Resident’s Corner
Ocular Cicatricial Pemphigoid
Stephenie Parker, O.D.

Ocular cicatricial pemphigoid (OCP) is a progressive autoimmune disorder that results in ulceration and scarring of mucous membranes. It is thought to result from an underlying genetic predisposition as well as an environmental “trigger” that leads to an immune attack on mucosal tissue. Without therapy, the disease results in keratinization of the ocular surface and dramatic scarring of the palpebral and bulbar conjunctiva over months to years¹.

Early disease is difficult to detect due to the fact that symptoms mirror those of many common and mild ocular complaints. This makes early diagnosis a significant challenge. In cases where OCP is suspected, the presence of mouth ulcers can be helpful, as this is present in early disease. Typical ocular signs include shrinkage of the conjunctiva, fornix narrowing, and the presence of symblepharon.

A conjunctival biopsy with direct immunofluorescence testing for IgG, IgM and C3 is the gold standard for diagnosis. The basement membrane of the conjunctival epithelium will show a linear pattern when positively stained¹.

Long term use of steroid-sparing immunomodulators are the main therapeutics used. While effective at controlling inflammation, oral corticosteroids are not typically used as symptoms recur once they are tapered. In terms of therapy specific to the ocular surface, aggressive lubrication is most important. Non-preserved artificial tears and ointments are preferred, as well as autologous serum if necessary¹. Bandage and scleral contact lenses have also shown to help in both patient comfort and maintaining the ocular surface to some degree². To reduce inflammation and increase tear production, topical corticosteroids, cyclosporine, and oral doxycycline can be used. In late stage OCP, amniotic membrane transplantation has shown some promise in delaying disease progression³. However, in severe, late-stage disease, the Boston keratoprosthesis may be the only way to preserve and restore vision¹,⁴.

Reference:

Dr. Parker is a graduate of the Illinois College of Optometry. She is presently the cornea and contact lens resident at Illinois College of Optometry. Her clinical interests include specialty contact lens fittings and the management of ocular surface disorders.

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