Bilateral Orbital Apex Syndrome Due to Granulomatosis with Polyangiitis

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A 55-year-old woman was presented to our Emergency Department (ED) with bilateral orbital apex syndrome (OAS). She had a 3-month history of headache, photophobia, subtle left ptosis, and bilateral forehead numbness, which prompted repeat ED visits. Cranial and orbital imaging on previous presentations was reported as normal. She had sensory complaints with a confirmed, mild sensory mononeuritis multiplex, and elevated myeloperoxidase autoantibodies at >8.0 U (normal < 1.0), raising suspicion of an ANCA-associated vasculitis. Over 1 month, her vision deteriorated with worsening retro-orbital pressure and pain with eye movements. Examination revealed visual acuity of 20/200 on the right and finger-counting on the left, left afferent pupillary defect, and bilateral inferior altitudinal hemianopia. Funduscopy showed raised left optic nerve with blurred nasal optic disc margins. There was severe bilateral ophthalmoplegia, bilateral periorbital swelling, left ptosis, and right V1 hypoesthesia. Remaining neurological examination was unremarkable. CT imaging revealed a soft tissue abnormality in the superior aspect of the left orbit, which was confirmed on MRI as an enhancing extraconal soft tissue mass (Figures 1 and 2). Enhancing infiltrates was also present in both orbital apices, yet more subtle on the right (Figure 2). Left orbital biopsy confirmed granulomatosis with polyangiitis (GPA). Treatment was initiated with IV steroids, followed by IV cyclophosphamide and ongoing oral prednisone. At follow-up, visual acuity and ductions returned to normal.

Orbital masses are a rare manifestation of GPA and associated with a refractory course and significant morbidity.[1] GPA, formerly Wegener's granulomatosis, is a rare autoimmune disorder characterized by granulomatous inflammation and necrotizing vasculitis of small- and medium-sized blood vessels. 50%–60% of GPA patients have ophthalmic involvement, with up to 16% presenting initially with ocular findings.[2] Of ocular presentations, orbital involvement is most frequent at 22%–45%.[3] Orbital manifestations may be secondary to primary granulomatous vasculitis or contiguous sinus involvement.[2] The latter mechanism is more common; infiltration from paranasal sinuses is demonstrated in 72% of orbital GPA cases.[1]

OAS comprises dysfunction of cranial nerves II, III, IV, VI and the ophthalmic branch of V and is reported in only a handful

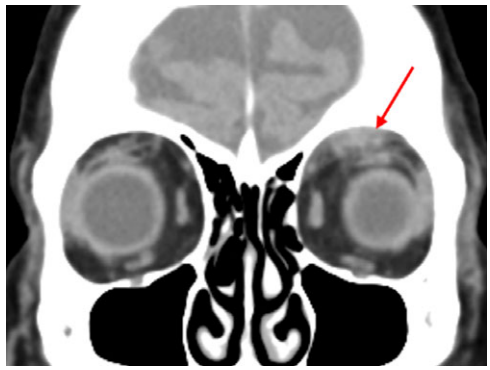

Figure 1: *Abnormal soft tissue is demonstrated on CT post-contrast (red arrow) involving the left superior orbit.*

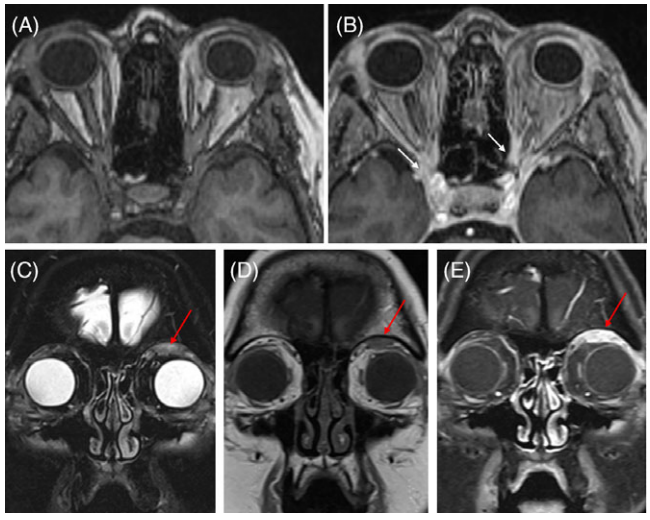

Figure 2: *MRI orbital imaging sequences showing T1 pre-contrast (A), T1 post-contrast (B), T2 fat-sat pre-contrast (C), T1 fat-sat pre-contrast (D) and T1 fat-sat post-contrast (E). Soft tissue mass involving the left superior extraconal fat tissue and superior rectus muscle (red arrows).The axial MRI MPR images show enhancing soft tissue extending from the orbital apices into the bilateral cavernous sinus (white arrows) with resultant apical crowding.*

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of cases of GPA. In most of these cases, OAS was caused by contiguous sinus or pachymeningeal infiltration.[3–8] OAS without contiguous sinus spread has been reported, but occurred in a patient with a pre-existing confirmed diagnosis of GPA.[9]

This case highlights several key points. First, despite profound ophthalmoplegia and severely compromised visual acuity, imaging findings were relatively subtle. In fact, to our knowledge, there has been only one previously reported case of ANCA-associated vasculitis causing OAS without corresponding radiographic abnormalities detected on MRI.[10] Second, despite a suspicion of systemic vasculitis given a mild sensory mononeuritis multiplex, an orbital biopsy was required to establish a definite histopathologic diagnosis. Finally, although the optic neuropathy and oculomotor dysfunction were severe at presentation, recovery to normal acuity and ductions demonstrates the robust long-term response to immunosuppression in this condition. In sum, close attention to the orbits is warranted when considering a diagnosis of ANCA-associated vasculitis. Imaging findings can be subtle, and diagnosis may be missed.[7]

Disclosures

All authors report no disclosures relevant to the manuscript.

Statement of Authorship

DK was involved in first draft, literature review, study concept, and design. CF was involved in literature review, critical review of manuscript for intellectual content, study concept, and design. MVDB and TV were responsible for creation of figures, figure legends, and critical review of manuscript for intellectual content.

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