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Bilateral keratoprostheses in a patient with ocular mucous membrane pemphigoid

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Abstract

Mucous membrane pemphigoid (MMP) with ocular involvement, also referred to as ocular cicatricial pemphigoid, is a rare, autoimmune blistering disease that can have devastating effects for patients. The resulting corneal neovascularization, recurrent corneal abrasions, and ulceration can ultimately result in severe vision loss. We present a 64-year-old man with recalcitrant ocular MMP and consequently advanced corneal involvement that received bilateral keratoprostheses. This case highlights an aggressive but successful placement of bilateral keratoprostheses in recalcitrant ocular MMP, which restored his vision and ultimately his quality of life.

Keywords: mucous membrane pemphigoid, ocular cicatricial pemphigoid, keratoprosthesis, K-Pro lens

Introduction

Mucous membrane pemphigoid (MMP) with ocular involvement, also referred to as ocular cicatricial pemphigoid (OCP), is a rare, autoimmune blistering disease that can ultimately result in severe vision loss. We present a particular patient with recalcitrant ocular MMP who received bilateral keratoprostheses, which helped to restore his vision and ultimately his quality of life.

Case Synopsis

A 64-year-old man developed mild symptoms of blurry vision and dry eyes in 2000. He underwent uneventful cataract surgery in 2008, which improved his vision somewhat, but he was subsequently noted to have bilateral symblepharon and conjunctivalization of his corneas, limiting both his eye movement and his vision. He began treatment for suspected ocular MMP with dapsone in 2012 and was ultimately diagnosed with MMP by an ophthalmology consultant based on conjunctival biopsy in 2014. Indirect immunofluorescence for IgG autoantibodies against the basement membrane zone and ELISA for anti-BP180, anti-BP230, and anti-type VII collagen autoantibodies were negative. Unfortunately, at that point he had already developed advanced corneal involvement; therefore, he underwent anterior segment reconstruction of the right eye with PROKERA® ring placement in 2014. He aggressively scarred over the PROKERA® ring, which ultimately had to be dissected out one week later. The following year he received a corneal transplant with keratoprosthesis (K-pro lens) placement for corneal involvement of the left eye, which improved his vision to 20/40 at only 32 days postoperatively. He was referred to the dermatology department for further medical management and achieved stabilization with



Figure 1. Successful placement of bilateral keratoprotheses in a patient with recalcitrant ocular cicatricial pemphigoid (OCP), restoring his vision (OS [oculus sinister] 20/40, OD [oculus dexter] 20/60) and ultimately his quality of life

mycophenolate mofetil 1gm daily, one round of rituximab infusions following the rheumatoid arthritis protocol (1gm repeated in two weeks), and tacrolimus eye drops. In 2018, a K-pro lens was placed in the right eye resulting in improved vision, from 20/500 to 20/60 (**Figure 1**).

He is currently maintained on mycophenolate mofetil 1gm daily as well as prednisolone, moxifloxacin, latanoprost, and tacrolimus eye drops. Since his last visit he developed glaucoma of the left eye, a known complication following K-pro lens placement [1]. This has caused a decrease in vision in his left eye but he currently maintains 20/60 vision in the right eye.

Case Discussion

Known target antigens for MMP include BP180, BP230, laminin 332, laminin 6, collagen VII, and integrin β 4, α 6 [2]. Previous reports suggest integrin beta 4 antibodies are more specific for

OCP, although these were not assessed in this case [3]. Use of combined serological testing may be appropriate when diagnosing MMP [4].

Conclusion

MMP with ocular involvement is a treatment challenge with potential for scarring and blindness. There are no available therapeutic options to reverse scarring and fibrosis once formed. Therefore, early detection and treatment is essential to prevent devastating complications [5]. This case highlights an aggressive but successful placement of bilateral keratoprotheses in a patient with advanced, recalcitrant ocular MMP, which restored his vision and ultimately his quality of life.

Potential conflicts of interest

The authors declare no conflicts of interests.

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