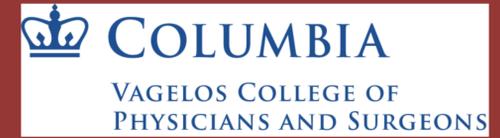


Mind The GPA: The Risk of Mortality in Granulomatosis with Polyangiitis

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Background

ANCA-associated vasculitis (AAV) frequently affects the small vasculature of the pulmonary and renal systems and can lead to alveolar hemorrhage and renal failure, amongst other complications. Granulomatosis with polyangiitis (GPA) is one of three ANCA-associated vasculitides. Treatment involves high dose corticosteroids and plasmapheresis for severe cases, however high dose steroids are associated with significant toxicity. Management has evolved due to several pivotal trials, and most recent data from PEXIVAS trial suggests lower prednisone doses maybe just as effective as the higher doses, with fewer complications. Here we present a case of a patient with GPA who required treatment with high-dose, long-duration steroids due to resistant pulmonary hemorrhage, and unfortunately succumbed to complications of a gastric perforation.

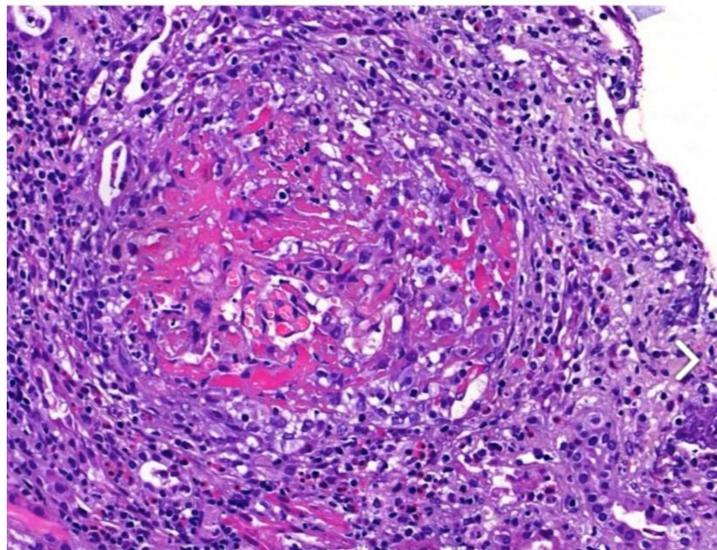


Figure 1: Anti-MPO related glomerulonephritis with segmental fibrinoid necrosis and destruction of the Bowman capsule is accompanied by inflammatory reaction (H&E).

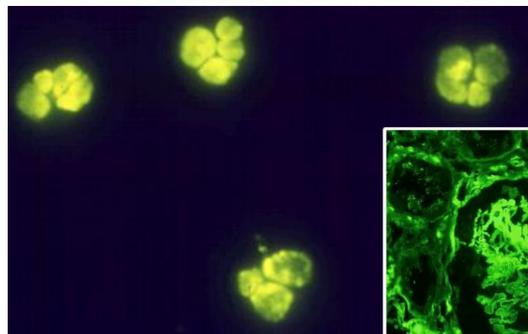


Figure 2: (P-ANCA) by indirect immunofluorescence with normal neutrophils.

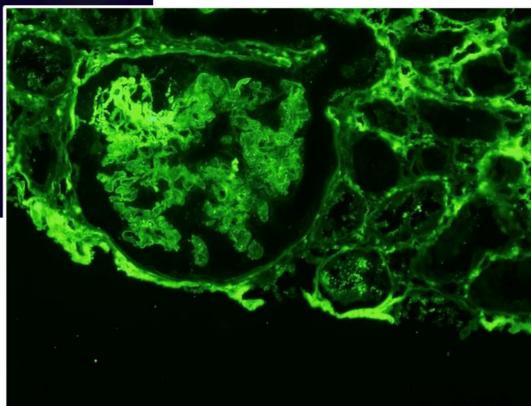


Figure 3: ANCA- Glomerulonephritis with segmental staining for fibrinogen (IF).

Case Presentation

A 76-year-old - previously healthy female presented with progressive shortness of breath and cough. She was treated as an outpatient with azithromycin for presumed atypical bacterial pneumonia without improvement in symptoms.

Physical exam was remarkable for:

RR of 24 and Oxygen saturation of 87% on room air
Crackles in bilateral lung bases

Labs were remarkable for:

Hemoglobin: 7.1 mg/dL
WBC: 15.3 k/mm³
Creatinine: 4.2 mg/dL from baseline of 1 mg/dL
Urinalysis: Moderate blood and 100mg/dL protein

IMAGE 1



CT Chest: Bibasilar ground-glass and consolidative opacities

IMAGE 2



CXR: Bilateral lower lobes pulmonary infiltrates. COPD.

Hospital course: On day 2 of admission, she developed new-onset hemoptysis. Kidney function deteriorated over the next several days and she required temporary dialysis. She was started on empiric pulse dose steroids and kidney biopsy confirmed severe diffuse crescentic, necrotizing pauci-immune glomerulonephritis with anti-MPO antibody/P-ANCA sero-positivity. She was treated with steroids and rituximab; however, she continued to have hemoptysis, prompting initiation of plasmapheresis and increase of her prednisone dose.

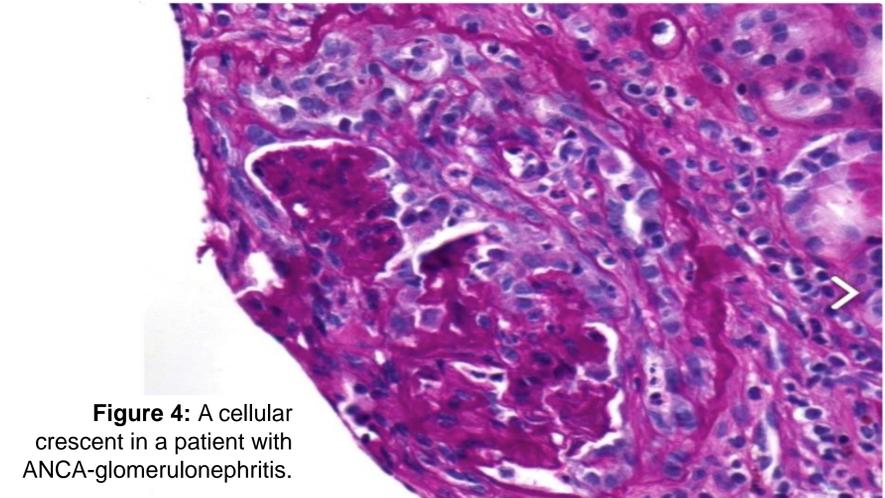


Figure 4: A cellular crescent in a patient with ANCA-glomerulonephritis.

Case Presentation

Due to the severity of her pulmonary disease, she was discharged on a 6-month course of prednisone, starting with 60mg daily with plan for a slow taper. She was also discharged with a plan for GI prophylaxis with omeprazole; however, she was not taking the medication. Despite improvement in her pulmonary and renal disease she was readmitted 1-month later for gastric ulcer perforation likely as a complication of steroid use, and unfortunately expired.

Discussion/Conclusion

This case outlines the severity and high risk for mortality in patients with GPA. Not only can the associated inflammation itself be fatal but immunosuppression is not without risk. Immunosuppressants have toxic adverse effects, such as the risk for peptic ulcer disease with steroid use as highlighted in this case. One needs to be vigilant about educating patients about the importance of prophylactic medications when on steroids. In addition, recent data from PEXIVAS suggests reduced dose glucocorticoid regimen may be as effective as the typical glucocorticoid regimen and should be considered, especially in older patients at higher risk for complications.

References

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