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CASE REPORT

A Stepwise Multimodality Treatment of Diffuse Angiolymphoid Hyperplasia of the Orbit

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ABSTRACT

The definite treatment of angiolymphoid hyperplasia is debatable. We report the case of a middle-aged man with an extensive angiolymphoid hyperplasia of the orbit that has been recalcitrant to multiple single-line treatments for 9 years. His previous treatment included several short courses of full-dose systemic steroids, debulking surgeries, and orbital radiotherapy. A stepwise multimodality treatment approach in this case could achieve a lasting satisfactory functional and cosmetic outcome.

Keywords: Angiolymphoid hyperplasia, multimodality, orbit

Angiolymphoid hyperplasia with eosinophilia (ALHE) is a clinicopathological entity that presents with chronic pruritic dermal nodules in the head and neck region, and may primarily involve orbital structures.[1–4] ALHE shares several clinical and pathological similarities with Kimura disease, which mainly affects Asian patients and may be associated with lymphadenopathy or nephrotic syndrome from immunoglobulin E deposition.[1,2] ALHE has been described as ''epithelioid hemangioma'' as it is characterized by vascular proliferation with plump endothelial cells, and chronic inflammatory cellular infiltration with predominance of eosinophils. The proliferating endothelial morphology distinguishes ALHE from Kimura disease.[3,4] We describe the treatment approach in a patient presented with massive orbital involvement with ALHE.

CASE REPORT

A 42-year-old Caucasian male was referred to evaluate right eye chronic proptosis. He had developed bilateral masses at the upper temporal quadrants of both orbits 9 years prior. The left orbital mass was surgically excised then, but an incision biopsy was obtained from the right orbital mass, which was followed by progressive enlargement of the mass causing significant right eye protrusion. The histology of the orbital biopsies identified ALHE, showing the typical features of proliferating vessels with large endothelial cells, and inflammatory infiltrates, composed mainly of lymphocytes and eosinophils. Peripheral blood eosinophilia and serum IgE elevation were not detected.

Along the years that followed, he had been treated with several short courses of 70–100 mg/day of oral prednisolone that temporarily improved proptosis, which recurred shortly after discontinuing the drug. He underwent 2 debulking surgeries of the retrobulbar mass at 3 and 5 years before referral that were followed by proptosis recurrence a few weeks after. One year before evaluation, he underwent fractionated stereotactic radiotherapy of 40 Gray over 10 sessions that produced a minor response.Examination revealed right eye proptosis of 9 mm on Hertel exophthalmometry, with 2 mm of lagophthalmos and minimal limited abduction. Right eye was resistant to retropulsion, and not associated with pulsations. Prominent subconjunctival vessels were noticed at the lateral rectus muscle insertion, extending to the lateral conjunctival fornix (Figure 1A). Best-corrected

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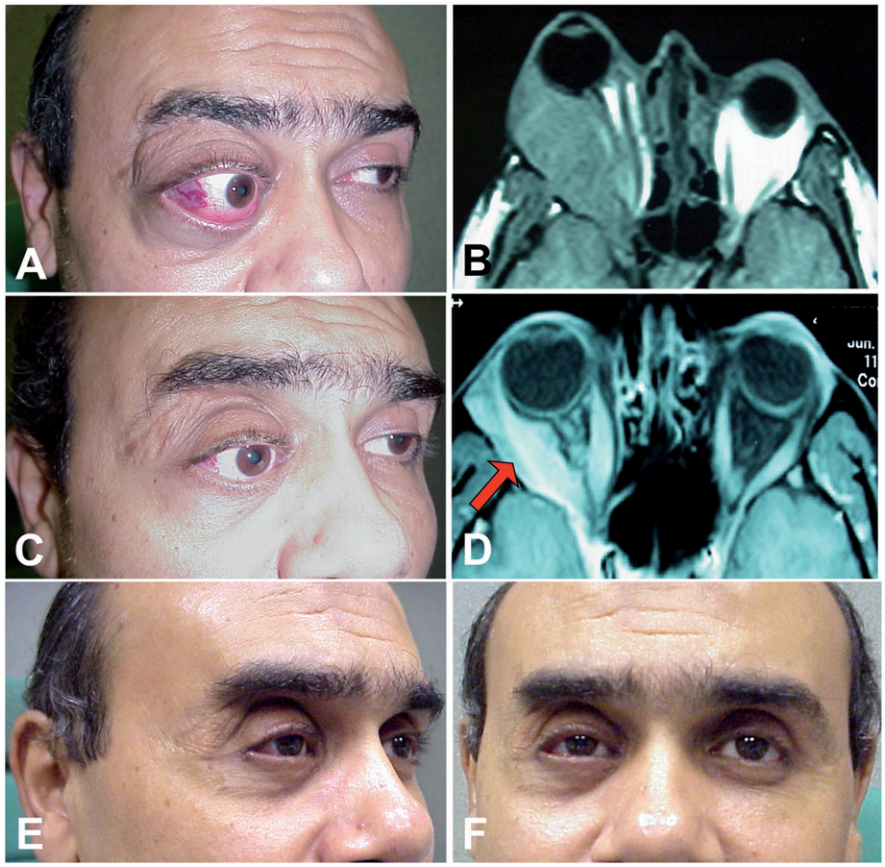

FIGURE 1. (A) Right eye proptosis associated with prominent subconjunctival engorged vessels at the lateral rectus muscle insertion; (B) T1-weighted MRI illustrates an extensive homogeneous isointense mass encompassing the right lateral rectus muscle; (C) Significant reduction of right eye proptosis after 3 weeks of combined treatment with steroids and Azathioprine; (D) T1-weighted MRI image with gadolinium enhancement and fat suppression shows significant reduction of the right orbital mass after 3 weeks of oral treatment. The lateral rectus muscle belly shows contrast enhancement within the residual orbital mass (arrow); (E) Lateral view; (F) Frontal view photographs at 3 weeks postsurgical excision of the residual lesion surrounding the right lateral muscle showing resolution of proptosis with minimal right enophthalmos.

visual acuity was 20/25 in both eyes, and bilateral ocular examination was otherwise normal. Orbital MRI revealed a large homogeneous isointense mass on T1 image, encompassing the lateral rectus muscle and extending between the lateral orbital wall and optic nerve (Figure 1B). Based on the earlier histopathologic diagnosis of ALHE, a combination oral therapy of prednisolone 25 mg/day and azathioprine 50 mg/day was prescribed to reduce tumour size.

A significant reduction in proptosis to 4 mm was noticed after 3 weeks from starting oral therapy (Figure 1C). There was no further proptosis reduction 3 weeks later. MRI was repeated then with gadolinium enhancement and fat suppression could highlight the lateral rectus muscle belly within the residual orbital mass (Figure 1D). Surgical excision of the remaining mass was then performed by careful dissection along the muscle fibres on the medial and lateral aspects of the lateral rectus muscle through transconjunctival approach. Three weeks after surgery there was right eye enophthalmos of 1 mm with bilateral full ocular motility (Figures 1E, 1F). The patient was subsequently maintained on a low dose of prednisolone 5 mg/day every 2 days that prevented relapse of proptosis for the subsequent 4 years of follow-up.

Comment

The ultimate treatment of ALHE is controversial. Complete surgical excision is typically the preferred treatment.[5–7] This was demonstrated in this patient by absence of proptosis recurrence following earlier surgical excision of the ALHE lesion in his left orbit. The right orbital lesion that progressed after the initial incomplete excision was considered too extensive to be surgically excisable at the time of referral. Other suggested treatments of ALHE mentioned in the ophthalmic and dermatologic literature provided variable outcomes, including systemic and local corticosteroids, cytotoxic agents, radiotherapy, carbon dioxide laser ablation, immunosuppressive agents, and oral retinoids.[5–9]

The lesion in this patient was refractory to all single-line treatments that included short courses of full dose systemic steroids, debulking surgeries, and orbital irradiation. This prompted stepwise multimodality treatment approach that yielded a final satisfactory outcome. The rationale of combining low dose steroids and a cytotoxic agent as an initial treatment was to reduce the lesion dimensions in preparation for surgical excision, while avoiding the adverse effects from using the full dose of a single agent for a prolonged period. Based on the MRI findings after medical therapy, surgical excision of the residual lesion around the muscle was achievable, without affecting the muscle function, to substantially reduce the disease burden. Lastly, a minimal dose of steroids as a maintenance therapy could check regrowth of any residual disease, without systemic sequelae.

In conclusion, this stepwise multimodality approach could achieve excellent functional and cosmetic outcome with no subsequent disease relapse in a case of extensive orbital involvement with ALHE.

DECLARATION OF INTEREST

The authors report no conflicts of interest. The authors alone are responsible for the content and writing of the paper.

REFERENCES

1. Buggage RR, Spraul CW, Wojno TH, Grossniklaus HE. Kimura disease of the orbit and ocular adnexa. *Surv Ophthalmol* 1999;44(1):79–91.
2. Baráková D, Sâch J, Kuchynka P, et al. Angiolymphoid hyperplasia with eosinophilia with bilateral involvement of the lacrimal glands. *Klin Monbl Augenheilkd* 2002; 219(5):376–379.
3. McEachren TM, Brownstein S, Jordan DR, et al. Epithelioid hemangioma of the orbit. *Ophthalmology* 2000; 107(4):806–810.
4. Fernandes BF, Al-Mujaini A, Petrogiannis-Haliotis T, et al. Epithelioid hemangioma (angiolymphoid hyperplasia with eosinophilia) of the orbit: a case report. *J Med Case Rep* 2007;25:1–30.
5. Francis IC, Kappagoda MB, Smith J, Kneale K. Kimura's disease of the orbit. *Ophthal Plast Reconstr Surg* 1988;4: 235–239.
6. Hidayat AA, Cameron JD, Font RL, Zimmerman LE. Angiolymphoid hyperplasia with eosinophilia (Kimura's disease) of the orbit and ocular adnexa. *Am J Ophthalmol* 1983;96(2):176–189.
7. Sánchez-Acosta A, Moreno-Arredondo D, Rubio-Solornio RI, et al. Angiolymphoid hyperplasia with eosinophilia of the lacrimal gland: a case report. *Orbit* 2008;27(3):195–198.
8. Baker MS, Avery RB, Johnson CR, Allen RC. Methotrexate as an alternative treatment for orbital angiolymphoid hyperplasia with eosinophilia. *Orbit* 2012;31(5):324–326.
9. Gencoglan G, Karaca S, Ertekin, B. Angiolymphoid hyperplasia with eosinophilia successfully treated with imiquimod. *Dermatology* 2007;215:233–235.