

Introduction

- Granulomatosis with polyangiitis (GPA) is a rare necrotizing vasculitis that primarily affects the upper airways, lungs and kidneys.
- We present a patient who was newly diagnosed with GPA with a history of recurrent sinus infections that were precipitated by a Covid-19 infection from 1 year prior.

Case

HPI

- A 41-year-old Caucasian male with history of sinus infections during childhood, obesity and alcohol abuse presented to the hospital for a 3-week history of hemoptysis, arthralgias, and a petechial rash over bilateral lower extremities.
- He denied any personal or family history of autoimmune conditions.
- Patient tested positive for Covid-19 one year ago and has since had recurrent episodes of sinusitis symptoms which he would manage with over-the-counter medications.
- Three weeks prior to admission, he started having joint pains which began in the feet and ankles and progressed to the wrists and hands, a petechial rash over the bilateral anterior shins, and oral aphthous ulcers.
- On initial evaluation, patient was hypoxic with oxygen saturation of 84% and was placed on 3L oxygen via nasal cannula. He was otherwise hemodynamically stable.

Case Continued

Diagnostic Workup

- Labs revealed creatinine 1.73 (baseline 1.26), RF 41, ANA negative, c-ANCA positive, proteinase 3 positive, GBM negative. Respiratory viral panel, hepatitis screen and Quantiferon TB were negative.
- CT pulmonary embolism study showed multifocal ground glass opacities (GGO) and a reverse halo sign with concerns for alveolar hemorrhage.

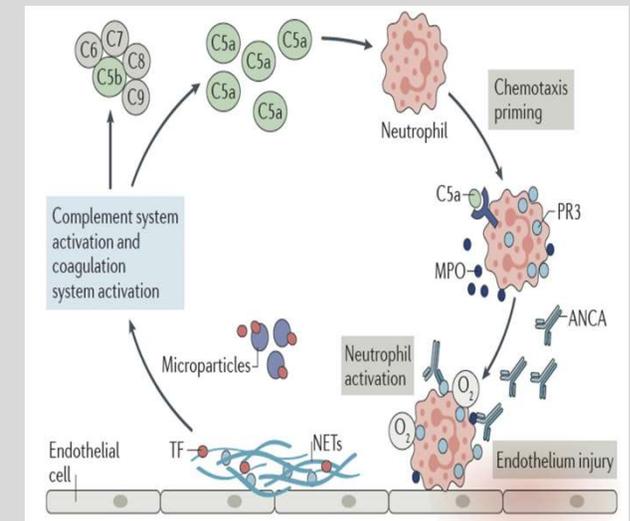


Treatment

- Patient was started on Rituximab and pulse steroid therapy. He received 3 sessions of plasmapheresis and a renal biopsy was performed; however, an insufficient sample was obtained.
- Patient was discharged on vitamin D and calcium supplementation and Bactrim for PJP prophylaxis with plan to taper prednisone by 10mg every 2 weeks.

Discussion

- This is a patient with no prior history of autoimmune disease who presented with classic symptoms of GPA and was diagnosed within 3 weeks of developing symptoms.
- GPA is an uncommon condition, and early diagnosis is imperative to start appropriate therapy and prevent irreversible complications.
- According to records, patient is scheduled to start Avacopan 30mg q12h in combination with Rituximab every 6 months.
- ADVOCATE Trial (2022)** demonstrated that avacopan was noninferior to using steroids for the treatment of ANCA-associated vasculitis.



References

Soulsby WD. Journal Club Review of "Avacopan for the Treatment of ANCA-Associated Vasculitis". ACR Open Rheumatol. 2022 Jul;4(7):558-561. doi: 10.1002/acr2.11412. Epub 2022 Feb 15. PMID: 35167187; PMCID: PMC9274376.