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Sir,
Rituximab in IgG4-related inflammatory disease of the orbit and ocular adnexae

The discovery of IgG4 implication in a subtype of previously-idiopathic orbital disease is beginning to change the disease management. In 2002, ‘the mainstay of therapy for idiopathic orbital inflammation (was) corticosteroids’,¹ with often excellent but unsustainable treatment response. Specific immunomodulatory therapy is now being investigated and early results show promise. Of particular interest is Rituximab (chimaeric monoclonal antibody against B-cell CD20).

The efficacy and safety of Rituximab has been demonstrated for systemic and intraocular inflammatory conditions.^{2,3} Since the submission of a review on adnexal IgG4-related disease,⁴ papers have been published on the use of Rituximab for systemic IgG4-related disease and data are now emerging on the use of Rituximab for IgG4-specific orbital disease.

The first is a case report of a 56-year-old lady with over 30 years of intractable orbital disease presumed to be idiopathic.⁵ Recent interest in IgG4 has led to further serum samples and tissue biopsy (lacrimal gland, extraocular muscles, intraconal fat, and trigeminal nerve) showing levels of IgG4 above the reference range. Six months after commencing, Rituximab proptosis improved, serum IgG4 normalised and orbital disease was deemed dormant.

The second paper is a review of 10 cases with IgG4-related orbital disease unresponsive to oral steroids and disease-modifying antirheumatic drugs (DMARDs).⁶ All patients received two infusions of Rituximab (1000 mg) 15 days apart. Nine of ten patients demonstrated ‘striking clinical improvement’ after 1 month of starting treatment. The remaining patients’

disease progression was halted, but no clinical improvement was evident. All 10 patients were able to discontinue oral steroid and DMARDs. Four patients required re-treatment at 6 months, with repeatable clinical improvement and serum IgG4 reduction.

Evidence is encouraging but of low scientific value, with no direct comparison to current standard care (prednisolone). Higher-level, prospective and randomised evidence investigating Rituximab against glucocorticoids would be beneficial. However, powering a study for a disease with such heterogenous clinical manifestations and poorly definable outcomes doubtless limits evidence supporting Rituximab to case-series data only.

Conflict of interest

The author declares no conflict of interest.

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Sir,
Subtarsal eyelid examination using an oblique slit lamp mirror in cases of eyelid shortening

We report a novel technique enabling examination of the superior fornix and tarsus in patients in whom the eyelid cannot be everted.