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Case Report

Orbital Inflammatory Disease Revealing Granulomatosis with Polyangiitis (Wegener's Disease): A Case Report

Soukaina Azib*, Nejjar Y, Laaouina S, Adil Khoyaali and Yassine Mouzari
Department of Ophthalmology, Military Hospital Mohammed V, Rabat, Morocco

***Corresponding author:** Soukaina Azib, Department of Ophthalmology, Military Hospital Mohammed V, Rabat, Morocco
Email : azib.soukaina92@gmail.com

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Abstract

We report the case of a 25-year-old woman presenting with painful redness of the eye,bilateral exophthalmos, and decreased visual acuity in a context of general malaise and weight loss. Her medical history included chronic cough with persistent rhinorrhea unresponsive to treatment. Ophthalmic examination revealed bilateral, reducible, non-pulsatile axial exophthalmos, chemosis, and ophthalmoplegia. Laboratory investigations showed elevated ESR and CRP with leukocytosis and positive cytoplasmic ANCA. Chest radiography revealed multiple bilateral pulmonary nodules, and nasal biopsy demonstrated granulomatous inflammation. Orbital CT confirmed bilateral myositis. The diagnosis of granulomatosis with polyangiitis (Wegener's disease) was established. The patient responded favorably to high-dose corticosteroid pulses followed by oral steroids and immunosuppressive therapy. This case illustrates how orbital inflammation may be the initial manifestation of a systemic vasculitis such as Wegener's disease.

Keywords: Orbital Inflammation; Granulomatosis with Polyangiitis; Wegener's Disease; ANCA-Associated Vasculitis; Corticosteroids; Immunosuppressive Therapy

Introduction

Orbital inflammatory disease, also known as idiopathic orbital inflammation or pseudotumor, is an inflammatory condition that can affect any structure within the orbit.The inflammatory response may be nonspecific, granulomatous, vasculitic, or due to reactive lymphoid hyperplasia. It can occur as part of an underlying systemic disease or in isolation. All age groups may be affected, and the process can be acute or chronic,sometimes with recurrent episodes.

Case Presentation

A 25-year-old female presented with painful redness and decreased vision in both eyes,associated with bilateral proptosis and progressive general deterioration marked by weight loss. Past history revealed chronic cough with persistent rhinorrhea unresponsive to medical treatment.Ophthalmologic examination showed a visual acuity of 4/10 in the right eye and 6/10 in the left eye. There was bilateral conjunctival chemosis, axial, reducible, non-pulsatile exophthalmos, and ophthalmoplegia. The corneas were clear with negative fluorescein staining. The anterior chambers were deep and optically empty. Intraocular pressure measured 22 mmHg in the right eye and 21 mmHg in the left eye. Lenses were transparent and fundus examinations were normal. Laboratory tests showed elevated ESR and CRP with leukocytosis. Serum protein electrophoresis revealed hypergammaglobulinemia. Renal function was normal. Serology detected positive cytoplasmic ANCA (c-ANCA). Chest radiography showed multiple bilateral pulmonary nodules. Nasal biopsy revealed granulomatous inflammation. Orbital CT demonstrated bilateral extraocular muscle enlargement consistent with orbital myositis.The diagnosis of granulomatosis with polyangiitis (Wegener's disease) revealed by orbital inflammatory disease was established. The patient received intravenous corticosteroid pulses followed by oral therapy and immunosuppressive agents, with marked clinical improvement.

Discussion

Orbital inflammatory disease and granulomatosis with polyangiitis (formerly Wegener's disease) are distinct entities that may share overlapping clinical manifestations. Both can present

Figure 1: Clinical appearance before treatment showing bilateral painful red eyes with conjunctival chemosis and axial proptosis.

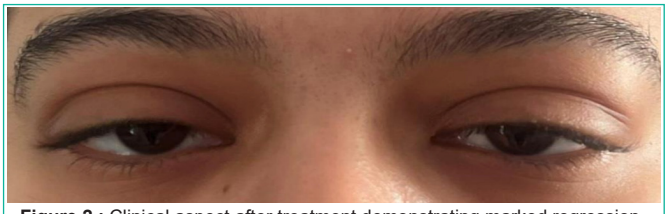

Figure 2 : Clinical aspect after treatment demonstrating marked regression of orbital inflammation and normalization of ocular appearance.

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with orbital pain, proptosis, ophthalmoplegia, and inflammatory signs.Granulomatosis with polyangiitis is a systemic necrotizing vasculitis affecting small- and medium-sized vessels, characterized by granulomatous inflammation of the respiratory tract and kidneys. When orbital inflammation represents the first manifestation, it can mimic idiopathic orbital inflammation, delaying diagnosis. The presence of systemic symptoms, pulmonary or sinus lesions, positive c-ANCA, and histological confirmation of granulomas are key diagnostic clues. Treatment relies on high-dose corticosteroids combined with immunosuppressive agents such as cyclophosphamide or rituximab, aiming to induce remission and prevent relapse. Early diagnosis and multidisciplinary management are crucial for visual and systemic prognosis.

Conclusion

Orbital inflammation encompasses a broad spectrum of inflammatory disorders involving intra-orbital structures, including anterior, diffuse, apical, myositic, and dacryoadenitis forms. Differentiating between idiopathic and specific inflammatory causes is essential.Granulomatosis with polyangiitis should be considered in young patients with orbital inflammation associated with systemic or sinonasal findings. Prompt immunosuppressive therapy leads to favorable visual and systemic outcomes.

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