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## Case Report

# Sclerosing angiomatoid nodular transformation of the spleen: A case report of thrombocytopenia and a hypervascular splenic mass

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## ABSTRACT

**Introduction:** Sclerosing Angiomatoid Nodular Transformation of the spleen is a benign vascular lesion with no known etiology.

**Presentation of Case:** We report a new case in a symptomatic twenty-one-year old female with thrombocytopenia and a hypervascular splenic mass discovered on ultrasound. Two MRIs were performed prior to hand-assisted laparoscopic splenectomy. The specimen was sent for histopathologic analysis with confirmation of final diagnosis from an outside facility.

**Discussion:** Sclerosing Angiomatoid Nodular Transformation of the spleen is most often discovered incidentally as a solitary splenic mass. The presence of a spoke-wheel pattern should alert the radiologist to this as a possibility.

**Conclusion:** Ultrasound and MR imaging findings can be used to accurately diagnose cases of splenic Sclerosing Angiomatoid Nodular Transformation. Susceptibility artifact within the lesion may be directly related to the amount of iron deposition.

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## Case report

A 21-year-old female presented to her primary care physician for renewal of oral contraceptive medication. At this visit the patient reported a 6-month history of daily headaches and fatigue. She also disclosed that she had previously attempted to

donate blood and was not allowed to do so because she found to be anemic. CBC with differential revealed a platelet count of 115,000/mcL with no other remarkable abnormalities. She was referred to hematology/oncology for workup of thrombocytopenia.

The initial hematology evaluation revealed a platelet count of 135,000/mcL. Additional labs were performed but revealed

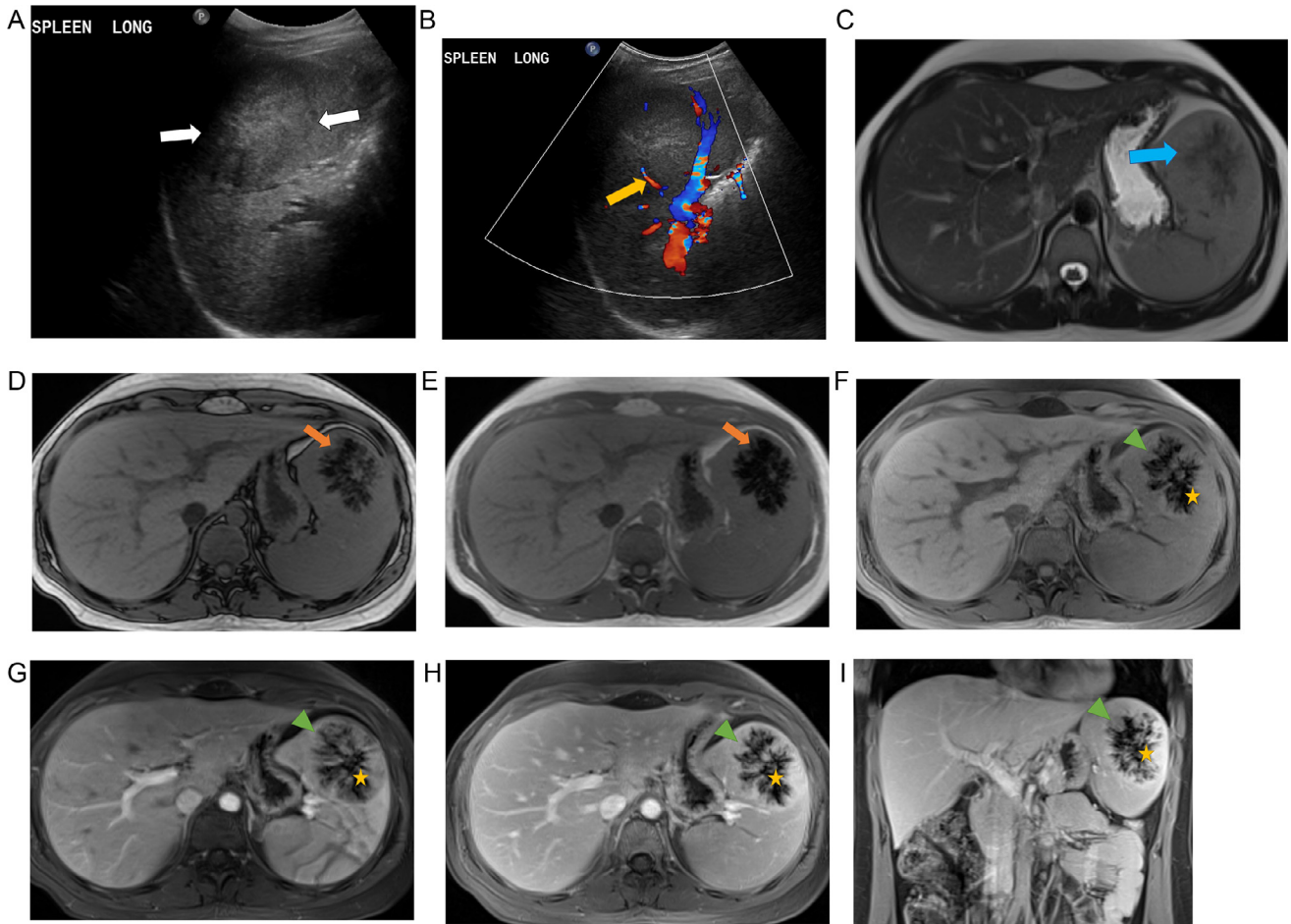
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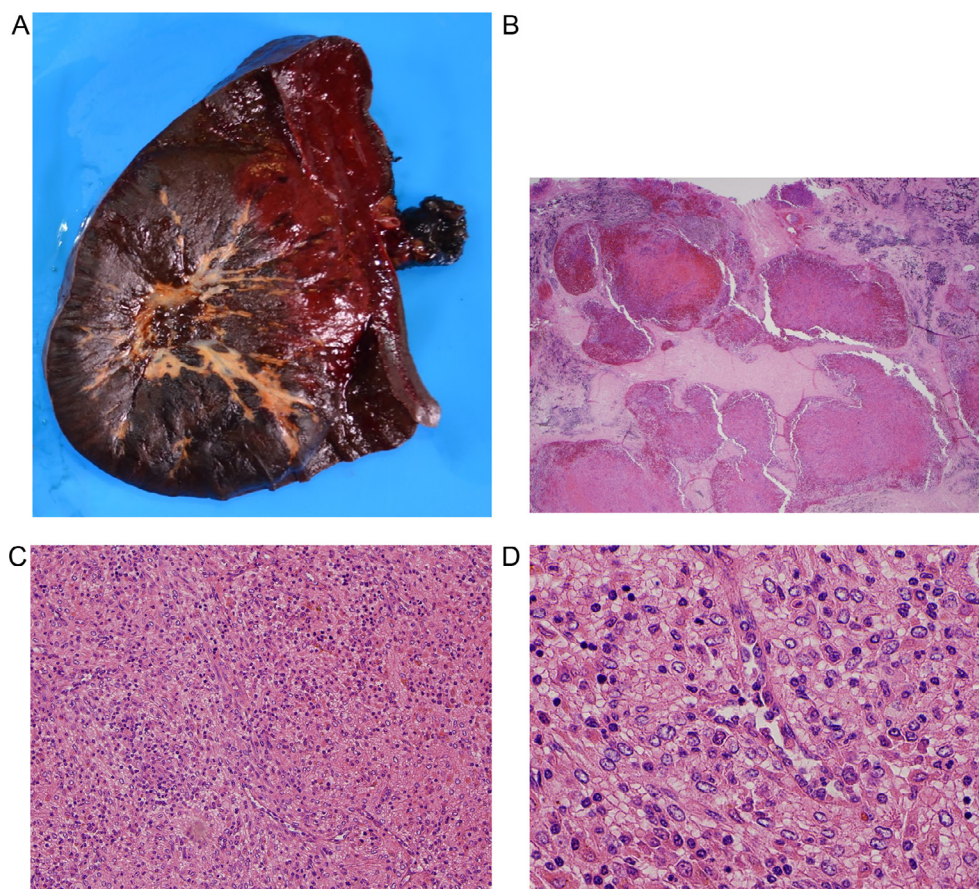
**Fig. 1 – Ultrasound and MR imaging findings of Sclerosing Angiomatoid Nodular Transformation of the spleen in a twenty-one-year-old female with thrombocytopenia.**

- A.** Long axis grey scale ultrasound image of the spleen showing a well-defined mildly hyperechoic splenic lesion (white arrows).
- B.** Long axis Doppler ultrasound image with color flow detected within the splenic lesion (yellow arrow).
- C.** Axial T2 weighted MR image without fat suppression of the splenic lesion showing peripheral isointensity and central hypointensity (blue arrow).
- D.** Axial T1 opposed-phase (OP) image showing peripheral isointense and central hypointense signal of the splenic lesion (orange arrow).
- E.** Axial T1 in-phase (IP) image showing increased central hypointensity of the splenic lesion as compared to the OP image (orange arrow).
- F.** Axial T1 fat-saturated MR image which demonstrates splenic lesion with peripheral isointensity (green arrow head) and central hypointensity (orange star).
- G.** Axial post-contrast T1 fat-saturated MR image in arterial phase demonstrating peripheral nodular enhancement of the lesion (arrow head) and lack of central filling (star).
- H.** Axial post-contrast T1 fat-saturated MR image in venous phase showing increasing peripheral enhancement (arrow head) and persistent lack of central filling (star).
- I.** Coronal post-contrast T1 fat-saturated MR image in the delayed phase showing peripheral enhancement (arrow head) and unchanged lack of central filling (star).

no significant abnormalities. Follow-up 1 month later revealed a platelet count of 115,000/mcL. The patient reported that between visits she had consumed 1-2 alcoholic beverages per week. She was instructed to refrain from alcohol consumption and to initiate an exercise routine. Serum B12 level was found to be below 400 so supplementation was started. At a 3-month

follow-up appointment her platelet count was 108,000/mcL despite B12 supplementation and minimal alcohol consumption.

An ultrasound of the spleen was performed and revealed a heterogeneous 7.1 × 5.7 cm solid mass with color Doppler flow within the mid pole of the spleen (Fig. 1 A-B). Diagnos-



**Fig. 2 – Pathologic findings of Sclerosing Angiomatoid Nodular Transformation in a twenty-one-year old female with thrombocytopenia.**

**A.** Gross macroscopic pathology picture of the spleen showing a single well-circumscribed mass with central radiating scar.

**B.** Microscopic picture of the splenic lesion (H&E stain, 2x) showing multiple angiomatoid nodules surrounded by thick dense fibrous bands.

**C.** Microscopic picture of the splenic lesion (H&E stain, 20x) showing angiomatous nodules composed of numerous blood vessels surrounded by collagenous stroma.

**D.** Microscopic picture of the splenic lesion (H&E stain, 60x) showing vessels lined by plump endothelial cells with large oval nuclei, vesicular chromatin and prominent nucleoli.

tic considerations included hemangioma, hamartoma, lymphoma, or metastasis and an MRI of the abdomen was suggested to better characterize the lesion.

MRI of the abdomen performed one week later demonstrated a rounded well-defined  $5.9 \times 7.8 \times 8.1$  cm anterior subcapsular mass present within the spleen. The mass demonstrated predominantly peripheral isointense T1 signal (Fig. 1F), minimally increased T2 signal (Fig. 1C), and a central stellate hypointense appearance on T1 and T2 sequences (Fig. 1 C-F). The lesion shows obvious drop of signal on the in-phase images (Fig. 1E) as compared to the opposed-phase images (Fig. 1D) reflecting internal susceptibility artifact suggestive of internal iron deposition. Postcontrast imaging of the mass demonstrated peripheral nodular enhancement which mildly increased on delayed phase imaging (Fig. 1 G-I). The central stellate portion of the mass had no significant enhancement (Fig. 1 G-I). No significant diffusion restriction was seen within the mass.

#### Figure 2.

Follow-up MRI of the abdomen performed 4 months later demonstrated a slight decrease in size ( $5.3 \times 7.2 \times 7.5$  cm) of the mass. The central stellate portion demonstrated more prominent T1 and T2 hypointensity with similar peripheral nodular enhancement on postcontrast delayed phase imaging. Both diffusion weighted imaging and apparent diffusion coefficient mapping demonstrated areas of hypointensity consistent with new internal susceptibility artifact suggestive of iron deposition.

The platelet count at that time was 96,000/mcL. She was given prophylactic vaccinations for encapsulated organisms and referred to Surgical Oncology for splenectomy evaluation.

Surgical Oncology agreed that the splenic mass was the most likely cause of her thrombocytopenia and she was scheduled for splenectomy. Laparoscopic hand-assisted splenectomy was performed approximately 11 months after initial presentation.

Pathologic analysis reported a 445 g spleen measuring  $14.5 \times 10 \times 7.5$  cm with a  $7.5 \times 7 \times 5.5$  cm well-circumscribed nonencapsulated dark red mass with tan-yellow to tan-white fibrous tissue expanding radially from the center of the lesion. Histopathologic and immunohistochemical evaluation of the splenic mass revealed the mass was composed of thick dense fibrous bands which coalesced to form angiomatous nodules composed of a fine network of numerous blood vessels surrounded by collagenous stroma. An increased number of plasma cells, rare small lymphocytes, rare neutrophils, and no eosinophils were seen within the vascular nodules. A population of bland spindle and/or plump endothelial cells with large oval nuclei, vesicular chromatin, and prominent nucleoli was noted. The dense fibrous bands contained numerous hemosiderin-laden macrophages and focal areas of microcalcifications. Numerous arteries of variable size with thickened hyalinized walls. These vessels stained positive for CD31, CD8, CD34, Factor VIII, and rare focal D240. CD68 highlighted an increased number of Littoral cells/histiocytes. The angiomatous nodules contained cells positive for CD31, CD8, CD34, vimentin, SMA, focal positive for EMA, rare positive S100, and negative for desmin. Approximately 10% positivity for Ki-67. Additional CD21 stains highlighted rare positive cells.

The histopathologic findings were consistent with sclerosing angiomatoid nodular transformation. Consultation from MD Anderson Cancer Center Department of Pathology was congruent with reported diagnosis. Notably, it was reported that this case had more hemorrhage, more calcium deposition, and more hemosiderin deposition than other cases they had previously evaluated.

The patient did well following surgery and was released by surgical oncology June 15, 2016. At her 6-week postoperative follow-up with hematology/oncology the patient reported feeling well and was anticipating returning to work. CBC with differential performed at this visit revealed a platelet count of 334,000/mcL and she was scheduled to return in 3 months for continued monitoring.

## Discussion

Sclerosing Angiomatoid Nodular Transformation (SANT) is a benign vascular lesion of the spleen initially described for its characteristic morphologic features and distinctive immunohistochemistry profile by Martel et al. in 2004 [1]. Since this initial description, fewer than 200 cases have been reported. In 2015 Cao et al. reviewed five patients with multifocal lesions [2], while other reports described a predominantly solitary lesion discovered incidentally [3]. Patients are commonly asymptomatic, but some present symptomatically, with nonspecific abdominal pain being the most common. Other presenting characteristics include, but are not limited to, cytopenias, flank pain, pelvic pain, and constitutional symptoms [4].

Hypervascular lesions of the spleen include hemangiomas, hamartomas, SANT, Littoral cell angiomatosis, metastases, lymphomas, angiosarcomas, malignant fibrous histiocytomas, and pleomorphic undifferentiated sarcomas. These lesions

can be difficult to diagnose and often require multiple imaging modalities in order to provide a confident differential diagnosis. Bowerson et al. recently reported the imaging characteristics, clinical features, and presentation of these hypervascular splenic lesions to better differentiate the various malignant and benign lesions [5]. Here we will focus on the ultrasound, computed tomography, and magnetic resonance imaging characteristics of SANT.

Ultrasound findings are nonspecific, commonly reported as a heterogeneous hypo- or hyperechogenic mass with Doppler detectable blood flow [6]. It has been suggested that the use of second-generation contrast enhanced ultrasound may provide additional information that can assist in visualizing the vessel structure and morphologic characteristics of SANT [7]. The ultrasound findings in this case are consistent with these reports, demonstrating a heterogeneous mass with Doppler blood flow. Contrast enhancement was not used in this case.

The first report of CT findings in SANT described a hypodense mass in early portal venous phase imaging which became homogenous postcontrast and indistinguishable from normal spleen on late portal venous phase images [8]. Others have reported findings of peripheral nodular enhancement on arterial phase imaging, hypervascular rim during portal venous phase imaging, and progressive enhancement toward isodensity on delayed imaging [9–13]. The radial scarring associated with the gross pathology may be visible on CT, but it is most prominent on MRI [14]. Our patient did not undergo CT scanning as it was determined MRI would be more appropriate for characterization given her age, concerns for radiation exposure, and prior ultrasound findings.

The dynamic MRI findings in a confirmed case of SANT were previously described as a spoke-wheel pattern by Karaosmanoglu et al. [14]. This pattern consisted of central hyperintensity on precontrast T1 fat-saturated images. T2 images demonstrated peripheral hyperintensity and central hypointensity. There were also areas of T2 hyperintensity radiating from the periphery to the center of the lesion which gives the characteristic spoke wheel appearance to the lesion. Early arterial phase T1 gradient echo images showed peripheral enhancement which progressed centrally but lacked central filling on late phase imaging.

Kim et al. proposed that decreased signal on longer gradient echo images may be secondary to old hemorrhage and hemosiderin deposition within the lesion. Hypointensity in both diffusion weighted images and apparent diffusion coefficient mapping was noted in one of their patients, which they suggested was due to susceptibility artifact from iron deposition [3]. The radiologic and histopathologic findings in our patient are concordant with this theory.

These findings correlate well with the pattern of fibrosis commonly seen on gross pathologic examination [1]. Subsequent studies have also reported the presence of this spoke-wheel pattern which may serve as a diagnostic clue for SANT although reports have been made where this pattern was not visible [15]. The images obtained in this case demonstrate the spoke-wheel pattern with remarkable clarity. This is likely due to the increased amount of hemosiderin deposition that was found within the mass. These findings support the suggestion that this pattern should heighten the interpreter's suspicion of this rare lesion.

In addition to the conventional MRI sequence findings described above, this case notably demonstrated no significant diffusion restriction within the lesion. Jang et al. demonstrated that the addition of diffusion weighted imaging to conventional MRI improved diagnostic confidence of both malignant and benign focal splenic lesions [16]. The interpreting physicians in our case were able to accurately diagnose this rare lesion prior to histopathologic confirmation based on these imaging characteristics.

## Conclusion

In summary, we reported a case of a symptomatic female with histopathologically confirmed SANT diagnosed accurately using ultrasound and MRI. The patient subsequently underwent hand-assisted laparoscopic splenectomy with resolution of symptoms soon after. In females of child-bearing age, MRI, and ultrasound should be considered first-line in the evaluation of splenic lesions. The presence of a spoke-wheel pattern on either study should alert the radiologist to the possibility of SANT.

## Teaching points

Opposed-phase and in-phase MR images demonstrating susceptibility artifact within the lesion may be directly related to the amount of iron deposition. Variations in the amount of iron deposition among lesions may explain why the spoke-wheel appearance is not always visualized.

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## Consent

No.

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