

Orbital Masses in Granulomatosis with Polyangiitis: A Call for Clinical Vigilance

Isabel Fonseca Silva¹ , Cristina Freitas² , Tomás Fonseca³ 

A 53-year-old female diagnosed with PR3-positive granulomatosis with polyangiitis (GPA), presented with symptoms including left upper eyelid swelling, proptosis, retro-orbital pain, and eye movement discomfort (Figures 1 and 2). Despite a history of severe disease activity leading to end-stage renal disease and subsequent kidney transplant, the patient had been managing well under long-term immunosuppressive therapy consisting of prednisolone, tacrolimus, and mycophenolate mofetil.

Imaging of the orbit revealed a significant mass in the extraconal orbital space, infiltrating the extraorbital muscles (Figure 3). Without signs of systemic GPA activity, a biopsy was performed, which revealed characteristic features of GPA, including leukocytoclastic vasculitis, fibrinoid necrosis, inflammatory infiltrate, and an epithelioid granuloma, with no evidence of neoplastic or infectious disease. The diagnosis of GPA pseudotumor was established, prompting the initiation of induction-remission treatment with steroids and rituximab. Given the clinical relapsing behavior of the pseudotumor, long-term maintenance therapy with rituximab was initiated, resulting in clinical improvement.

After kidney transplant and immunosuppressors, GPA relapse is rare.¹ This case highlights the importance of prolonged follow-up in patients with GPA, particularly in those who are PR3-positive, as they tend to have a higher relapse rate.² Orbital masses are a rare manifestation of GPA, but refractory and potentially devastating in their clinical presentation; therefore, it is crucial to accurately identify them, distinguish them from mimickers, and pursue an aggressive treatment.³

ORCID iDs of the authors:

I.F.S. 0000-0001-8499-0897;
C. F. 0009-0002-1101-7740;
T. F. 0000-0002-9386-1250.

Cite this article as: Fonseca Silva I, Freitas C, Fonseca T. Orbital masses in granulomatosis with polyangiitis: A call for clinical vigilance. *Eur J Rheumatol.* 2024;11(2):64-65.

¹ Department of Internal Medicine, Unidade Local de Saúde de Santo António, Porto, Portugal

² Department of Nephrology, Unidade Local de Saúde de Santo António, Porto, Portugal

³ Clinical Immunology Unity, Unidade Local de Saúde de Santo António, Porto, Portugal

Corresponding author:
Isabel Fonseca Silva
E-mail: isabelfonsecasilva93@gmail.com

Received: March 30, 2024
Accepted: May 23, 2024
Publication Date: September 5, 2024

Copyright@Author(s) - Available online at
www.eurjrheumatol.org.

Content of this journal is licensed under a Creative Commons Attribution-NonCommercial 4.0 International License.

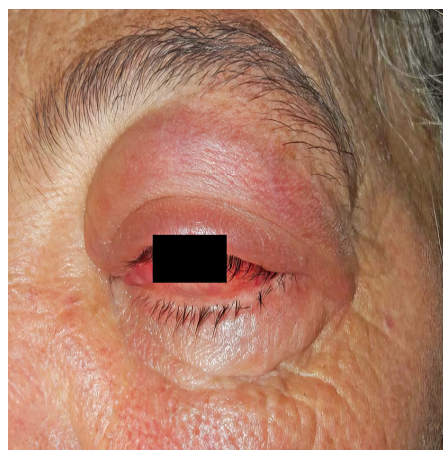


Figure 1 and 2. Marked left upper eyelid swelling.



Figure 3. Computed tomography showing on the left orbit, a mass of soft tissues, in the extra-conical topography, extending anteriorly and laterally to the subcutaneous tissues.

Informed Consent: written informed consent was obtained from the patients/patient who agreed to take part in the study.

Peer-review: Externally peer-reviewed.

Author Contributions: Concept – I.F.S., C.F., T.F.; Design – I.F.S., T.F.; Supervision – C.F., T.F.; Resources – I.F.S., C.F., T.F.; Materials – I.F.S., C.F., T.F.; Data Collection and/or Processing – I.F.S.; Analysis and/or Interpretation – I.F.S., C.F., T.F.; Literature Search – I.F.S., T.F.; Writing – I.F.S., T.F.; Critical Review – C.F., T.F.

Declaration of Interests: The authors have no conflicts of interest to declare.

Funding: The authors declare that this study received no financial support.

References

1. O'Brien FJ, Abdalla A, Wong L, et al. Recurrence of anti-neutrophil cytoplasmic antibody-associated vasculitis in appropriately immunosuppressed renal transplant patients: a discussion of two cases. *Case Rep Nephrol Urol.* 2013;3(1):16-21. [\[CrossRef\]](#)
2. Pierrot-Deseilligny Despujol C, Pouchot J, Pagnoux C, Coste J, Guillevin L. Predictors at diagnosis of a first Wegener's granulomatosis relapse after obtaining complete remission. *Rheumatol (Oxf Engl).* 2010;49(11):2181-2190. [\[CrossRef\]](#)
3. Muller K, Lin JH. Orbital granulomatosis with polyangiitis (Wegener granulomatosis): clinical and pathologic findings. *Arch Pathol Lab Med.* 2014;138(8):1110-1114. [\[CrossRef\]](#)