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Orbital pseudotumor: An unusual complication in granulomatosis with polyangiitis

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Abstract
Orbital pseudotumor is a rare but serious manifestation of granulomatosis with polyangiitis (GPA). This complication can lead to severe ocular functional impairment and poses significant diagnostic and therapeutic challenges.
We report the case of a patient with GPA presenting an orbital pseudotumor, detailing the clinical, radiological, and therapeutic aspects. This case highlights the importance of a multidisciplinary approach to optimize patient management and, most importantly, the need to perform a biopsy of any suspicious mass to confirm the diagnosis.

Keywords: Granulomatosis with polyangiitis, orbital pseudotumor, systemic vasculitis

Introduction
Granulomatosis with polyangiitis (GPA) is a necrotizing granulomatous vasculitis of unknown etiology, with a particular tropism for the upper respiratory tract, lungs, and kidneys. However, certain localizations remain rare, or even exceptional. Among these, ophthalmologic involvement can manifest as an inflammatory orbitopathy in the form of intraorbital masses or orbital inflammatory pseudotumors, which typically occur during the course of the disease. This case of orbital pseudotumor complicating GPA illustrates the clinical polymorphism of the disease and highlights the diagnostic and therapeutic challenges it may present.

Case report
We report the case of a 54-year-old patient who has been followed for 5 years for granulomatosis with polyangiitis (GPA), diagnosed based on ENT and pulmonary manifestations associated with positive cANCA, and treated with long-term corticosteroid therapy. The patient was admitted to our service for painful left exophthalmos, progressively worsening, associated with a red eye and decreased visual acuity, without diplopia.
The ophthalmological examination, including fundus examination, revealed decreased visual acuity and left exophthalmos with limitation of ocular movements, resulting in a visual field defect. No signs of conjunctivitis, scleritis, uveitis, or retinal vasculitis were observed.
The orbitocerebral CT scan, performed with contrast injection, showed a heterogeneous mass in the left intra-orbital area extending into the adjacent soft tissues, without intracerebral extension or associated vascular complications.
Orbital and Cerebral Magnetic Resonance Imaging (MRI) revealed an ill-defined lesion involving both intra- and extra-conical areas, infiltrating the extraocular muscles and encasing the left optic nerve.
Histological examination of the lesion showed an inflammatory orbital pseudotumor, thus excluding a neoplastic, lymphomatous, or infectious origin.
The extension workup to search for other localizations revealed ENT involvement in the form of thickening of the sinuses, with no renal, pulmonary, or cardiovascular involvement.
The biological workup showed no significant abnormalities, except for positive cANCA.
The diagnosis of an inflammatory orbital pseudotumor complicating GPA was established. The patient was treated with oral corticosteroids, combined with cyclophosphamide boluses, in accordance with the standard vasculitis protocol. Given the moderate clinical improvement and partial regression of exophthalmos, the decision was made to escalate the treatment to rituximab.

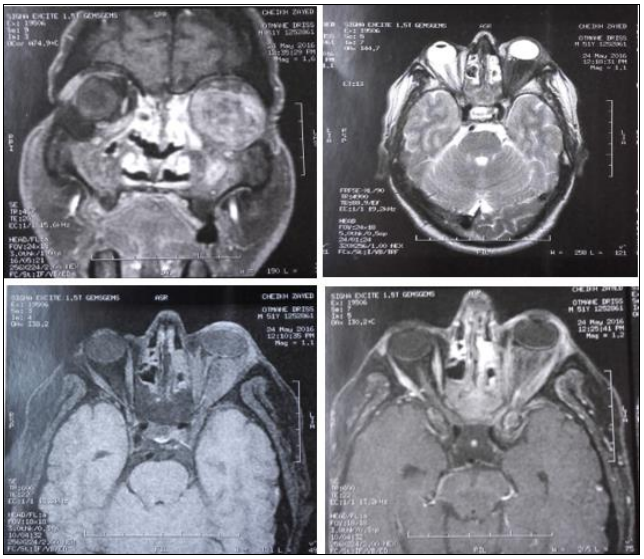

Fig 1: Orbital-cerebral MRI slices obtained in different T1 and T2 weightings sequences before and after gadolinium injection revealed an infiltrating mass in the left intra-orbital region involving intra- and extra-conical fat as well as the extraocular muscles, with extension to the orbital apex causing optic nerve compression and grade 3 exophthalmos

Discussion
Granulomatosis with polyangiitis is a multi-system necrotizing vasculitis of unknown etiology, histologically characterized by granulomatous inflammation, tissue necrosis, and vasculitis affecting the walls of medium- and, predominantly, small-caliber vessels. It primarily involves the upper and lower airways and the kidneys, but many

other organs and tissues can also be affected, including all orbital and ocular structures [1].

The frequency of ocular involvement in Wegener's disease varies between 28% and 87% [2, 3, 4, 5]. This is largely explained by the anatomical proximity of the eye to the upper airways.

All structures of the eye and its adnexa may be involved, either through contiguous extension of a nearby granulomatous lesion or due to focal necrotizing involvement of small vessels.

Inflammatory pseudotumors of the orbit are relatively rare ophthalmologic complications of the disease. They usually occur during an active phase of the disease and are often associated with involvement of other organs. However, they can sometimes be the dominant or even the sole manifestation of the disease [1].

These pseudotumors most commonly present as progressively worsening pain and unilateral proptosis [2, 6], as observed in our patient.

Other ocular manifestations may include ptosis, eyelid edema, epiphora, partial or complete limitation of ocular movements, and diplopia, which may result from a mass effect caused by the pseudotumor.

Orbital involvement, which is more often unilateral than bilateral, can have a poor visual prognosis due to optic nerve compression [7] and potential loss of visual function. This may occur when proptosis leads to complications such as exposure keratitis, corneal ulceration, or corneal perforation [8].

Painless ophthalmoplegia, causing diplopia, can occur in the absence of orbital pathology, due to vasculitis affecting either the ocular nerves or the extraocular muscles [6, 9].

Orbito-cephalic imaging, particularly CT and especially MRI with T1- and T2-weighted sequences, is an essential diagnostic tool. It helps reveal the nature of the orbital lesion, such as intraorbital fibrosis [8], and determine its extent.

The diagnosis can be confirmed or even established through targeted biopsy, which may demonstrate histological features characteristic of the disease [2]. Histologically, these lesions show replacement of inflammatory and necrotic areas by fibrotic tissue. However, it is critical to rule out other granulomatous pathologies and orbital conditions, such as idiopathic causes, lymphomas, or thyroid-related diseases [7].

The treatment approach does not differ between systemic and localized forms of the disease. In cases of ophthalmologic involvement, a favorable response is typically achieved with corticosteroids and/or immunosuppressants [6, 7, 10]. However, visual acuity outcomes may sometimes remain unfavorable.

For refractory forms, biologic therapy should be considered if the visual prognosis is at risk. If medical treatment fails, other options may be explored, including percutaneous radiotherapy, alcoholization of the ciliary ganglion for intractable pain [10], or orbital decompression in cases of optic nerve compression during acute inflammatory phases [11].

Conclusion

Granulomatosis with polyangiitis (GPA) can affect the entire orbital contents and all ocular tunics, either through contiguous extension or as localized vasculitis. Orbital pseudotumors are ophthalmologic complications of the disease that can lead to significant functional and aesthetic sequelae. This underscores the importance of regular ophthalmologic monitoring to detect subtle early signs, enabling timely and appropriate management, including immunosuppressive therapy, to preserve not only the visual prognosis but also the patient's overall survival.

Furthermore, it is essential to consider GPA in cases of atypical presentations and, most importantly, to perform a biopsy of any suspicious mass to confirm the diagnosis

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