Multiple cavitary pulmonary nodules and episcleritis of the left eye: Granulomatosis with polyangiitis (GPA) masquerading as infection

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Introduction

Granulomatosis with Polyangiitis (GPA), previously known as Wegener's Granulomatosis, is characterized by the presence of granulomatous inflammation and necrotizing vasculitis involving small and medium sized vessels in various organs**.**

Most common initial manifestation:

- Chronic sinusitis (67%)
- Pulmonary infiltrates (71%)
- Conjunctivitis/Episcleritis /Uveitis (50%)
- Renal disease (17%)

Hospital Course

HPI: 77 year old Caucasian male with a PMHx of Squamous Cell Carcinoma of the lip (s/p resection & radiotherapy), DMII, CKD Stage III, HTN, Hyperlipidemia, & Vertigo presented to NGMC with 3 week history of:

- Productive cough
- Shortness of breath
- Dyspnea on exertion

ED Vitals: T:98.1 HR:69 BP: 132/65 RR: 20 O2: 92% RA

PE: Erythematous conjunctiva of the left eye and decreased breath sounds with rhonchi & rales throughout all lung fields

Labs:

12.2	12.7 / 39.7	338

135	101	52	264
4.7	28	1.71	

Microbiology: Blood, Sputum, AFB 3/3, and Urine Cultures, Urine antigens (Streptococcus Pneumoniae & Legionella), Influenza PCR, Mycobacterium Tuberculosis PCR all (-)

Started IV Vancomycin, Piperacillin/Tazobactam, and Azithromycin, transitioned to Ceftriaxone and Metronidazole due to (-) MRSA & Pseudomonas findings

Transferred to the ICU due to acute hypoxic respiratory failure with imaging showing pulmonary cavitary lesions [**Figure 1**]

Serine protease-3 (PR3) was elevated at 1,081 & Rheumatology was consulted due to high suspicion of Granulomatosis with polyangiitis

Methylprednisolone 500 mg IV qd for 3 days with a prednisone taper, and first dose Rituximab 375 mg/m^2 infusion (once a week x 4 weeks) improved overall respiratory symptoms and left eye irritation

Readmission

6 weeks s/p hospitalization, presented to ED with shortness of breath

Patient readmitted with saddle pulmonary artery embolus and multiple right lower extremity DVT's on Venous Duplex [**Figure 2**]

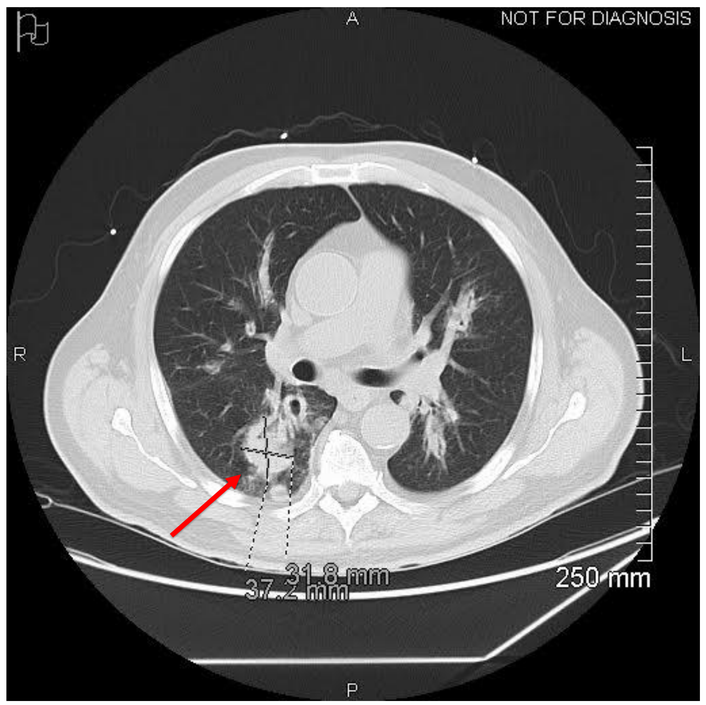

Figure 1 (Pre-treatment): CT of Chest w/o contrast depicting multiple cavitary lung nodules and b/l perihilar infiltrates; Largest area is in the RLL measuring 3.7 x 3.2 cm *(Image used with permission from Northeast Georgia Medical Center Diagnostic Radiology)*

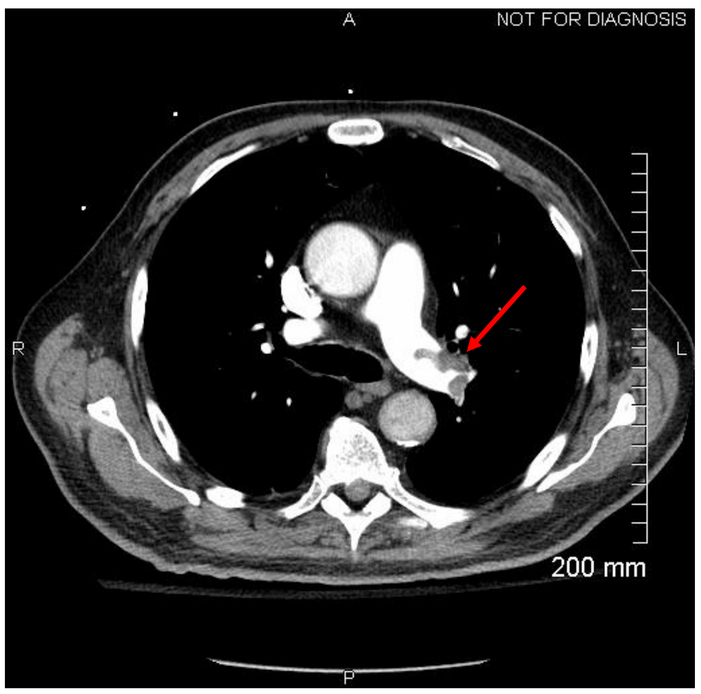

Figure 2 (Post-treatment): CTA depicting new saddle PE, cavitary lesion in the RUL measuring 16 mm (*Image used with permission from Northeast Georgia Medical Center Diagnostic Radiology)*

Discussion

This case demonstrates that the ACR 1990 Classification Criteria would have caused a missed diagnosis of GPA.

ACR/EULAR 2017 Provisional Classification Criteria for Granulomatosis with Polyangiitis (GPA) is more concentrated on clinical findings, reflects current investigative practices, and includes ANCA, but not critically dependent on a biopsy.

Need >=5 of the following to meet GPA criteria:
(Our patient Score: 8)

- Bloody nasal discharge, ulcers, crusting or sinonasal congestion (3)
- Nasal Polyps (-4)
- Hearing loss or reduction (1)
- Cartilaginous involvement (2)
- Red or painful eye (1)
- C-ANCA or P-ANCA (5)
- Eosinophil count ≥1 x10^9/L (-3)
- Nodule, mass or cavitation on chest imaging (2)
- Granuloma on biopsy (3)

Conclusion

Our patient predominantly had pulmonary and ocular involvement with no symptoms suggesting involvement of the upper respiratory tract or renal system.

Thorough workup and clinical diagnosis helped our patient to briskly recover from this rare and life-threatening disease.

C-ANCA/anti-PR3 has a high specificity (>95%) for the diagnosis of GPA.

Notably, GPA has an increased long-term risk of certain vascular events and should be evaluated for thrombotic complications if relevant symptoms develop [**Figure 2**].

This case illustrates the flaws of the ACR 1990 Classification Criteria for GPA, and highlights the need for an updated model, such as the ACR/EULAR 2017 Provisional Classification Criteria, where diagnosis can be made without need for biopsy.

References

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