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# Sarcoidosis Presenting with Massive Splenomegaly and Severe Epistaxis

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10/25/2019

# Introduction

- Sarcoidosis is an idiopathic multisystem disorder characterized by formation of non-caseating granulomas
- >90% of cases isolated to the lungs
- Extrapulmonary involvement:
  - Skin, lymph nodes, eyes, heart, GI tract, liver, and spleen

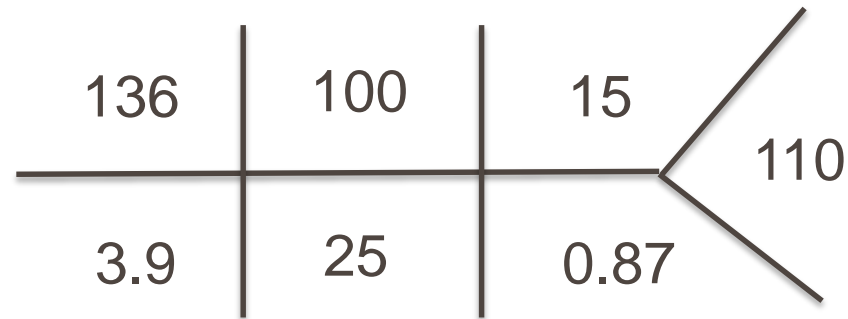
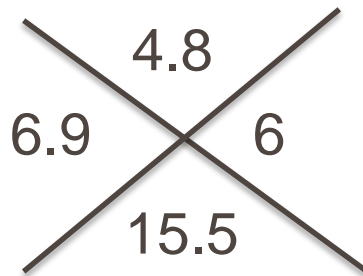
# History

- 58 yo AAF presented from urgent care with hemoglobin of 4.8. Two days prior to admission she had spontaneous profuse nosebleed that lasted 6 hours. Associated symptoms: palpitations, dizziness, nausea, vomiting, fatigue.
  - PMH: menorrhagia, uterine fibroids, LLE DVT x2 (1<sup>st</sup> unprovoked, 2<sup>nd</sup> traumatic, tx with Warfarin x 6 mos), iron deficiency anemia, first trimester miscarriage, epistaxis requiring cauterization
  - PSH: C-section, bilateral tubal ligation
  - FHx: Sister with bleeding and clotting disorder
  - Social: Never smoker, denies EtOH, smokes marijuana daily, married with 2 children
  - Medications: Asprin 81mg daily
  - Allergies: None

# Physical Exam

- Vitals: T 97.8, P 129, R 17, BP 140/65, SpO2 100% (room air)
- General appearance: alert, awake, oriented, no acute distress
- Head/Eyes: atraumatic, clear cornea, EOMI, **pale conjunctiva**
- ENT: moist mucosal membranes, **no blood visualized in nares**
- Neck: non-tender, no lymphadenopathy
- Cardiovascular: **tachycardia**, normal heart sounds, BP/pulses equal bilat.
- Respiratory: clear to auscultation, no distress, no tenderness
- Abdomen/GI: **splenomegaly**, active bowel sounds, soft, non-tender, no distention
- Extremities: no edema-all extremities, no evidence of DVT
- Musculoskeletal: normal inspection, no CVA tenderness
- Neuro/CNS: alert, oriented X 3, normal speech
- Skin: dry, intact, no gross abnormalities
- Psychiatry: normal affect, normal judgment/insight, normal mood

## Lab Findings



- PT 14.5 H, INR 1.23, aPTT 24.7 L
- Fibrinogen 271 H
- Abs Retic count 0.1730 (0.11H)
- LDH 262 (190 H), Haptoglobin 112

### Peripheral smear:

- Marked anemia with normocytic indices
- Profound thrombocytopenia
- Atypical lymphocytes and increased metamyelocytes and myelocytes and rare NRBC
- No identifiable blast cells
- No schistocytes.

# Investigations

- CT abdomen/pelvis showed massive splenomegaly with spleen measuring 25 cm craniocaudad, 8 cm transverse, and 18.5 cm anterior-posterior but without discrete splenic lesions.
- CT chest was void of hilar adenopathy or pulmonary infiltrates but showed bibasilar scarring fibrosis and bronchiectasis.
- Bone marrow biopsy and flow cytometry was negative for malignancy but iron stains showed complete depletion of storage and sideroblastic iron.

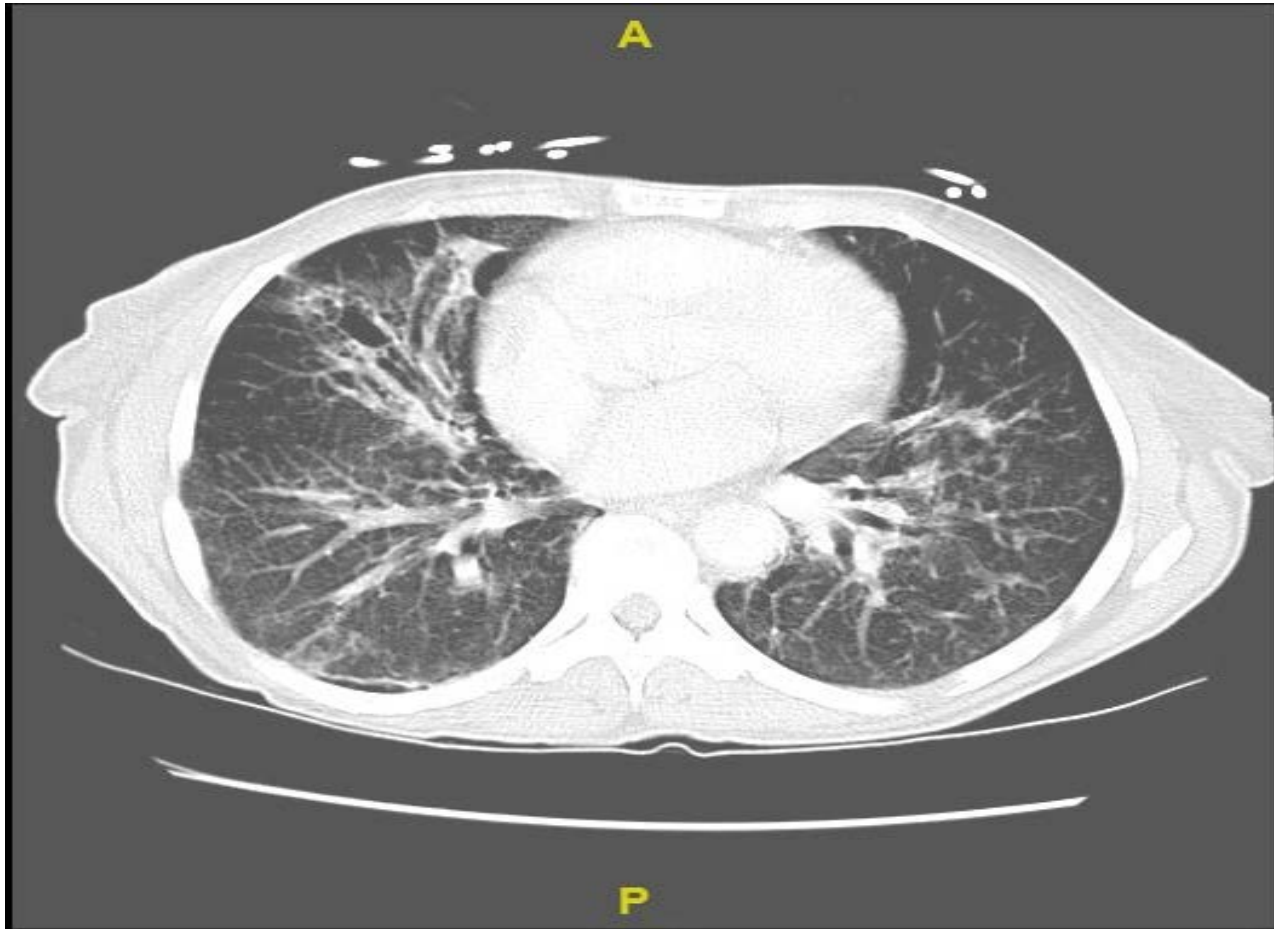
# Imaging



CT abdomen and pelvis with contrast on admission demonstrated marked splenomegaly, spleen measuring 25 cm craniocaudad, the 8 cm transverse, a 18.5 cm AP. Marked enlargement of the splenic veins. Portal vein is patent. The spleen is displacing the left kidney medially.



# Imaging



CT chest with contrast: Stable bibasilar scarring, bronchiectasis, and patchy consolidation compatible with chronic inflammatory changes.

# Hospital Course

- Hospital day #3: severe epistaxis for several hours requiring nasal packing
- She continued to have intermittent persistent nasal bleeding and received multiple platelet transfusions without improvement in her platelet count.
- Open splenectomy was deemed unsafe given the impressive size of the spleen.
- The patient underwent partial splenic artery embolization which revealed multiple splenic artery aneurysms. Within three days the platelet count normalized and epistaxis resolved.
- Partial splenic artery embolization allowed diagnostic splenectomy and splenic artery repair to be safely performed.
- Surgical pathology demonstrated extensive non-caseating granulomas consistent with sarcoidosis.

# More Imaging



- Gross specimen of spleen after open splenectomy

# Discussion

- The liver and spleen are the primary organs involved in abdominal manifestations of sarcoidosis. In the spleen, massive splenomegaly is the most common presentation, followed by multiple splenic lesions.
- This case of massive splenomegaly was confounded by both primary splenic sarcoidosis and multiple splenic artery aneurysms leading to aggressive platelet sequestration.

# Discussion

- Despite consecutive platelet transfusions, platelet counts remained under 10 thousand, and hemostasis was unable to be achieved resulting in life-threatening epistaxis.
- After splenic embolization, splenic artery aneurysm repair, and open splenectomy, platelet count quickly normalized and her epistaxis resolved.

# Conclusion

- In patients presenting with massive splenomegaly and thrombocytopenia, splenic sarcoidosis should be considered in the differential along with other causes of splenic sequestration including infection, autoimmune cytopenias, leukemia, lymphoma, liver disease, vascular obstruction, and extramedullary hematopoiesis.

# Thank You