Sero-Negative Granulomatosis with Polyangiitis Masquerading as a Hypermetabolic Left Upper Lobe Lung Mass
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INTRODUCTION
- Granulomatosis with Polyangiitis (GPA) is a small vessel vasculitis that usually affects the upper respiratory tract (URT, nasopharynx), lower respiratory tract (LRT, lungs), and the kidneys [1] (See Image 1).
- Chest X-ray findings commonly show bilateral, lingular opacities with hilar lymphadenopathy; less commonly, the disease can also present as a discrete lung nodule.
- Pathophysiology of GPA has been linked to c-ANCA, an auto-antibody that binds to proteinase-3 within the cytoplasm of neutrophils [2, 3] (See Image 2).
- c-ANCA is both specific and sensitive for GPA, but about 10% of patients with GPA are c-ANCA negative [3].

CASE DESCRIPTION
- HPI: A 54-year-old male fashion designer presented to the hospital with complaints of worsening myalgia, sinuses, and blurry vision.
- ROS: + Dry mouth, + blood in stool (past two days).
- PPH: Significant for a hypermetabolic left upper lobe nodule recently discovered on PET scan and for similar symptomology four months prior that improved with a prednisone taper.
  - Prior admission revealed no unifying diagnosis and had negative rheumatological workup (neg. RF, ANA, SSB, SSB, ANCA, and myositis antibodies & negative temporal artery biopsy).
- Vitalts (on admission): BP: 166/83 | Pulse: 113 | Temp: 98.9°F (Oral) | SpO2: 94% Resp: 20 | BMI: 30.8 kg/m²
- Physical Exam: Patient in mild distress, but overall exam was unremarkable with no significant findings.
- Labs (on admission): Markedly elevated ESR/CRP (118 mm/hr; 199.5 mg/dL), lymphocytosis (12.4 x 1000/μL) and thrombocytosis (750 x 1000/μL); rheumatological workup negative (as before).
- Imaging (CXR): Showed confluent opacity in the left lung apex corresponding to the focal mass recently found on prior PET/CT (See Image 3a, 3b, and 3c).
- Pathology Report: Lung mass was biopsied and demonstrated lymphoplasmacytic infiltrate, fibrosis and areas of neutrophilic necrosis and occasional multinucleated giant cells consistent with changes of granulomatosis with polyangiitis (GPA).
- Plan: A preliminary diagnosis of GPA was made, and that led to a treatment with a prednisone taper with scheduled doses of outpatient rituximab. Four months after treatment, CT and CXR had shown resolution of lung mass (See Image 3d).

DISCUSSION
- 82 to 94 percent of patients with GPA are positive for c-ANCA, but about 10% are also c-ANCA negative [3].
- Generally, LRT presentations of GPA include nodules, cavitations or infiltrates, but less commonly can also include hypermetabolic lung nodules (as seen here).
- Though our patient’s clinical symptomology suggested GPA, an underlying vasculitis (sinusitis, myalgias, and blurry vision), the negative serological workup (-RF, -ANA, -ANCA) led us to disregard rheumatological causes and consider causes like infection and malignancy.
- This case study suggests that even though serological markers can help systematically diagnose conditions like GPA, these markers are neither necessary nor sufficient for diagnosis.
- Definitive diagnosis, when suspecting GPA, should be defined definitively with biopsychological presentation and infectious causes should be ruled out (as seen in path reports).
- Clinicians should remain cognizant of the varied presentations of GPA and understand that its diagnosis, while guided by certain serological markers, should not solely rely on serological markers.

CITATIONS