelial cells of normal vessels. No white pulp is present within these tumors, a characteristic that allows the diagnosis of hamartoma to be excluded.

Radiologic Features
In a report of multiple hemangiopericytomas, the lesions were seen as hypoechoic nodules on sonograms but were not identified on either CT scans or technetium-99m sulfur colloid scintigrams (79). However, these lesions were observed on MR images and had low signal intensity with T1-weighted pulse sequences and high signal intensity with T2-weighted pulse sequences (79). Reported CT findings in hemangiopericytoma include a large splenic mass with polylobular contours and smaller disseminated lesions throughout the spleen (16). In addition, speckled calcification may be seen on CT scans, and contrast-enhanced studies show discrete hyperattenuation of solid portions and septations (14). These tumors often bleed because of their hypervascular nature and expansive growth, findings that can be identified at CT.

Hemangioendothelioma

Clinical Features
The clinical characteristics of hemangioendothelioma are often nonspecific, with patients presenting with left upper quadrant pain or a palpable mass. Patients may also have hematologic abnormalities, evidence of hypersplenism, and metastatic disease. Splenic hemangioendothelioma appears to occur more frequently in the young adult population, but pediatric cases have been reported (81). No apparent gender predilection has been described.

Pathologic Features
Hemangioendothelioma is a very rare primary vascular tumor of the spleen and has variable malignant potential, as reflected in both its histologic appearance and biologic behavior (82). From a histopathologic and clinical standpoint, hemangioendothelioma is thought to represent an intermediate entity between hemangioma and angiosarcoma (81,83,84). However, because the morphologic appearance of hemangioendothelioma is highly variable, there is some debate as to its existence as a distinct pathologic entity. It is thought that many cases previously reported in the literature as splenic hemangioendothelioma are in fact angiosarcomas (1,85,86).

In a patient without evidence of metastatic disease, the gross pathologic appearance of hemangioendothelioma has been described as that of a large well-circumscribed, nonencapsulated solid splenic mass (83). Its histologic appearance may range from patterns that are well differentiated to highly undifferentiated forms, and the tumor is composed of vascular and stromal elements. Immunohistochemical studies demonstrate positive staining with antibodies to factor VIII-related antigen and Ulex europaeus lectin of the endothelial-lined cells in the more defined vascular channels.
Findings that allow hemangioendothelioma to be distinguished from angiosarcoma are absence of dissecting growth and lack of striking cellular atypia.

Radiologic Features
The diagnosis of splenic hemangioendothelioma is not likely to be made on the basis of imaging characteristics alone, since these findings are non-specific. On sonograms, hemangioendothelioma is typically seen as a hypoechoic mass that is distinct from the surrounding splenic parenchyma. Anechoic areas may be present and reflect intratumoral necrosis. On color Doppler images, hemangioendothelioma appears with disordered vascularization and high-velocity arterial flow and a low resistive index in the solid areas of the tumor. These findings are thought to reflect tumoral neoangiogenesis (84).

The typical CT appearance is that of a low-attenuation mass with enhancement of the solid portions of the tumor that may appear hypovascular relative to the normal splenic parenchyma. Findings suggestive of malignancy, such as areas of necrosis and hemorrhage, may be present and will not show evidence of enhancement. Signs of infiltration of the surrounding splenic parenchyma and evidence of metastatic disease may also be identified on CT scans. Calcification has not been described as a specific feature of this tumor. A unique feature of hepatic hemangioendothelioma not seen in the splenic counterpart is retraction of the overlying capsule by peripheral tumors (87). At MR imaging, splenic hemangioendothelioma appears as a heterogeneous solid lesion that may exhibit low signal intensity with both T1- and T2-weighted pulse sequences, an appearance that suggests the presence of hemosiderin.

Angiosarcoma

Clinical Features
Primary angiosarcoma of the spleen is a very rare vascular neoplasm, but it represents the most common nonhematolymphoid malignant tumor of the spleen. It is found more frequently in older patients, with few patients being less than 40 years of age, and no gender predilection is apparent (86,88). Unlike angiosarcoma of the liver, splenic angiosarcoma has no documented association with exposure to carcinogens such as thorium dioxide, vinyl chloride, or arsenic. However, there have been case reports of splenic angiosarcoma associated with previous chemotherapy for lymphoma and radiation therapy for breast cancer (89,90).

Typical symptoms at the time of presentation include abdominal pain, which may be accompanied by constitutional symptoms of fever, fatigue, and weight loss. Clinical complaints may be accompanied by hematologic disorders such as anemia, thrombocytopenia, or other coagulopathy. At physical examination, splenomegaly is almost always identified, and a focal left upper quadrant abdominal mass may be present as well (91). Patients with splenic angiosarcoma may also present with signs and symptoms of hemoperitoneum, since spontaneous rupture is a known complication in up to 30% of patients (86,88,92).

Metastatic disease is common and typically involves the liver, lungs, bone, bone marrow, and lymphatic system (93). Splenectomy is typically performed, but chemotherapy has not been shown to be effective (86). Prognosis is poor and almost all patients die within 1 year of diagnosis.

Pathologic Features
Patients with splenic angiosarcoma typically have massive splenomegaly, with splenic weights often exceeding 1,000 g (88). Cut specimens usually reveal poorly defined nodular masses that are purple or red (Fig 28a). Diffuse involvement of the spleen is common, and replacement of the entire splenic parenchyma with tumor may be seen. Solitary masses do occur, but they are a less common manifestation. Prominent areas of hemorrhage and necrosis are frequently seen within the tumor.

The histologic features of splenic angiosarcoma are similar to those of angiosarcomas seen in other locations. Splenic angiosarcoma appears to arise from splenic sinus endothelial cells, a finding that has been confirmed with immunohistochemical techniques (94). As seen at microscopic evaluation, the tumor consists of disorganized anastomosing vascular channels lined by plump, atypical endothelial cells with large, irregular, hyperchromatic nuclei and a high mitotic rate (Fig 28). Papillary formations and solid areas within the tumor are common. Within the same tumor, the degree of cellular differentiation may vary, but significant nuclear pleomorphism is at least focally present (Fig 28c). Well-differentiated areas appear as splenic sinuslike structures, whereas poorly differentiated areas have sarcomatous features.
Radiologic Features

The imaging appearance of splenic angiosarcoma is that of an aggressive splenic mass, frequently with associated metastatic disease at the time of diagnosis. Sonography, CT, and MR imaging all reveal evidence of marked splenomegaly. On sonograms, the most common appearance is that of a complex mass with heterogeneous echotexture (Fig 29). Cystic areas within the mass are frequently identified and likely reflect areas of necrosis and hemorrhage. Increased Doppler flow may be seen in the more solid echogenic portions of the tumor.

On CT scans, the most common appearance is that of an ill-defined splenic mass with heterogeneous contrast enhancement and areas of necrotic degeneration. Evidence of intraperitoneal hemorrhage is seen in lesions that spontaneously ruptured, and in the acute setting, hemorrhage will appear hyperattenuating on unenhanced images (Fig 30). Scattered punctate calcifications may
**Figure 29.** Sonographic features of splenic angiosarcoma. (a) Transverse sonogram of the spleen in a 72-year-old woman who complained of left upper quadrant pain shows a well-defined mass (arrowheads) of heterogeneous echotexture. (b) Contrast-enhanced CT scan shows a rim-enhancing splenic mass (arrows) and multiple enhancing liver metastases (arrowheads). (c, d) Longitudinal gray-scale (c) and color Doppler (d) sonograms in a 62-year-old woman who presented with Kasabach-Merritt syndrome show splenomegaly with multiple hypoechoic masses (arrows in c) throughout the spleen. Color Doppler US shows increased vascularity (arrow in d) within the masses.

**Figure 30.** Spontaneous hemorrhage within a splenic angiosarcoma in a 58-year-old man who presented with syncope. (a) Unenhanced CT scan shows focal areas of high-attenuation hemorrhage (asterisk) within a splenic mass. (b) Contrast-enhanced CT scan shows a heterogeneously enhancing mass within the spleen.
occasionally be seen (Fig 31), but massive calcification in a radial pattern in a splenic angiosarcoma has also been reported (95). Hypervascular metastasis to the liver as well as metastatic disease to the lungs, bones, and lymphatic system are well demonstrated at CT (Fig 29b).

The MR imaging appearance of splenic angiosarcoma reflects the hemorrhagic nature of the tumor. Areas of increased and decreased signal intensity may be seen on images obtained with both T1- and T2-weighted pulse sequences, findings that are consistent with the presence of blood products and necrosis (Fig 32). Low-signal-intensity areas on MR images have also been shown to represent siderotic nodules that were confirmed at histopathologic examination (96). Contrast-enhanced MR imaging reveals heterogeneous enhancement within the tumor, corresponding to the pathologic findings of solid tumor with areas of necrosis (97).

**Incidental Splenic Mass**

The finding of an incidental splenic mass is frequently encountered at imaging studies that are performed on patients for other reasons. In the absence of findings that suggest metastasis or lymphoma, a primary vascular neoplasm should be considered.

At sonographic evaluation, the most common echogenic solid or complex cystic mass in a patient without symptoms is the splenic hemangioma. Echogenic calcifications, when present, may be helpful in establishing the diagnosis. However, if increased blood flow is present on color Doppler images in association with a homogeneous solid echogenic mass, the diagnosis of splenic hamartoma may be suggested.
A focal splenic abnormality identified at sonography should be further evaluated by means of CT or MR imaging performed with and without contrast material. With contrast-enhanced techniques, small capillary hemangiomas typically reveal early uniform enhancement that persists on delayed images. Cavernous hemangiomas have peripheral enhancement but without well-defined peripheral nodules. It must be emphasized that the MR imaging appearance of splenic hemangiomas may be complex and that differentiation of these lesions from malignant disease may not be possible. However, other findings suggestive of a malignant process should be sought. Any invasion of the surrounding splenic parenchyma indicates a more aggressive or malignant process.

A large subcapsular solitary cystic abnormality with internal septations and tiny mural nodules discovered incidentally in a child favors the diagnosis of lymphangioma. Mural calcifications are difficult to detect on MR images, and intracystic proteinaceous contents appear bright on T1-weighted images. No significant contrast enhancement should be seen in splenic hemangiomas.

Any atypical or unexplained imaging feature identified in an incidental splenic abnormality requires additional imaging evaluation or follow-up. Because several of the primary vascular tumors of the spleen have uncertain biologic behavior, splenectomy may be required for definitive assessment. In patients with an aggressive symptomatic splenic process, splenectomy will almost certainly be performed. Percutaneous biopsy of a suspected vascular splenic tumor should be performed only after careful assessment of the patient’s clotting function, because a higher incidence of complication has been reported in the literature (98).

Conclusions
Primary vascular neoplasms of the spleen represent the majority of nonhematolymphoid tumors of the spleen. The benign primary vascular tumors include hemangioma, hamartoma, and lymphangioma, whereas those of variable or uncertain biologic behavior include lilloar cell angioma, hemangioendothelioma, and hemangipercyctoma. The primary malignant vascular neoplasm of the spleen is angiosarcoma. Peliosis is a rare lesion of unknown cause that is usually found incidentally in patients without symptoms but it may be associated with hematologic disease or metastatic disease.

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References


