# Early Ocular Involvement in Granulomatosis with Polyangiitis

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## BACKGROUND

- **Granulomatosis with polyangiitis (GPA)** is an anti-neutrophil cytoplasmic antibody (ANCA) associated necrotizing vasculitis of small and medium blood vessels
- GPA most commonly affects the upper respiratory tract, lungs and kidneys
- **Ocular GPA is common** and has been described as one of the initial presentations of disease[1-4]
- Before modern immunomodulators, GPA had a median survival of 5 months and 1-year survival rate of less than 20%[5]

## OBJECTIVE

To highlight the importance of proper clinical workup in early ocular GPA to ensure swift initiation of remission induction and avoid serious complications

## CASE PRESENTATION

- 58-year-old hispanic male presenting to the ED with **vision loss, eye pain, conjunctivitis, headache**, night sweats, and productive **cough**
- Past medical history of **psoriatic arthritis** with joint and eye pain, vision changes, nail pitting, perioral erosions, and renal dysfunction secondary to **amyloidosis**
- Following **Bright scan** positive for **scleritis**, patient was begun on **60mg prednisone daily** and admitted for expedited workup

## HOSPITAL COURSE

- Upon presentation, the patient had **severely injected conjunctiva**, visual acuity 20/400 in his right eye and total blindness in his left eye (Image 1)
- Initial lab work showed **elevated CRP and ESR**; urinalysis showed **proteinuria** and **hematuria**.
- Bright scan and MRI orbits and brain were **concerning for lymphoma**
- Lymphoma workup identified a large **4.5 cm lung mass** with several smaller 1-2 cm masses with biopsies showing **necrotizing granulomatous inflammation** with **vasculitis** and negative stains and cultures
- Lab work found a positive **cytoplasmic ANCA (c-ANCA)** 1:320 as well as proteinase 3 (PR3) elevated at 8.0

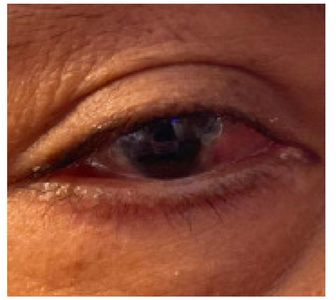

**Image 1.** Injected conjunctiva

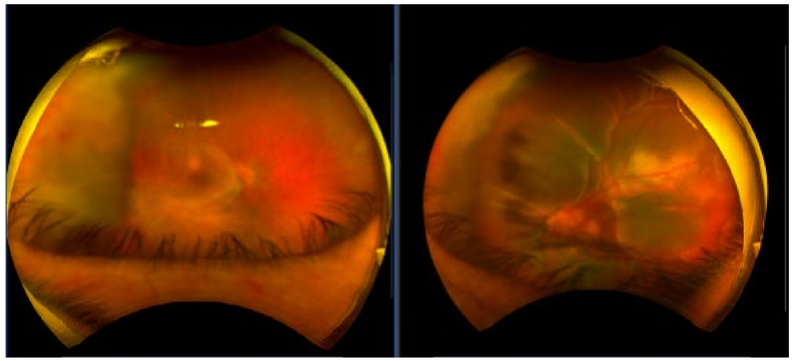

**Image 2.** MRI Orbits

## DIFFERENTIAL DIAGNOSIS

| | |
|---|---|
| **Ocular Lymphoma** | **Rheumatoid Arthritis** |
| **Infection** | **Systemic Lupus Erythematosus** |
| **Microscopic Polyangiitis** | **Sarcoidosis** |
| **Churg-Strauss Syndrome** | **Drug Toxicity** |

## IMAGING

- **MRI orbits** (image 2) with left eye retinal detachment
- Asymmetric enhancement and thickening of the left **globe choroid** and asymmetric thickening of the left **sclera** with minimal enhancement
- Enlargement and inflammatory changes with restricted diffusion involving the **left lacrimal gland**
- DDx: **Inflammatory** process versus neoplastic change d/t **lymphoma**

## CONCLUSIONS

- The presence of **ocular manifestations** of GPA (conjunctivitis, scleritis, and optic neuropathy) in the presence of other **multisystemic** signs of **vasculitis** should prompt screening for **c-ANCA** titers, which are present in 90% of patients with GPA[4]
- The 2022 American College of Rheumatology **diagnostic criteria** of GPA include[6]:
  - ✓ Bloody **nasal** discharge, nasal crusting, or sino-nasal congestion
  - ✓ **Cartilaginous** involvement
  - ✓ **Hearing** loss
  - ✓ **c-ANCA** or anti–PR3 ANCA positivity
  - ✓ **Pulmonary** nodules, mass, or cavitation on chest imaging
  - ✓ **Granuloma or giant cells** on biopsy
  - ✓ Inflammation or consolidation of the **nasal/ paranasal sinuses** on imaging
  - ✓ Pauci-immune **glomerulonephritis**
- Recognition of these criteria is essential in **reducing** the **high risk of severe sequelae** associated with untreated GPA.

## REFERENCES

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