I’m a Mystery: An Unusual Case of a Neck Mass

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THE CASE

HPI 81 yom presents with 1 day h/o enlarging right-sided neck mass
- Ice application provided mild reduction in size
- Reports no associated sxs, denies fever, sore throat, chest pain, SOB, dysphagia
- Hospitalized at least twice in the previous 6 months in Florida, where he resides, for swelling in his neck, denies h/o intubation
- States “I’m a mystery, no one knows what’s going on with me.”

ROS
- Intentional weight loss - 60 lbs over past 6 months via decreasing meal sizes.
THE CASE

PMH
- T2DM-not on insulin
- Essential HTN
- Paroxysmal atrial fibrillation
- Osteoarthritis
- DVT-first diagnosed in 2010, with recurrent DVTs in the last 6 months
- Basal cell carcinoma of left hand
- Colonoscopy with polyp removal 3-4 years ago

PSH
- Carpal tunnel release
- Femur fracture surgery
- 2 knee replacements
- Bowel resection, etiology unspecified
THE CASE

**Meds**
- Glipizide 5mg
- Coumadin 5mg
- Metoprolol 25mg
- Aspirin 81mg
- Celebrex 200mg
- Ferrous sulfate 325mg
- Niacin 500mg
- Simvastatin 40mg

**FH**
- Mother died of MI
- Father died of MI, h/o lung cancer
- 3 sisters died of cancer, 1 from lymphoma, 2 unspecified

**SH**
- Retired for 24 yrs, lives in Florida, in Wisconsin for the summers
- Quit smoking in 1968
**PE**

- **VS:** T98.4F BP 102/58 P89 RR20 SpO2 100% RA
- **General:** normal-appearing elderly gentleman in NAD
- **HEENT:** oropharynx without erythema, exudate or lesions. Dentures
- **Neck:** normal ROM. **Right-sided non-pulsatile tender neck mass over the SCM muscle.** 6cmx4cm, rubbery, warm, pink, no other lymphadenopathy/masses in head, neck or nodes
- **Cardiovascular:** RRR, normal S1, S2, no murmurs
- **Respiratory:** CTAB. No inspiratory stridor
- **Abdomen:** no HSM
- **Skin and Neuro:** no abnormal findings
IMAGING

CT scan of neck showed findings consistent with

- Myositis
- Cellulitis
- Parotitis
HOSPITAL COURSE

- CBC showed mild normocytic anemia, INR 1.5
- BMP, UA, CPK and CXR normal
- Started on Vanc/Zosyn
- ENT consulted, presentation c/w parotitis
- Discharged on 10 day course of amoxicillin/clavulanate, to f/u with ENT after finishing antibiotic
PATIENT RETURNS

- 3 days later, complains of difficulty swallowing and increased swelling on right side of neck
- Re-admitted from ENT clinic
- O2 sat 90% RA
- PE significant for enlarging right-sided neck mass
- CBC unchanged, INR at 1.2
DIFFERENTIAL DIAGNOSIS

- Congenital
- Inflammatory
  - Skin abscess
  - Bacterial lymphadenitis
  - Reactive viral lymphadenitis
  - Autoimmune vasculitis
  - Focal myositis
  - Fibrosing mediastinitis
- Neoplastic
  - Lymphoma
  - Head and neck metastases from squamous cell cancer
  - Salivary gland
  - Thyroid
  - Benign skin cyst
  - Lipoma
RE-IMAGING
WORKUP

- Abnormal lab results
  - ESR 104 CRP 11.5
  - Positive lupus anticoagulant (while on anticoagulation)
  - Elevated IgA

- Normal lab results
  - Cardiolipin antibody
  - Beta 2 glycoprotein 1 antibody
  - ACE level
  - Aldolase
  - ANA with ELISA, ANA with indirect immunofluorescent assay
  - ANCA
  - Anti-SSA/Ro
  - Anti-Jo 1
  - Lyme titer
  - Homocysteine level
  - Quantitative IgG and IgM
  - Histoplasmosis complement fixation antibody level
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BIOPSY

Dense Fibrosis
(NO SIGNS OF MALIGNANCY)
Fibrosing Mediastinitis

- Disorder characterized by excessive fibrotic reaction in the mediastinum.
- Can result in the compromise of the airways, great vessels and other mediastinal structures
- Symptomology includes cough, chest pain and dyspnea due to compression of vital structures, pulmonary artery obstruction, SVC syndrome or constrictive pericarditis
- Commonly a sequel of histoplasmosis, or can be sequel of tuberculosis or sarcoidosis

Pathophysiology

- Usually results from excessive host response to a prior infection that involves the mediastinal lymph nodes
- Fungal antigens from lymph nodes leak into mediastinum, leading to hypersensitivity and fibrotic response (higher risk with HLA-A2 antigen)
- Divided into two major classes
  - Focal-most cases secondary to Histoplasmosis (82%)
    - Will see calcifications in mediastinum
  - Diffuse-autoimmune/idiopathic (18%)
    - Not associated with calcifications in mediastinum
- Idiopathic type often requires biopsy due to inability to r/o other etiologies such as lymphoma

Treatment

- Glucocorticoids have not been shown to be effective in the focal type, although controlled trials have not been performed
- 1 case report demonstrated steroids halted progression of autoimmune fibrosing mediastinitis

For our patient

- For his neck mass, Pt was given 60mg daily prednisone and his swelling decreased
- Was seen during 1 week f/u and reported no symptoms.
- Was referred to a rheumatologist at Froedert who confirmed idiopathic fibrosing mediastinitis at 3 week f/u

Summary

- Fibrosing mediastinitis can be included on the differential for a neck mass
- Autoimmune fibrosing mediastinitis may benefit from glucocorticoid therapy
References

- Weinberger, SE. Fibrosing mediastinitis. UpToDate. Jul 2013
Cervical manifestations of FM

- 2 cases reported of fibroinflammatory and fibrosclerosing lesions originating in mediastinum which extended superiorly to involve soft tissues of the neck

CT CHEST

- Superior vena cava diameter of 12mm. Right brachiocephalic and right IJV are not visible. Mediastinum has no adenopathy or masses.
- Impression: SVC stenosis with involvement of distal segment of both brachiocephalic veins in the right IJV
MRI NECK

- Extensive inflammatory stranding in the right neck with swelling of SCM with adenopathy
FOCAL MYOSITIS

- Uncommon inflammatory pseudotumor of skeletal muscle
- Cause is unknown, trauma considered
- Review of 115 cases\(^1\) reported location as lower extremities (n=39), sternocleidomastoid (n=8).
- Histology showed variable myopathic (93%) and focal neurogenic changes, fibrosis, and inflammation (97%), occasionally accompanied by prominent eosinophils (n=20)

Auerbach, A et al. Focal Myositis: Clinicopathologic Study of 115 cases of an Intramuscular Mass-Like Reactive Process
DENSE FIBROSIS

- Idiopathic pulmonary fibrosis
- Cirrhosis
- Mediastinal fibrosis
- Myelofibrosis
- Retroperitoneal fibrosis
- Old MI
- Keloid
- Scleroderma