

Uncommon Debut: Orbital Pseudotumor as the Initial Presentation of IgG4-Related Disease

Pseudotumor Orbitário como Apresentação Inicial de Doença Relacionada com IgG4

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Palavras-chave: Doença Relacionada a Imunoglobulina G4; Pseudotumor Orbitário.

Keywords: Immunoglobulin G4-Related Disease; Orbital Pseudotumor.

A 73-year-old man was admitted due to a 4-year bilateral periorbital edema, foreign body sensation, significant weight loss, and anorexia. He presented significant bilateral upper eyelid edema with proptosis of the left eye (Fig.1 A, B). Blood

analysis showed eosinophilia, and elevated Immunoglobulin E and G4 (IgG4). Magnetic resonance imaging of the orbits showed marked thickening of the retro-orbital muscles and prominence of the pre-septal periorbital soft tissues bilaterally (Fig.1 C, D). A biopsy of the lacrimal gland showed lymphoplasmacytic infiltrate with a significant number of IgG4 plasma cells. IgG4-related disease was diagnosed. Prednisolone was started, followed by rituximab, with sustained clinical remission.

IgG4-related disease (IgG4-RD) is a rare fibroinflammatory disorder characterized by diffuse organ infiltration.¹ IgG4-RD in the eye and ocular adnexa (IgG4-ROD) usually

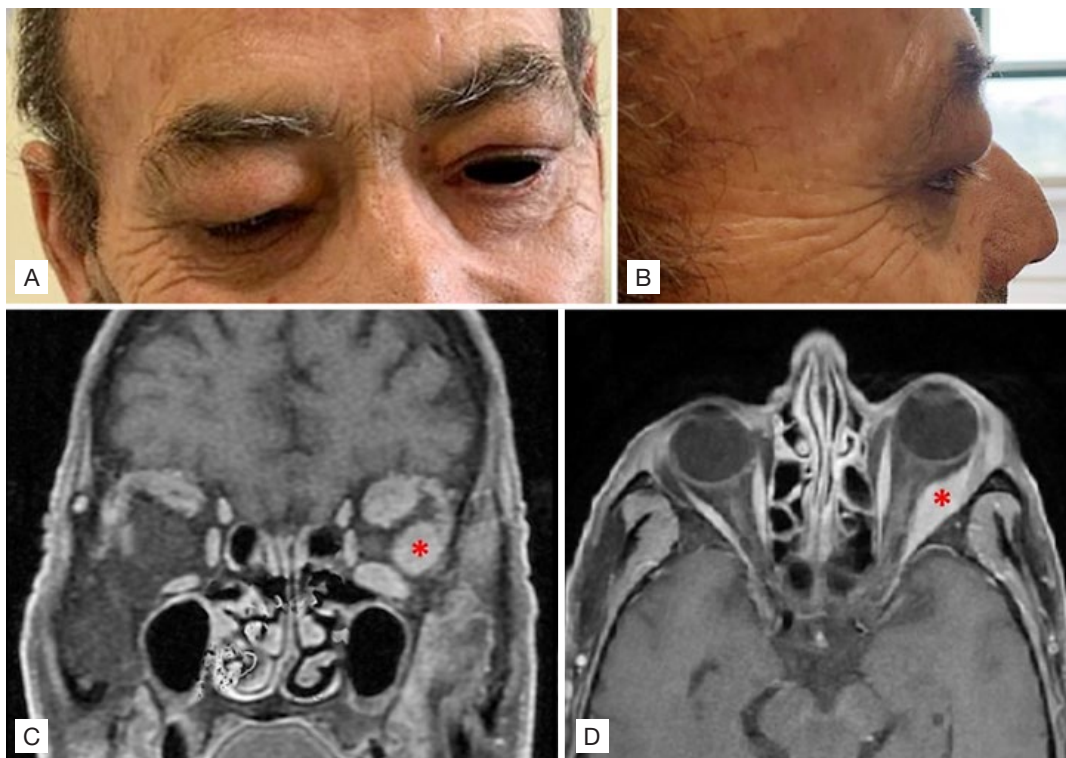


Figure 1: A and B: Clinical presentation: significant bilateral upper eyelid edema with masses in the superior temporal orbit suggestive of enlarged lacrimal glands and proptosis of the left eye. C and D: Magnetic resonance imaging of the orbits: bilateral thickening and increased enhancement of the pre-septal periorbital soft tissues and extraocular muscles (*) that is more pronounced on the left, compatible with inflammatory changes.

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presents as a chronic, painless swelling of eyelids or proptosis of the orbit. The mean time to diagnosis is 4–5 years due to its unspecific manifestations, slow progression and the need to exclude other common mimickers (such as thyroid-associated orbitopathy or malignancy).² Providers must have a high level of suspicion of this syndrome and red flags to early diagnosis are an unmet need, since adequate therapy is critical for IgG4-RD prognosis.³ ■

Declaração de Contribuição

ACM - Pesquisa bibliográfica, avaliação do paciente e redação do artigo.
ARC - Aquisição e interpretação das imagens, revisão crítica do manuscrito.
TF - Revisão crítica do manuscrito.
Todos os autores aprovaram a versão final a ser publicada.

Contributorship Statement

ACM - Literature search, patient assessment and writing the article.
ARC - Acquisition and interpretation of images, critical revision of the manuscript.
TF - Critical revision of the manuscript.
All authors approved the final draft.

Responsabilidades Éticas

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