

LETTERS TO THE EDITOR

Local interferon alfa-2b for ocular cicatricial pemphigoid

EDITOR.—Ocular cicatricial pemphigoid (OCP) is a relatively uncommon, chronic, fibrotic, progressive, bilateral, potentially blinding disease of the ocular surface.¹ The finding in most cases of antibodies binding the basement membrane of conjunctival epithelial cells indicates an immune pathogenesis. Currently, systemic immunotherapy is required to achieve remissions of active OCP,² risking significant morbidity and potential mortality. Our usual treatment is a combination of corticosteroids with either methotrexate, 10–30 mg per week, or cyclophosphamide, 1–3 mg/kg per day. Side effects of steroids include osteoporosis, infection, weight gain, and myopathy, while immunosuppression increases the risk of leucopenia, malignancy and internal organ toxicity.

Interferon alfa-2 (IFN α) may offer significant advantages over current treatments for OCP, since it has effects which may be active against both the immune³ and the fibrotic^{4, 5} components of the disease. The apparent ability of IFN α to reduce the permeability of microvascular endothelium might also be beneficial.⁶

CASES

We have administered IFN alfa-2b (Schering-Plough, Kenilworth, NJ, USA) by subconjunctival injection to five patients suffering from OCP. All patients had active conjunctival inflammation with symblepharon formation in the treated eye and were poor candidates for systemic immunosuppression because they were either too old and infirm (patients 1 and 2) or they had been unable to tolerate it (patients 3–5). Interferon alfa-2b was given in doses of 1.5×10^6 IU twice weekly for 3 weeks then weekly for another 3 weeks for a mean total dose of 12.8 (range 9–16) $\times 10^6$ IU. After topical anaesthetic had been given, the drug was injected in a volume of 0.2 ml beneath the posterior bulbar conjunctiva, preferably in an area uninvolved by symblepharon. Only the eye that displayed the more advanced disease was treated in each patient, the other eye serving as a control.

Table 1 shows the results of a pretreatment conjunctival biopsy and the pre- and post-treatment (most recent) OCP scores⁷ for each patient. This OCP grading scheme, devised by us, takes into account inflammation, acuity, and the sequelae of both dryness and fibrosis. Patients 1 and 2 had OCP related to topical glaucoma medication⁸ which had already been

discontinued. The scores of the treated eyes were unchanged in two patients and improved in three. Interestingly, no progression was found in untreated eyes. All patients reported a gradual easing of their symptoms of chronic ocular discomfort in the treated eye during the months after treatment. No significant adverse events were found, nor has any patient required subsequent treatment in either eye apart from occasional topical steroid preparations.

COMMENT

The mechanism of action of IFN α against OCP is uncertain. The lack of progression in untreated eyes may be simply because the disease was less active in that eye, as it was in each of our cases, but it also raises the possibility of an effect of IFN α on the systemic immune response. This would be consistent with ability of IFN α to downregulate class II MHC antigen expression³ resulting in a reduced antigen load visible to immune surveillance mechanisms. The greater effect in the treated eye, however, suggests that local factors, such as its antifibrotic effect, might also be important.

When combined with the data from a previous study,⁹ these results show that subconjunctival IFN α as we administered it appears to be relatively safe in humans. Systemic problems are very unlikely at this low dose, as are local side effects. Although systemic IFN α has occasionally been associated with the development of autoimmune disease, no such ocular association has ever been described. Similarly, we have not found any evidence of retinal vascular disease in our patients.

While these results are encouraging with respect to efficacy, the number of patients is too small to allow any firm conclusions to be drawn. We recommend that interferon alfa be used for OCP only in the context of a prospective clinical study.

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Unilateral pellucid marginal degeneration in an elderly patient

EDITOR.—Pellucid marginal degeneration (PMD) of the cornea is a disorder characterised by bilateral, clear, inferior, peripheral corneal thinning.^{1,2} It is usually diagnosed between 20 and 40 years of age.¹ I present the unusual case of unilateral pellucid marginal degeneration in a 60-year-old patient.

CASE REPORT

A 60-year-old black man reported normal vision in both eyes as a child. Around the age of 30 he noted progressive blurring in his right eye, which he attributed to mild blunt trauma he suffered to his eye around that time. No ocular surgery or treatment was performed after the trauma. His vision gradually worsened in the right eye. At no time was the eye inflamed. He denied any problems in his left eye. His medical history was notable for atherosclerotic cardiovascular disease, asthma, and hypertension. He denies arthritis and seasonal allergy. There is no family history of eye disease, including keratoconus or corneal dystrophy.

On examination, best corrected visual acuity was 20/200 in the right eye and 20/20 in the left with $-10.75 +7.25 \times 180$ right and $-0.25 +0.75 \times 180$ left. Laser interferometry revealed 20/40 potential acuity right. There was no afferent pupillary defect. Motility was normal, but there was a 15 dioptre exotropia in the right eye. Confrontation visual fields were intact in both eyes. Slit-lamp examination was remarkable for a narrow band of corneal thinning inferiorly in the right eye with central bulging above the thinned zone and an area of normal thickness between the thinned section and the limbus (Fig 1). Faint vertical stromal striae were present in the right eye. No iron lines, lipid deposition, or vascularisation were present in the cornea in the right eye. The left cornea appeared normal, without any trace of iron line, striae,

Table 1 Pathology and response to treatment with subconjunctival interferon alfa-2b in each patient with ocular cicatricial pemphigoid (OCP)

Patient No	Age, sex	Conjunctival biopsy			OCP scores		Duration of follow up (months)
		Squamous metaplasia	Lymphocytic infiltration	Ig binding	Pre T, C	Post T, C	
1	84, F	Yes	Yes	No	22, 14	16, 14	30
2	89, M	Yes	Yes	No	16, 14	16, 14	6
3	77, M	Yes	Yes	Yes	18, 16	18, 14	30
4	65, F	Yes	Yes	Yes	18, 16	12, 12	23
5	63, F	Yes	Yes	Yes	18, 14	16, 11	10

Ig = immunoglobulin; T = treated eye; C = control eye.