# Frozen Eye in Granulomatosis with Polyangiitis (Wegener's)

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**ABSTRACT**

Ocular involvement in Granulomatosis with Polyangiitis (GP or Wegener Granulomatosis) is a potentially serious disease manifestation which needs prompt diagnosis and treatment. We report a case of a 59-year-old man with known GP for nine years who presented with a left frozen eye. Definite histological diagnosis with biopsy allowed prompt exclusion of infections and neoplasms and permitted institution of increased immunosuppression to avoid further progression of disease.

*Keywords: Wegener Granulomatosis, Ocular involvement, Histology, Diagnosis, Immunosuppression*

## INTRODUCTION

A 59-year-old man with known granulomatosis with polyangiitis (Wegener's) (GP) presented with sudden left eye visual loss. Examination revealed a left frozen eye with ptosis, proptosis, conjunctival injection, and absent pupillary light reflex (Fig. 1, please see overleaf). His GP presented nine years previously with symmetric polyarthritis, granulomas at nasal biopsy, and a positive anti-PR3. He was treated serially with cyclophosphamide, mycophenolate mofetil (MMF), then azathioprine 2.5 mg/kg/day, and prednisolone 7.5 mg/day maintenance therapy, with no respiratory or renal involvement at or after diagnosis. Computed tomography of the paranasal sinuses at the time of visual loss showed tissue thickening throughout the paranasal sinuses and bilateral medial orbital masses (arrows), resulting in lateral displacement of the medial rectus muscle and compression of the optic nerve (Fig. 2, please see overleaf).

## DISCUSSION

The differential diagnosis of such a mass in a patient with GP includes a relapse, infection (particularly orbital aspergillosis), or neoplasm (e.g. lymphoma). Histological diagnosis is needed for definitive diagnosis in this situation as noninvasive investigations have limited utility: e.g. serum galactomannan antigen assay to diagnose invasive aspergillosis has limited sensitivity (71%) and specificity (89%)[1], and a rise in anti-neutrophil cytoplasmic antibody titres does not confirm relapse of GP. Left medial orbital mass incision biopsy showed fibrosis with focal active leucocytoclastic vasculitis no granulomas or necrosis nor evidence of malignancy, fungal or mycobacterial infection. The patient was treated with high dose corticosteroids (1 mg/kg/day) and elected to use oral cyclophosphamide rather than rituximab[2], with the aim of preventing visual loss in the right eye. The patient's left periorbital swelling subsided. Loss of vision on the left eye persisted while right eye vision remained stable.

## CONCLUSION

This vignette illustrates the need for definite histological diagnosis in this clinical situation, biopsy allows prompt exclusion of infection and neoplasm, confirms the clinical diagnosis of GP

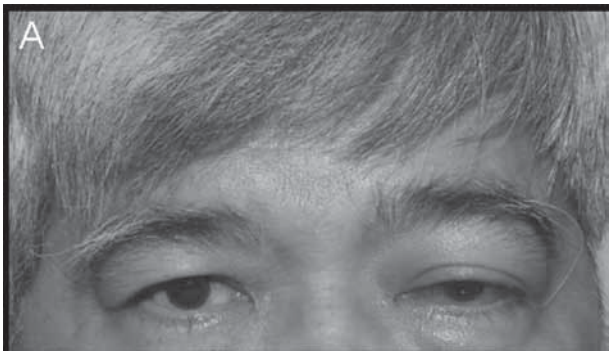


Fig. 1. Photograph of the patient's face shows a left frozen eye with partial ptosis, proptosis, and conjunctival injection.

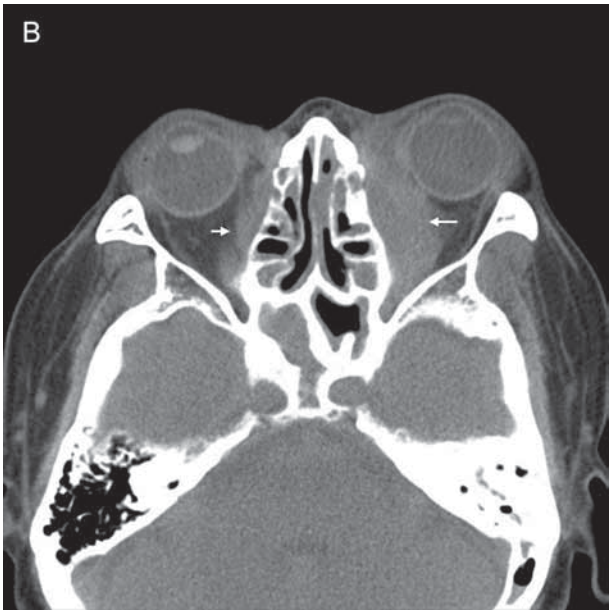


Fig. 2. Computed tomography of the paranasal sinuses shows tissue thickening throughout the paranasal sinuses and bilateral medial orbital masses (arrows).

relapse and permits institution of increased immunosuppression to avoid further progression of disease.

## REFERENCES

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