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Images in Rheumatology

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

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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 neoplasic 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]

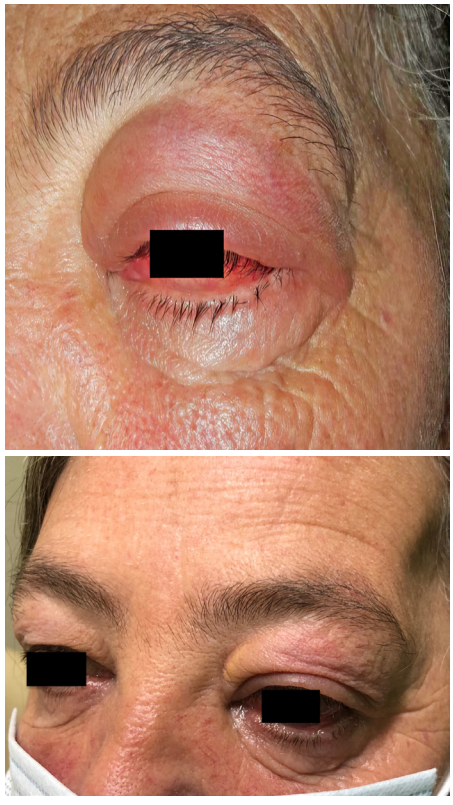

Figure 1 and 2. Marked left upper eyelid swelling.

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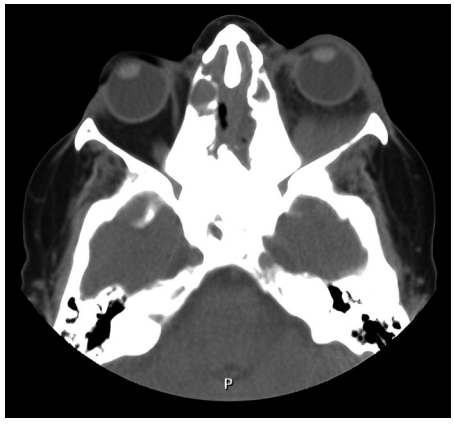

Figure 3. Computed tomography showing on the left orbit, a mass of soft tissues, in the extra-conical topography, extending anteriorly and laterally to the subcutaneous tissues.

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