Calcified Splenic Lesions: Pattern Recognition Approach on CT With Pathologic Correlation

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OBJECTIVE. Incidental splenic lesions, often found on CT images of the abdomen, may often be ignored or mischaracterized. Calcified splenic lesions are often presumed to be granulomas; however, understanding the broader differential diagnostic considerations can be useful.

CONCLUSION. Determining the cause of splenic lesions is essential to guide appropriate management; the pattern of calcification together with other imaging and clinical findings can aid with differentiation.

Keywords: calcified lesions, cancer, granulomatous disease, spleen

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Incidental splenic lesions such as are found on CT images of the abdomen are often presumed benign and therefore ignored or are sometimes mischaracterized. A study by Siewert et al. [1] found that among incidentally discovered splenic masses greater than 1 cm in diameter, 84% were benign and 16% were malignant. The most common malignancy in the spleen is lymphoma, with the spleen being one of the most commonly involved organs; however, it is rarely the only affected organ [1]. Splenic lesions may also reflect benign systemic abnormalities such as disseminated granulomatous infection or underlying portal venous congestion.

In cases in which splenic lesions on CT may not have a characteristic pattern of attenuation or enhancement, the morphologic features and pattern of calcifications can help the differential diagnosis [2]. Using the imaging characteristics of a splenic mass, radiologists can recommend further imaging, surveillance, or more invasive management [3]. Whereas most calcified splenic lesions are presumed to be sequelae of prior granulomatous infection, there is a broader differential diagnosis for these lesions. An algorithmic approach to splenic lesions according to their characteristic calcification patterns can therefore narrow the differential diagnosis and help guide management.

Splenic Hemangiomas

Splenic hemangiomas are the most common benign primary neoplasm in the spleen, and can be of cavernous or capillary subtype [4]. Patients generally show no symptoms, but when the hemangiomas are large, rupture can occur in up to 25% of cases [4–8]. Preemptive partial splenectomy may therefore be considered for large or symptomatic lesions [5, 6]. Splenic hemangiomas are usually small and exhibit slow growth related to nonencapsulated proliferations of blood vessels [5, 9].

The calcifications within hemangiomas can appear as central punctate, curvilinear, or speckled in areas of thrombosis [6, 9, 10]. Multiphase CT of a splenic hemangioma will typically show a hypoattenuating lesion on unenhanced CT, sometimes with early peripheral discontinuous enhancement with uniform delayed enhancement mirroring the blood pool, although this pattern can be obscured because of background parenchyma enhancement [6] (Fig. 1). Smaller lesions may show flash-filling enhancement, especially among the capillary subtype, and larger lesions will often show centripetal progression of enhancement with a persistently enhancing central fibrous scar [6, 9, 11].

The main consideration for differential diagnosis of these lesions is a malignant lesion with mixed solid and cystic components that presents with inhomogeneous enhancement and central nonenhancement because of necrosis [5, 6, 9].

True Epithelial Cysts

True epithelial cysts present incidentally and are generally asymptomatic; they are also called benign epithelial cysts [12]. True epithelial cysts form during development with outer cuboidal epithelium and inner endothelium and result from infolding of peritoneal mesothelium within the spleen [13].

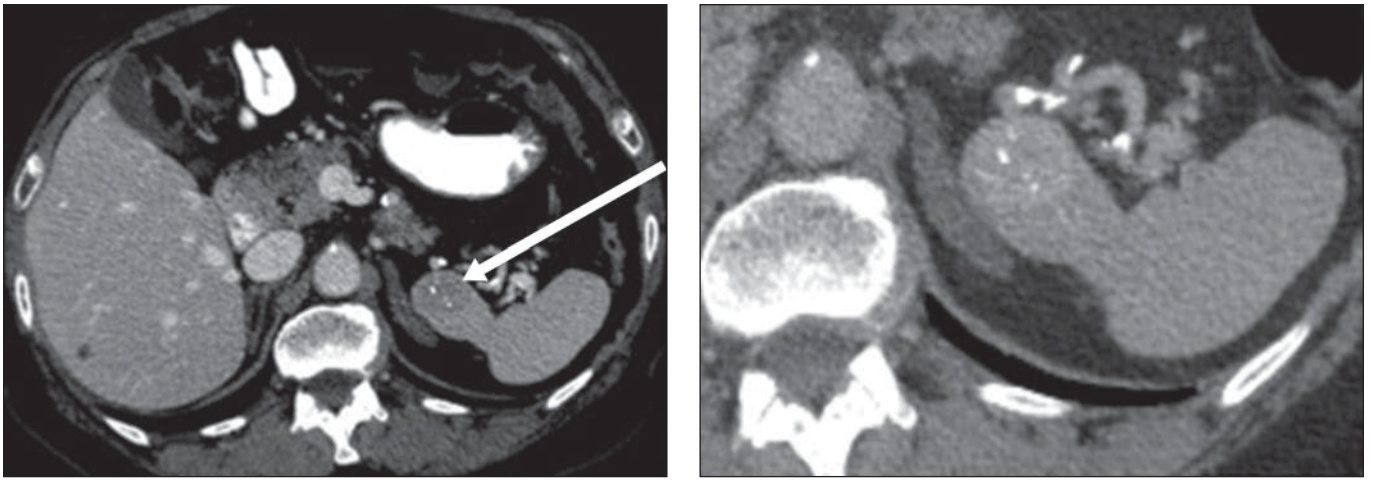

Fig. 1—73-year-old man who presented for staging workup of known prostate cancer.
A and **B,** Axial contrast-enhanced CT images of abdomen and pelvis show incidental splenic mass (*arrow,* **A**) with punctate calcifications and peripheral discontinuous mild enhancement on portal venous phase (**A**) with progressive fill-in on magnified view of delayed phase (**B**). Appearance of lesion was most consistent with small cavernous hemangioma given its imaging characteristics.

Imaging shows a well-defined unilocular cystic mass with walls that can be partially calcified but are otherwise imperceptible. The internal fluid is hypoattenuating and nonenhancing [10, 13]. Some cysts may have internal septations that enhance with contrast administration [10]. Up to 14% of true epithelial cysts can have thin curvilinear wall calcifications, but calcifications can also be seen within septations [12, 13]. When compared with pseudocysts, which are the primary differential diagnostic consideration for these lesions, true epithelial cysts are more likely to have internal enhancing septations but are less likely to have wall calcifications [14].

Cysts that are symptomatic because of large diameter and mass effect may require further management. Spleen-sparing surgery or minimally invasive drainage can be considered for cysts at increased risk of rupture due to large size [7].

Epidermoid Cysts

Epidermoid cysts are among the rarest type of true cysts and are thought to develop from mesothelial cells entrapped within the spleen during early embryonic development [14, 15]. Patients are often children or young adults who typically show no symptoms.

Histopathologic analysis shows an inner lining of stratified squamous epithelium and a wall of fibrous tissue around a fluid-filled unilocular or multilocular cyst [8]. CT shows a well-circumscribed unilocular or multilocular cyst with internal fluid attenuation and rare calcifications [8, 15]. If present, the calcifications will be thin and curvilinear and along the peripheral wall or along the internal septations [8] (Fig. 2A). However, calcifications are much less common in these cysts than in pseudocysts [16].

Epidermoid cysts are managed conservatively. If the patient is symptomatic or if the cyst is greater than 4–5 cm in diameter with increased risk of rupture, spleen-sparing surgery may be considered [7, 8]. Complete splenectomy is avoided because of the resultant increased risk of infections in younger patients [7].

Pseudocysts

Pseudocysts, or false cysts [3], make up 80% of all splenic cystic lesions [10]. Histologically, pseudocysts have no endothelium and may have internal hemorrhage and debris [12, 14]. Unlike other cysts, approximately 50% of pseudocysts show calcifications, whereas calcifications are rarer in true cysts. Pseudocysts occur as a result of prior trauma, pancreatitis, infection, or iatrogenic causes [12].

Splenic injury accounts for approximately 50% of organ injuries in blunt abdominal trauma [16]. A chronic hematoma may result in the formation of a posttraumatic pseudocyst that commonly has peripheral curvi-

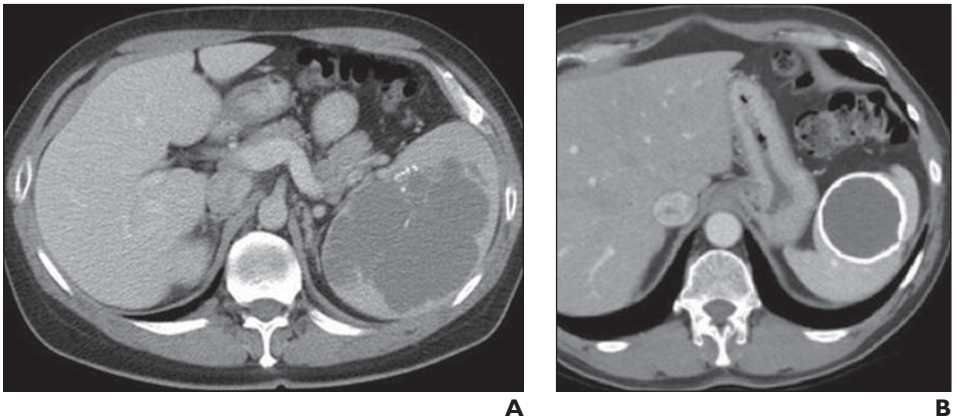

Fig. 2—Patients with asymptomatic cysts.
A, 41-year-old man with no symptoms with incidental splenic mass. Axial contrast-enhanced CT image shows septate, multilocular cystic mass with thin curvilinear calcifications and diffuse splenomegaly. This mass was most consistent with epidermoid squamous cyst given its imaging characteristics.
B, 52-year-old woman with known remote trauma from motor vehicle accident presented for follow-up of known splenic hematoma. Axial contrast-enhanced CT image shows round, well-circumscribed hypoattenuating mass with thick calcified rim, which is consistent with posttraumatic pseudocyst.

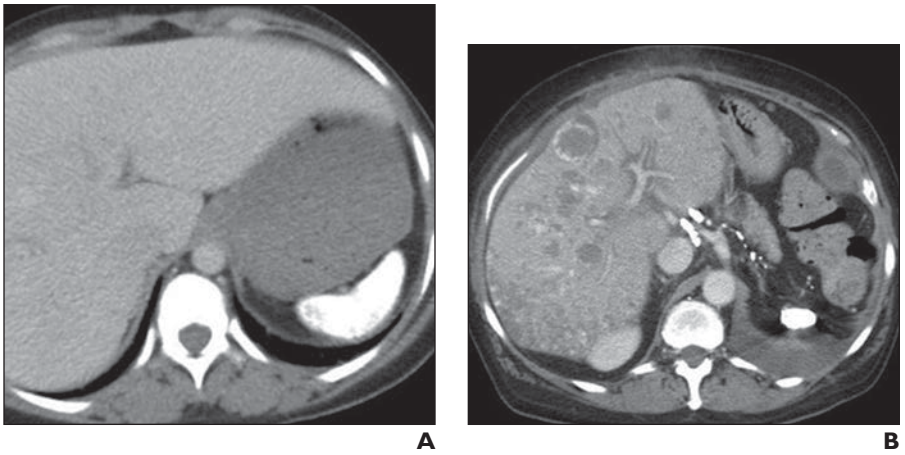

Fig. 3—Patients with shrunken spleen.
A, 20-year-old man with known sickle cell disease at routine follow-up. Axial abdominal contrast-enhanced CT image shows small, dense, calcified spleen resulting from chronic autosplenectomy with underlying sickle cell disease.
B, 64-year-old woman who presented with hepatic angiosarcoma and prior exposure to thorium dioxide (Thorotrast). Axial contrast-enhanced CT image shows peripherally calcified hypoenhancing lesions in liver, multiple calcified intraabdominal lymph nodes, and shrunken calcified spleen.

linear calcifications (Fig. 2B). In addition to those with underlying traumatic causes, splenic hematomas can occur as a result of earlier splenic biopsy or anticoagulation therapy [16]. These posttraumatic cysts may show differential attenuation because of layering blood products and thick, curvilinear or plaquelike calcifications within a thick fibrous wall [12–14].

Patients may need partial or total splenectomy for management because of the high rate of recurrence with percutaneous drainage, especially if the cause is trauma [17, 18].

Autosplenectomy and Splenic Infarction

Splenic infarction in younger patients most commonly occurs in the setting of sickle cell disease, whereas in older patients splenic infarction may be seen with cardiac thromboembolic disease, endocarditis, or atrial fibrillation [3, 19]. Other causes of thromboemboli in patients older than 40 years old may be thrombi that arise from the splenic artery or sinusoidal venous thrombosis in the setting of splenomegaly in the setting of leukemia, lymphoma, or sickle cell disease [14, 19–22].

In early sickle cell disease, abnormal polymerized hemoglobin leads to perfusion-reperfusion injury and resultant slow microcirculation with splenic sequestration [22]. This progresses to congestion, splenomegaly, and eventually multiple small splenic infarcts [10, 22–24]. At this stage, CT may show multiple peripheral wedge-shaped areas of hypoattenuation; however, this ultimately advances to autosplenectomy with little residual functional splenic tissue and diffuse ferrocalcinosis that appears as a small shrunken spleen with coarse confluent calcifications [3, 10, 14, 20–24] (Fig. 3A). Other CT findings in sickle cell disease include hepatic hemosiderosis resulting from transfusional iron overload, extramedullary hematopoiesis, cardiomegaly, vertebral endplate infarcts, osteonecrosis or osteomyelitis, and cholelithiasis resulting from hemolysis with pigmented gallstones [24, 25]. Complications of splenic sequestration resulting from sickle cell disease may rarely include splenic rupture with subcapsular hematoma and hemoperitoneum [26]. Early surgical intervention with splenectomy is preferred to reduce the mortality associated with complications or from the subsequent splenic infarction [19].

Gamna-Gandy Bodies

Gamna-Gandy bodies are asymptomatic nodules that affect 9–12% of patients with portal hypertension and that occur as a result of the deposition of hemosiderin [3, 16]. Pathologically, portal hypertension leads to congestive splenomegaly and then internal foci of hemorrhage and necrosis, in which hemosiderin, calcium, and fibrotic tissue accumulate [27, 28]. The differential diagnosis includes hematologic malignancy, acquired hemochromatosis, and paroxysmal nocturnal hematuria [3, 27].

Gamna-Gandy bodies are best visualized on CT when calcified; they appear as areas of hyperattenuation on unenhanced imaging. They are diffuse, punctate, and less than 1 cm in diameter [28] (Fig. 4). Accompanying findings may include cirrhosis, splenomegaly, varices, or ascites [3]. On MRI, the presence of hemosiderin will result in a loss of signal intensity on both T1- and T2-weighted imaging [28].

The differential diagnosis for these lesions includes sarcoidosis and other granulomatous infections [27].

Calcified Granulomas

Calcified splenic granulomas are a common incidental finding, most commonly resulting from tuberculosis or histoplasmosis and less commonly from *Pneumocystis carinii* pneumonia or brucellosis [29, 30]. A risk factor for many of these infections is HIV infection. In all cases, the granulomas are preceded by mul-

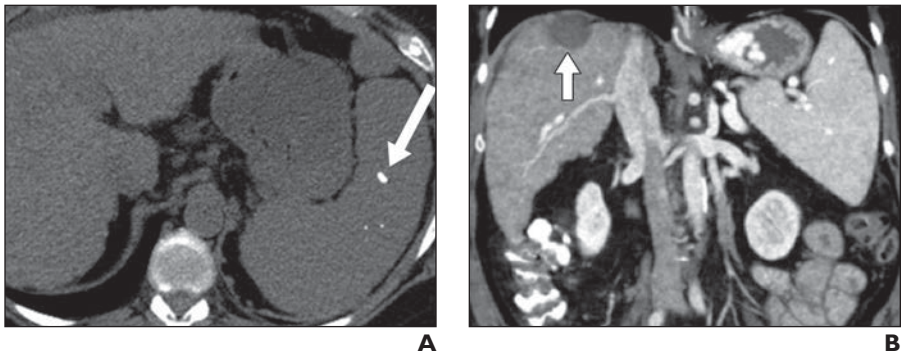

Fig. 4—54-year-old man with cirrhosis and portal hypertension at follow-up after ablation of segment VIII hepatocellular carcinoma.
A, Axial unenhanced CT image shows multiple punctate calcifications within spleen (*arrow*).
B, Coronal reconstruction image from contrast-enhanced CT scan shows scattered punctate splenic calcifications. Cirrhotic liver with area of hypoattenuation (*arrow*) after ablation in hepatic segment VIII is shown.

tiple splenic microabscesses that are hypoattenuating lesions less than 2 cm in diameter. When the microabscesses heal, they may calcify with or without preceding regression. Those that calcify after regression, in cases of histoplasmosis or tuberculosis, may appear as stippled, diffuse calcifications in an otherwise normal spleen [3] (Figs. 5A and 5B). More than five punctate calcifications in the spleen has been described to be more characteristic of histoplasmosis, whereas less than five calcifications may be reflective of splenic involvement in tuberculosis [31]. Meanwhile, ringlike calcifications with splenic infiltration are characteristic of *Pneumocystis carinii* pneumonia resulting from fibrosis and calcification of the microabscesses with central caseating necrosis without preceding regression [3, 29, 30] (Fig. 5C).

Granulomatous infections may present with disseminated disease, including calcified lesions in the liver, lymph nodes, and kidneys, both before and after antimicrobial treatment [10, 12, 29, 30]. Disseminated *Pneumocystis carinii* pneumonia may additionally present with punctate calcifications in the adrenal glands and pleural or peritoneal effusions that subsequently calcify [12, 29].

Echinococcal Cysts

Approximately 1–8% of patients infected with *Echinococcus* species show splenic involvement, although more typically echinococcal cysts are seen in the liver or lungs [3, 16, 32]. The infectious agent is most commonly the *Echinococcus granulosus* tapeworm or less commonly the highly invasive *Echinococcus multilocularis* tapeworm [33]. Patients are infected by *Echinococcus granulosis* tapeworm eggs in contaminated food or from contact with dogs in endemic regions such as the Mediterranean and Middle East, South America, Australia, and New Zealand. *Echinococcus* eggs invade the gastrointestinal mucosa and colonize in the liver via portal venous transmission, from which they can seed the spleen via peritoneal or hematogenous routes [3, 33, 34].

Cysts typically measure up to 8–16 cm in diameter [32]. On CT, these hydatid cysts are typically well-marginated with internal fluid attenuation and occasional air-fluid levels [14, 33]. At all stages, hydatid cysts may have curvilinear, ringlike peripheral calcifications of the pericyst in up to 20–30% of cases [14, 33, 34]. In early

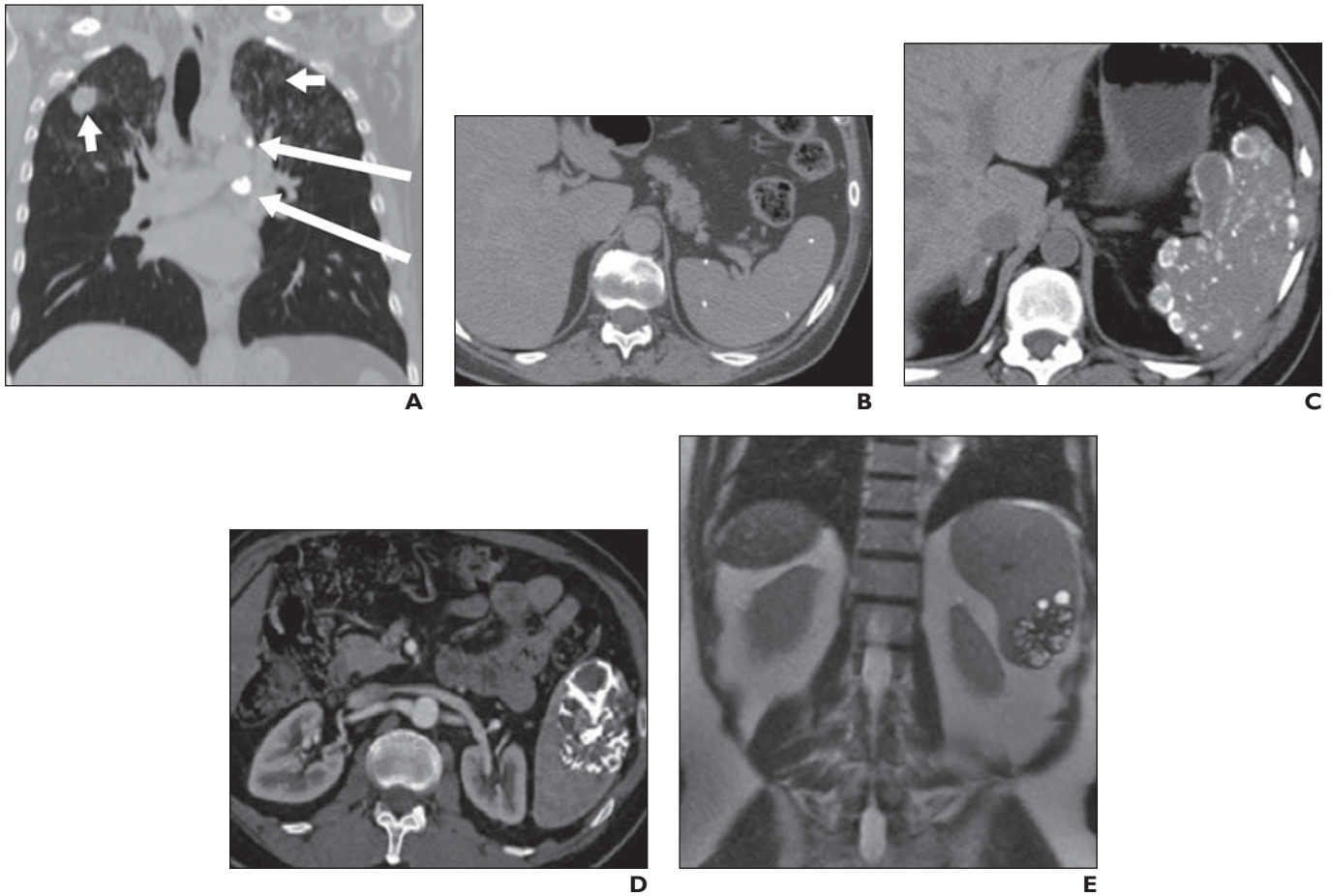

Fig. 5—Cases with infectious causes.
A and **B**, 67-year-old man with pathologically proven histoplasmosis on bronchoscopy and acid-fast culture. Coronal unenhanced CT image shows calcified left hilar nodes (*long arrows*, **A**) and bilateral pulmonary nodules (*short arrows*, **A**) with peribronchial interstitial disease, which is consistent with resolving histoplasmosis. Axial unenhanced CT image (**B**) shows more than five diffuse punctate splenic calcifications, which is consistent with earlier treated granulomatous disease.
C, 55-year-old man with known *Pneumocystis carinii* pneumonia infection at follow-up imaging. Axial contrast-enhanced CT image shows numerous calcified lesions with mild central hypoattenuation that corresponds to caseating necrosis.
D and **E**, 42-year-old woman with known splenic hydatid cyst. Axial contrast-enhanced CT image (**D**) shows multiple daughter cysts and peripheral thick rim calcifications. Coronal T2-weighted MR image (**E**) shows internal increased signal intensity, which is consistent with fluid-filled daughter cysts, and loss of peripheral rim signal intensity, which is consistent with coarse calcifications and blooming artifact.

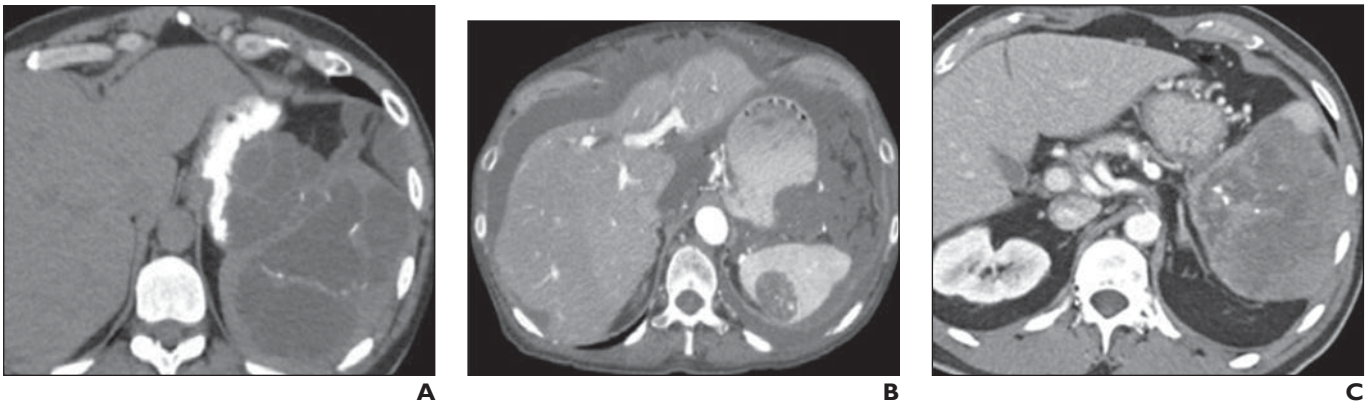

Fig. 6—Cases with secondary neoplasm.
A, 54-year-old man with known metastatic appendiceal mucinous neoplasm. Axial unenhanced CT image of abdomen shows thin peripheral calcifications within walls and septations of complex cystic splenic mass, which is consistent with metastatic disease from primary mucinous appendiceal neoplasm.
B, 57-year-old man with known metastatic colorectal cancer. Axial contrast-enhanced CT image of abdomen shows low-attenuation mass in spleen with stippled internal calcifications and diffusely increased attenuation of intraperitoneal ascites, consistent with peritoneal carcinomatosis.
C, 71-year-old man with high-grade B-cell lymphoma with abdominal pain. Axial contrast-enhanced CT image of mid abdomen shows hypoattenuating complex cystic splenic mass with ill-defined margins, thin calcifications along septations, and associated splenomegaly, consistent with lymphomatous involvement.

disease, type I cysts are unilocular with internal simple fluid attenuation with variable internal calcification or attenuation resulting from hydatid sand from living scolexes [33]. Type II cysts have internal septa, slightly higher internal attenuation resulting from dense debris, and small peripheral wheellike daughter cysts that often have lower internal attenuation than the mother cyst [14, 16, 32, 34]. Type II cysts can also have serpiginous calcifications resulting from collapsed serpentine membranes, producing a ring-and-arc appearance (Figs. 5D and 5E), also called the water lily sign [16, 32]. Type III cysts may be densely calcified with dead scolexes [3, 33, 34]. Type IV cysts are associated with parasitic membrane degeneration that leads to fissures in the cyst wall and rupture of the cyst with superinfection in 50% of cases because of pleural, peritoneal, or biliary seeding [33].

Concurrent imaging findings may include hepatic cysts in 80% of cases [33, 34], which increase the risk for secondary infection resulting from peritoneal seeding or biliary or portal venous spread. Echinococcal infection of the liver and spleen can spread alternatively by direct extension with abdominal wall invasion or into the thorax via the diaphragm [33, 34].

The differential diagnosis includes other cysts, such as epidermoid cysts and pseudocysts. Hydatid cysts can rupture either spontaneously or as a result of trauma, therefore the standard of treatment is spleen-conserving surgery when possible or total splenectomy [35].

Thorotrast Exposure

Exposure to thorium dioxide (Thorotrast) is the most common iatrogenic cause of hepatic angiosarcoma and it can also cause splenic angiosarcoma. Thorotrast is a highly radioactive contrast agent used until the 1950s [36]. Because Thorotrast has a half-life of 22 years, it has great radioactive potential, leading to fibrosis and carcinogenesis, and may eventually lead to malignancy in patients up to 65 years after injection, especially in the liver and spleen because of the biodistribution and clearance [36]. In some cases, patients may present with concurrent spontaneous hemoperitoneum resulting from rupture of the highly vascular tumor [36].

High-density thorium dioxide deposits can be seen in the liver, spleen, and lymph nodes on CT. Radioactivity leads to cell inflammation and fibrosis, which can cause splenic atrophy with a shrunken calcified spleen [36, 37] (Fig. 3B). The liver may show reticular and lacelike calcifications with sur-

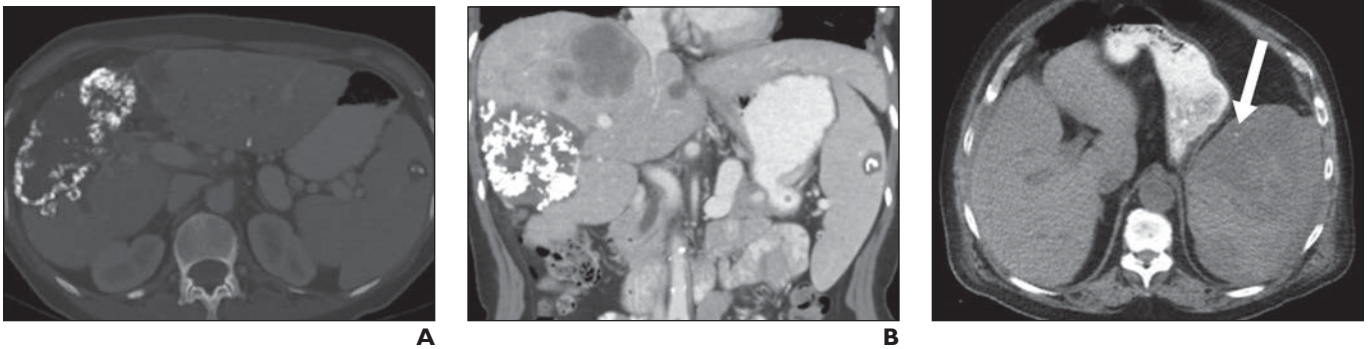

Fig. 7—58-year-old woman with pathologic pelvic fractures resulting from biopsy-proven metastatic epithelioid hemangioendothelioma.
A, Axial contrast-enhanced CT image with bone window shows large liver mass with coarse internal calcifications and smaller partially calcified lesion in spleen.
B, Coronal reconstruction of contrast-enhanced CT image shows multiple hypoattenuating masses of liver, with dominant mass showing coarse calcifications as described in **A** and smaller hypoattenuating, partially calcified mass in spleen.

Fig. 8—40-year-old man with primary splenic angiosarcoma presented with polycythemia vera. Axial unenhanced CT image shows some subtle scattered calcifications at pathologically proven angiosarcoma (*arrow*).

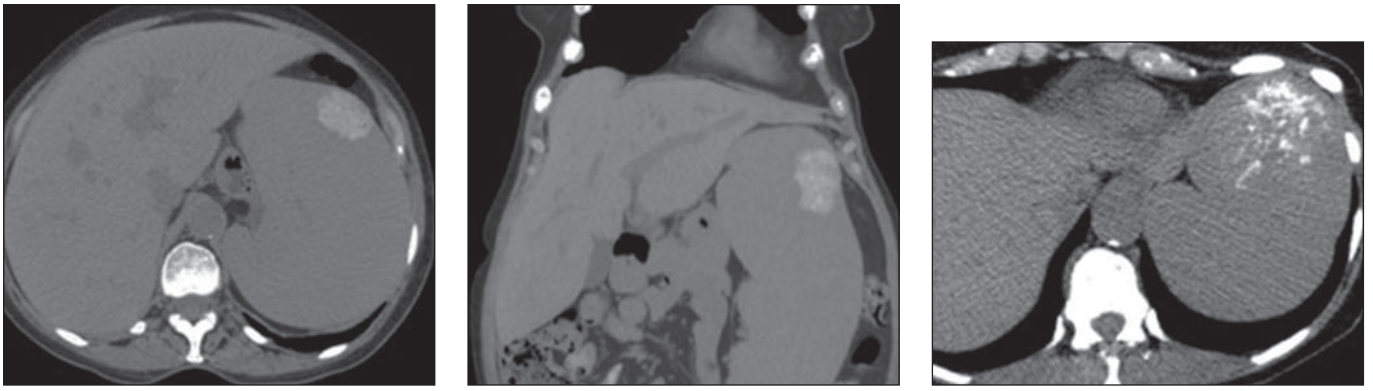

Fig. 9— Myelofibrosis.
A and **B,** 75-year-old woman with known myelofibrosis and massive splenomegaly. Axial unenhanced CT image (**A**) in bone window shows hypoattenuating lesion in anterosuperior spleen with coarse calcifications. Coronal reconstruction of axial CT image (**B**) shows splenic calcified lesion.
C, 69-year-old woman with known myelofibrosis and hepatosplenomegaly. Axial unenhanced CT image shows round anterosuperior splenic lesion with multiple internal coarse calcifications.

rounding hyperattenuating, hemorrhagic ascites [36, 37]. Densely calcified epigastric lymph nodes may be noted [36].

Calcified Splenic Metastases

On autopsy, 7% of patients with a known malignancy were found to have splenic involvement, which is typically asymptomatic in approximately 60% of patients [12, 38]. Among all splenic masses incidentally discovered on CT examination, approximately 1% of the masses were found to be metastatic in origin [1]. The most common malignancies with splenic metastases include melanoma, followed by lymphoma breast carcinoma, lung carcinoma, colorectal carcinoma, ovarian cancer, and gastric cancer, most commonly via hematogenous or peritoneal dissemination [1, 16, 38].

The imaging appearance of a metastatic lesion in the spleen varies with the type of primary cancer [38, 39]. The typical appearance is a hypoattenuating lesion with occasional calcifications and mild enhancement [38], which may be associated with splenomegaly [40]. Metastatic melanoma may present as a cystic lesion [12]. Solitary lesions are most commonly from primary colorectal or ovarian neoplasm [40]. Macronodular, micronodular, or diffuse infiltration patterns are also possible [38, 39]. Surface nodular metastases are most commonly a result of mucinous neoplasm that disseminates throughout the peritoneal cavity, studding the splenic hilum with hypoattenuating cystic lesions that may have faint, linear, or coarse calcifications [38] (Fig. 6A). Ovarian mucinous neoplasm in particular may involve psammomatous calcifications that invade the splenic parenchyma. Associated findings with ovarian cancer or other metastatic cancers may include malignant ascites, peritoneal carcinomatosis, and omental cake [38] (Fig. 6B).

Lymphoma

Lymphoma is the most common malignant tumor in the spleen [12]. Non-Hodgkin lymphoma, particularly diffuse B-cell lymphoma, is the most common subtype of lymphoma in the spleen, with 30–40% of patients with non-Hodgkin lymphoma having splenic involvement, especially those who are at least 60 years old [12, 41, 42]. Most patients already have disseminated lymphoma at presentation [43, 44]. Clinically, patients present with splenomegaly in up to 80% of cases [14], although splenic rupture is a rare complication [45].

Lymphoma infiltrates the splenic white pulp to a greater extent than the red pulp, forming large nodular masses that typically measure greater than 3 cm in diameter [44]. Other common imaging features of lymphoma include splenomegaly and splenic infarcts [16, 44]. Solitary lesions are usually hypo- or isoattenuating with areas of necrosis and rarely have calcifications [41, 44]. However, when present, calcifications can be dystrophic and de novo (Fig. 6C) or, after treatment, secondary to necrosis, hemorrhage, or fibrosis [12]. Splenic involvement is often accompanied by retroperitoneal or localized lymphadenopathy in the splen-

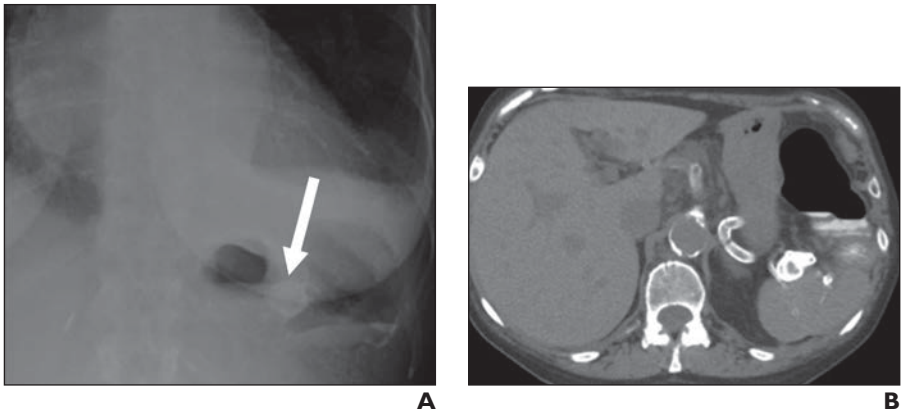

Fig. 10—56-year-old woman showing no symptoms.
A, Chest radiograph shows peripherally calcified lesion (*arrow*) in left upper quadrant.
B, Axial unenhanced CT image shows peripherally calcified hypoattenuating mass in splenic hilum with adjacent thick tram-track calcifications of tortuous splenic artery. Calcified lesion corresponds to splenic artery pseudoaneurysm according to typical location and imaging characteristics.

TABLE 1: Algorithmic Approach to Calcified Splenic Lesions According to CT Findings

Predominant Morphologic Features, Pattern of Calcification	Other Characteristics	Cause
Mass with wall calcifications		
Thin peripheral calcifications	Unilocular cyst; multilocular, septate cyst; hypoattenuating mass, multifocal metastatic involvement (gastrointestinal tract, ovaries); splenomegaly, splenic infarcts, lymphadenopathy	True epithelial cyst; epidermoid cyst; peritoneal metastasis of nonsplenic neoplasm; lymphoma (especially untreated)
Thick curvilinear peripheral calcifications	Unilocular cyst; located at the splenic hilum; multiseptated cystic mass with possible debris and daughter cysts	Pseudocyst; splenic artery aneurysm; echinococcus granulosis infection
Mass with both wall and internal calcifications		
Complete calcification with central clearing	Multiple masses in the spleen with possible ringlike appearance due to central clearing	*Pneumocystis carinii* infection
Mass with internal calcifications		
Coarse calcifications	Solitary hypoattenuating soft-tissue mass, splenomegaly; hypoattenuating mass, multifocal metastatic involvement (lungs, liver, bones); hypoattenuating mass, multifocal metastatic involvement (gastrointestinal tract, ovaries); splenomegaly, splenic infarcts, lymphadenopathy	Myelofibrosis; epithelioid hemangioendothelioma; peritoneal metastasis of nonsplenic neoplasm; lymphoma (especially treated)
Punctate calcifications	Progressive centripetal fill-in of contrast, rapidly growing; progressive centripetal fill-in of contrast, slow growing; increased vascularity with rapid contrast enhancement	Pleomorphic sarcoma; hemangioma; angiosarcoma
Diffuse calcifications without associated mass		
Coalescent calcifications	Shrunken spleen, associated with a known thromboembolic cause; shrunken spleen, associated with contrast exposure before the 1950s	Autosplenectomy thorium dioxide (Thorotrast) exposure–related fibrosis
Scattered punctate calcifications	Splenomegaly, ascites, cirrhosis; granulomata in the lungs, < five splenic calcifications; granulomata in the lungs, ≥ five splenic calcifications	Gamna-Gandy bodies; *Mycobacterium tuberculosis*; *Histoplasma capsulatum* infection

ic hilum and hepatomegaly [41]. Solitary or multifocal lesions are less common than miliary or diffuse infiltration, therefore CT and MRI are not sensitive for detecting splenic involvement. However, splenic involvement does not generally affect staging [12, 40].

Primary splenic lymphoma will present with spleen-predominant disease with either diffuse uniform infiltration by masses less than 1 cm or a solitary mass that is ill-defined, hypoattenuating with possible mild enhancement, and invading the splenic capsule [2, 41, 43]. The mass may additionally have central necrosis and invasive features with infiltration of the splenic capsule and neighboring visceral organs [2, 41, 43, 45].

Epithelioid Hemangioendothelioma

Epithelioid hemangioendothelioma is a rare connective tissue neoplasm that can arise from various tissues, including liver, bones, skin, soft tissues, lungs, and spleen. It is more common in young adults, and splenic epithelioid hemangioendothelioma can present as a left upper quadrant abdominal mass or pain [6, 9]. It is a malignant vascular tumor that is usually multifocal with metastatic involvement of multiple other visceral organs and connective tissues [46]. It has intermediate malignant potential, with both vascular and stromal components [6, 40].

In the bones, epithelioid hemangioendothelioma presents as mixed sclerotic-lytic lesions, whereas in the visceral organs it grows as coalescing, often calcified masses and then metastasizes to involve other organs [29, 46]. Multifocal lesions in the liver may appear as hypoattenuating lesions with rim enhancement and associated capsular retraction [46].

Imaging of the spleen may show a nonencapsulated yet well-defined solitary mass with hypoattenuating areas of necrosis, enhancing areas of hemorrhage, and possible calcifications without capsular retraction (Fig. 7). Extrasplenic findings may include hepatic epithelioid hemangioendothelioma with characteristic capsular retraction [6, 40]. Lesions will be hyperintense on T1- and T2-weighted imaging because of hemosiderin [2].

Management of splenic hemangioendotheliomas involves surgical resection with partial splenectomy when possible, or total splenectomy if necessary [47].

Splenic Pleomorphic Sarcoma

Undifferentiated pleomorphic sarcoma is one of the most common sarcomas, especially at sites of prior irradiation [48]. However, these lesions are rare in the spleen, with only 20 such cases reported in the literature. These lesions most commonly occur in men 20–40 years old. The tumor is aggressive, commonly metastasizes to the liver, and has a high rate of recurrence. Histopathologic findings show polymorphic, multinucleated, poorly differentiated cells with areas of necrosis [48].

On CT, pleomorphic sarcomas are round, hypoattenuating soft-tissue lesions that closely resemble hemangiomas, but show rapid growth. They have only minimal internal vascularity and can have peripheral serpiginous enhancement. Because of osseous or chondroid metaplasia in 16% of cases, these lesions can develop punctate or coarse calcifications [48, 49]. Hemoperitoneum has been reported as a possible complication [48]. The management strategy for this aggressive splenic malignancy includes surgical removal and chemotherapy [48].

Splenic Angiosarcoma

Angiosarcoma is the most common primary malignant splenic solid tumor, but it is very rare overall. It presents in adults 40–79 years old and is not related to chemical exposures (unlike hepatic angiosarcoma) [12, 16, 40, 50]. This malignancy arises from highly mitotic splenic sinus endothelial cells along disorganized, anastomosing vascular channels [6, 50]. There is a high 1-year mortality rate associated with this malignancy [51].

Imaging may show a solitary mass, multiple splenic masses, or diffuse splenic involvement with splenomegaly. Lesions are heterogeneously hypoattenuating with hyperenhancement in areas of necrosis and areas of hemosiderin deposition with calcification on imaging [6, 40, 50, 52] (Fig. 8). Some lesions may have coarse central calcifications in a radial pattern as a result of underlying fibrosis [52]. Additional findings may include hepatic or bone metastatic lesions and splenic rupture in 30% of patients [12, 51]. Metastatic disease occurs in 69–100% of patients with hypervascular masses in the liver, lung, and lymph nodes [50].

The main consideration for differential diagnosis is a hemangioma, which is best ruled out in favor of angiosarcoma with evidence of metastatic disease [52]. The management strategy for angiosarcoma is splenectomy and chemotherapy after diagnosis is confirmed with fine-needle aspiration biopsy. Chemotherapy is required because of the high proportion of cases with metastatic disease, and splenectomy is also required due to the high risk of rupture [50].

Myelofibrosis

Myelofibrosis is one of multiple myeloproliferative neoplasms that can transform into leukemia, with approximately 14% of primary myelofibrosis cases undergoing leukemic transformation [53]. Other myeloproliferative neoplasms include polycythemia vera and essential thrombocythemia. Approximately 50–60% of patients with these neoplasms have gain-of-function mutations in the Janus kinase 2 gene (*JAK2*) [53, 54].

Two patients with myelofibrosis at MD Anderson Cancer Center had hypoattenuating solitary splenic masses with coarse internal calcifications with concurrent splenomegaly, with no other cases of splenic masses in myelofibrosis described in the literature (Fig. 9). The development of a splenic mass represents a dysregulated tumor microenvironment within the spleen as a result of extramedullary hematopoiesis and increased endothelial cell proliferation [55]. Initially, ineffective hematopoiesis occurs as a result of bone marrow fibrosis and resultant endothelial-to-mesenchymal transition of greater than 30% of endothelial cells in the bone marrow microvasculature. Eventually, this phenotype distribution also occurs in the myelofibrotic spleen, leading to splenic fibrosis and then ineffective hematopoiesis involving megakaryocytes, myeloid cells, fibroblasts, and endothelial cells [54]. This may explain an underlying mechanism for splenic masses in patients with myelofibrosis. However, the pathophysiologic features for splenic lesions in myelofibrosis are not well elucidated in the current literature, much less the presence of calcifications in splenic lesions. Splenomegaly is known to be common.

Treatment of myelofibrosis involves allogeneic hematopoietic stem cell transplant, which is effective in approximately 50% of patients [53].

Splenic Artery Aneurysm

The true prevalence of splenic artery aneurysm is thought to be as low as 0.2% or as high as 10.4%. Among splanchnic artery aneurysms, they are the most common at around 60% [56]. Causes may include atherosclerosis, portal hypertension, trauma, or pancreatitis [39, 57]. Up to 20% of patients may present with left upper quadrant discomfort symptoms, but otherwise report no symptoms [57].

On CT, aneurysms are well-defined enhancing lesions that can have mural thrombus and peripheral calcifications [58]. Many splenic artery aneurysms may contain thromboses and calcifications, with 24% found to have peripheral wall calcifications in a study done by Therakathu et al. [57] (Fig. 10). Aneurysms are usually less than 2 cm in diameter [56].

The differential diagnosis for a splenic artery aneurysm includes an enhancing pancreatic mass or a tortuous vessel. Angiography is the diagnostic method reference standard; however, it is invasive [58]. On angiography, half of aneurysms are fusiform and half are saccular, with greater incidence of pancreatitis when saccular [58]. Splenic artery aneurysms are related with a risk of rupture in 2% of cases [56].

Conclusion

Incidental splenic calcifications have a broad differential diagnosis; however, they can be readily categorized into a clinically useful shortened list according to the imaging patterns and clinical context. This article proposes an algorithmic approach to diagnosing splenic lesions with calcifications (Table 1) that should be used in conjunction with the patient's history and other imaging findings. In general, splenic lesions are managed conservatively even when the patient is showing symptoms of disease. Additional treatment is best determined after diagnosing the underlying abnormality.

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