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Sclerosing Angiomatoid Nodular Transformation of the Spleen

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Background	A 26-year-old female presented with two weeks of left upper quadrant abdominal pain; a splenic mass was discovered on computed tomography (CT) and magnetic resonance imaging (MRI).
Summary	A 26-year-old female presented to our emergency department with two weeks of left upper quadrant abdominal pain. Abdominal CT scan demonstrated a 3.8 cm splenic lesion. Subsequent MRI revealed a well-circumscribed hypointense splenic lesion with indeterminate features favoring the diagnosis of a hemangioma. Laparoscopic splenectomy was performed, and morcellation of the spleen was avoided to prevent the inadvertent spread of a potential malignant lesion. We extended the umbilical port site incision and removed the intact spleen in a large laparoscopic specimen bag. Pathologic examination established the diagnosis of sclerosing angiomatoid nodular transformation (SANT) of the spleen, a benign lesion. The patient reported resolution of her pain at her four-week follow-up.
Conclusion	SANT of the spleen is a rare vascular lesion that can present with vague abdominal discomfort, with definitive diagnosis requiring histologic examination. We present a case of SANT of the spleen that we successfully treated with laparoscopic splenectomy and removed intact by extending a port site into a small laparotomy incision.
Keywords	Sclerosing angiomatoid nodular transformation, splenectomy, laparoscopic

DISCLOSURE:
The authors have no conflicts of interest to disclose.

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Case Description

Sclerosing angiomatoid nodular transformation (SANT) of the spleen is a rare benign lesion that was first characterized by Martel et al in 2004.[1] SANT typically presents asymptomatically or with vague abdominal discomfort or pain, but may also be an incidental finding on imaging. Definitive treatment and pathologic examination is with splenectomy. Here we report a case of SANT that presented with abdominal pain and discuss the clinical presentation, imaging, immunohistochemical profile, treatment, and outcomes.

A 26-year-old Hispanic woman with a history of molar pregnancy presented with two weeks of constant left upper quadrant pain associated with nausea, iron-deficiency anemia (Hg=12.9 g/dL, MCV=76 fL, Ferritin = 4.20 ng/dL) and negative serial pregnancy tests. The patient's initial CT imaging in the emergency department demonstrated a 3.8 x 3.8 x 3.7 cm splenic lesion (Figure 1a). Subsequent MRI of the abdomen demonstrated a 3.7 cm well-circumscribed hypointense splenic lesion with indeterminate features favoring a diagnosis of hemangioma (Figure 1b). Further workup revealed no other potential source of the pain. We opted to remove the spleen laparoscopically. Due to the uncertainty of the diagnosis, the intact spleen was extracted by extending one of the port site incisions. The postoperative course was uncomplicated, and the patient was discharged home on postoperative day four after administration of postsplenectomy vaccinations.

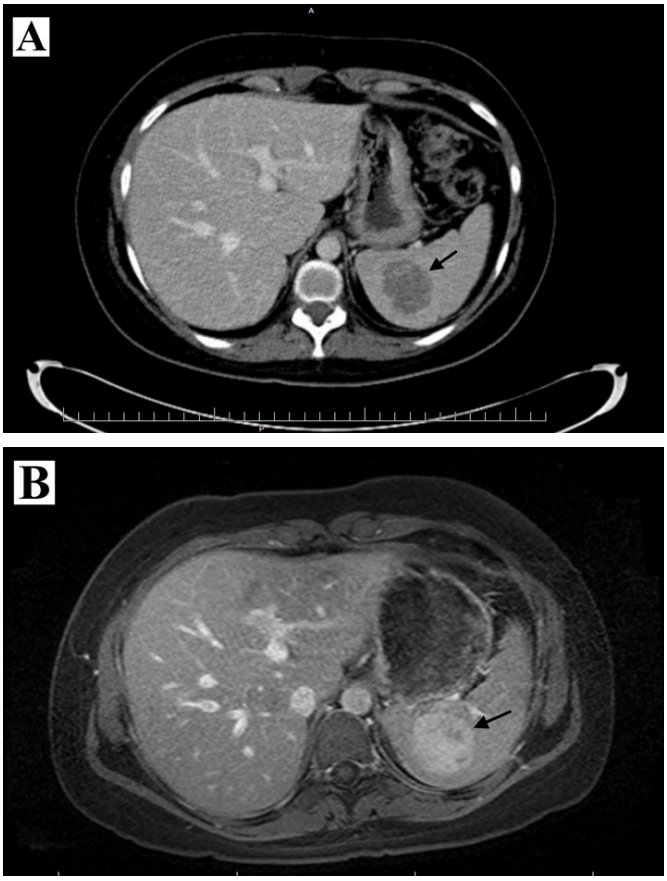

Figure 1. Radiologic findings in SANT. A: Arterial phase of CT with contrast demonstrates a well-circumscribed 3.8 x 3.8 x 3.7 cm lesion with nodular enhancement at the periphery of the lesion. **B:** T1DI MRI demonstrates well-circumscribed hypointense splenic lesion with indeterminate features favoring a diagnosis of hemangioma.

Gross pathological examination of the lesion demonstrated a variegated well-circumscribed lesion with tan-red hemorrhagic areas intermixed with areas of tan-white fibrosis and with no necrotic areas (Figure 2).

The uninvolved spleen displayed a tan-red, unremarkable cut surface. Microscopic examination revealed a benign, vaguely nodular lesion composed of vascularized nodules surrounded by diffusely sclerotic stroma containing a prominent lymphoplasmacytic infiltrate (Figure 3).

Vascular structures were positive for both CD31 and CD34 and negative for CD8. Of note, no evidence of Epstein-Barr virus (EBV) infection was seen on EBV-encoded ribonucleic acid (EBER) in-situ hybridization, and no increased IgG4+ plasma cells were identified on IgG and IgG4 immunohistochemistry. Collectively, these findings are compatible with the diagnosis of SANT.

At her four-week follow-up appointment, the patient denied ongoing symptoms of pain. She reported occasional nausea for which resolved after an additional four weeks.

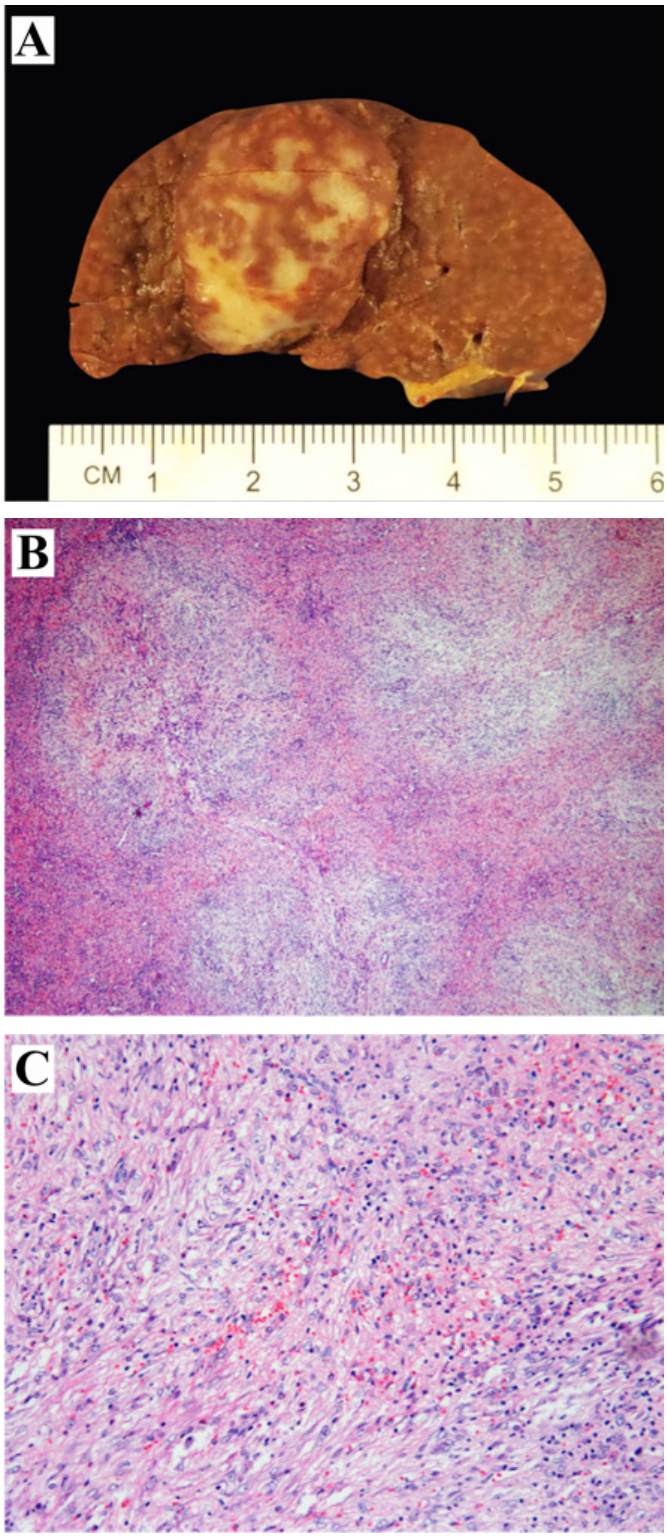

Figure 2. Pathologic findings in SANT. A: Gross examination reveals a well circumscribed, bulging mass with a variegated cut surface comprised of alternating red, hemorrhagic areas and tan white, fibrous areas. **B:** Hematoxylin and eosin stain—microscopic examination reveals a nodular architecture, with fibrous bands delineating paler nodules. **C:** Hematoxylin and eosin stain—on high power, many capillary-like vascular spaces lined by plump endothelial cells are appreciated in a background of fibroblastic stroma with abundant lymphoplasmacytic infiltrate.

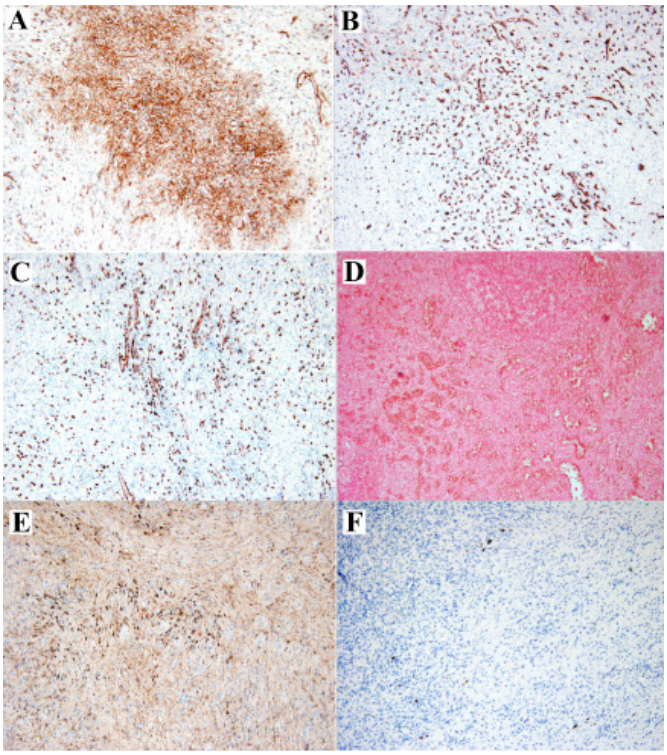

Figure 3. Ancillary testing. A–C: Immunohistochemistry, peroxidase. Immunohistochemical stain reveals that the capillary-like vascular spaces are positive for CD31 (A), CD34 (B) and CD8 (C), a profile consistent with splenic vasculature. **(D): In-situ hybridization (ISH).** No Epstein-Barr virus-positive cells were identified via EBER-ISH. (**E–F) Immunohistochemistry, peroxidase**. A significant number of IgG positive plasma cells was seen (E), but only rare IgG4 positive cells were identified (F) estimated to represent less than five percent of the total IgG positive plasma cells.

Discussion

SANT is a benign, nodular, vascular proliferation of splenic red pulp that is often associated with sclerosis. Its exact etiology is not fully understood; however, one theory is that SANT is the end-stage of a variety of benign splenic conditions, including inflammatory pseudotumor, hamartoma, and hematoma.[2] Although SANT was originally cited to occur more often in females, this preponderance has declined as more reports are published.[3]

The radiographic features of SANT have been previously described on CT and MRI, which closely correlates with our findings.[4, 5] On CT, the lesion appears as a hypodense mass with heterogeneous enhancement. It has a well-demarcated border and nodular peripheral enhancement suggestive of a hypervascular rim and is without calcifications. On T1WI contrast-enhancing MRI, the lesion shows centripetal and progressive enhancement without signs of cys-

tic changes or necrosis. These radiologic findings may be suggestive of benign vascular lesions such as hemangiomas; however, they can also be malignant hemangiomas such as angiosarcomas. Therefore, it is important to definitively characterize these lesions by histopathology.[6]

Most reported cases of SANT are diagnosed after splenectomy following an incidental radiographic finding; however, there is one report of diagnosis by ultrasound-guided percutaneous core needle biopsy.[7] To date, there have been no reported asymptomatic cases or instances where SANT has been observed without surgical intervention. This may be, in part, due to the requirement of histology for the definitive diagnosis.

On gross examination, SANT is usually a solitary, well-circumscribed nodule that is distinct from the surrounding splenic parenchyma. Histologically, it has a vaguely lobular architecture surrounded by a hyaline shell, with markedly cellular blood vessels similar to those seen in hemangiomas.[8] Its stroma consists of myxoid and sclerotic fibrous tissue with some myofibroblasts, lymphoplastmacytic cells, and hemosiderin-containing macrophages. Immunohistochemical analysis has suggested SANT to be a predominantly polyclonal reactive lesion, lacking evidence of any relationship with IgG4-related sclerosing lesions, or EBV-positive stromal cells.[9] SANT has traditionally demonstrated three distinct patterns of immune staining: CD34+/CD31+/CD8-, CD34-/CD31+/CD8+ , and CD34-/CD31+/CD8-, which indicates derivation from capillaries, splenic sinusoidal lining cells, or small veins, respectively.[10] Early lesions have also demonstrated perivascular concentric fibrosis, as was seen in this case.[6]

Splenectomy is the treatment of choice for SANT, and no instance of recurrence has been reported. It is important, however, to note that the initial presentation can be somewhat ambiguous. In our case, we opted for a laparoscopic splenectomy to minimize length of stay and to shorten the postoperative recovery time. Morcellation to extract the spleen was avoided due to the uncertain diagnosis of the lesion and to prevent inadvertent spread of a potential malignancy. We extracted the intact spleen in a large laparoscopic bag by extending the umbilical port. This may be of special pertinence given that there have been a few reported instances of multifocal lesions involving anywhere from two to eight nodules, as these presentations may further enhance a clinician's suspicion of a malignant etiology.[3]

Lastly, it is important to ensure proper vaccination to prevent overwhelming postsplenectomy sepsis secondary to encapsulated bacteria. The timing of vaccinations has been the topic of a longstanding debate, and there is no Class I data identifying the appropriate timing for presplenectomy. It is generally accepted that patients undergoing elective splenectomy should be vaccinated two weeks prior to the procedure; this guideline is supported by Class III data.[11] Those undergoing emergent or urgent splenectomy should receive vaccinations prior to discharge.

Conclusion

SANT is a rare splenic lesion and cause of left upper quadrant pain. The surgical approach should consider the possibility of malignancy, and morcellation of the spleen should be avoided. No recurrent lesions have been reported after surgical removal.

Lessons Learned

Sclerosing angiomatoid nodular transformation of the spleen is a rare, benign vascular growth the can present with vague symptoms. Imaging suggests vascular involvement, but diagnosis requires histologic evaluation. Here we report a case managed by laparoscopic splenectomy with port site extension for intact removal of the spleen.

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