

# An extremely rare case of symptomatic multifocal Sclerosing Angiomatoid

## Nodular Transformation of Spleen (SANT) Suburban Community Hospital Extraordinary People. Extraordinary Care.

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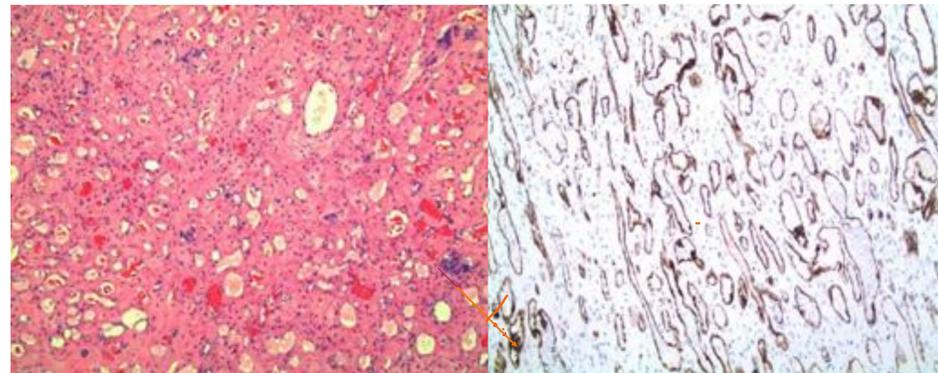
### Background

- Sclerosing Angiomatoid Nodular Transformation (SANT) is an extremely rare benign vascular lesion of the spleen with unclear etiology which can mimic a malignant splenic neoplasm on imaging.
- Most cases were detected incidentally as they are mostly asymptomatic and rarely symptomatic.
- Diagnosis confirmed with histopathology and treatment is splenectomy

### Case Details

- A 62-year-old African-American male presented with left upper abdominal pain for 2 days and weight loss of 40 pounds over 6 months.
- Physical exam: Non tender Grade 3 Splenomegaly on palpation
- CT Abdomen: Splenomegaly with multiple heterogeneously hypodense masses, suspicious for splenic neoplasm.
- Abnormal laboratory values:
  - Leukocyte count :4,400 cells/ $\mu$ L with neutrophilic predominance.
  - Hemoglobin :13.1 g/dL
  - Low platelets :29,000 cells/ $\mu$ L,
  - Fibrinogen : 537 mg/dL
  - Peripheral smear : Pancytopenia.
- Pre-operative Histopathology : Ultrasound-guided core biopsy of the spleen revealed features most consistent with a vascular neoplasm, favoring littoral cell angiodysplasia.
- Treatment :
  - Platelet transfusions to maintain the platelet count above 100,000/ $\mu$ L for surgery.
  - Splenectomy was performed without complications.
  - Postoperatively : Symptoms and thrombocytopenia resolved.
- Histopathology of the surgical specimen: Vascular derived lesion consistent with sclerosing angiomatoid nodular transformation(SANT)

### Imaging & Histopathology



### Discussion

- To date, less than 200 cases of SANT of the spleen have been reported.
- Most cases were diagnosed incidentally in asymptomatic patients based on incidental findings on imaging.
- For symptomatic patients, abdominal pain or discomfort was the predominant symptom; other symptoms were palpable mass, flank pain, pelvic pain, anemia, long-standing low-grade fevers, and night sweats.
- SANT usually presents as a solitary lesion but can be multifocal in rare cases such as this case.
- Radiological imaging cannot differentiate SANT from other malignant tumors.
- Definitive diagnosis is contingent on histopathological and immunohistochemical examination of the lesion. Immunohistochemistry shows vessels of spleen positive for CD34/CD31/CD8 markers
- The gold standard treatment is splenectomy.

### Conclusion

- SANT is an extremely rare benign vascular lesion of the spleen and this case is even rare because of multifocal lesions and symptomatic presentation
- A definitive diagnosis can be missed on Ultrasound guided core needle biopsy and it can be established only with surgical histopathology and immunohistochemistry.
- Despite the established benign nature of the tumor, the fact that this patient had significant unexplained weight loss (a concerning feature for malignancy), we conclude that further detailed insight is warranted to understand the Benign Vs Malignant nature of the SANT of spleen.

### References

- 1) <https://www.pathologyoutlines.com/topic/spleensclerosingangiomas.html>
- 2) <https://www.ncbi.nlm.nih.gov/pmc/articles/PMC7809664/#>
- 3) <https://www.ncbi.nlm.nih.gov/pmc/articles/PMC5607801/>