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Introduction

Ocular cicatricial pemphigoid (OCP) is an autoimmune ocular disease that can cause severe dry eye syndrome, conjunctival scarring, symblepharon formation, trichiasis, and corneal involvement.¹⁻² In advanced stages of the disease, keratinization of the cornea may occur.² Scarring of the conjunctiva and cornea can damage the corneal nerve endings, leading to the development of neurotrophic keratitis.³ Neurotrophic keratitis is an ocular disease characterized by a reduction or absence of corneal sensitivity, which may lead to epithelial defects, corneal stromal ulceration, and eventually corneal perforation.⁴ Scleral lenses are an effective treatment option for ocular cicatricial pemphigoid and neurotrophic keratitis, providing therapeutic, protective, and refractive benefits.

Case Description

A 70-year-old female presented with matted shut eyelashes due to severe ocular dryness and discomfort leading to obstruction of peripheral vision. The patient also has unknown systemic conditions that caused open wounds on her skin and around her eyes. The patient had previously sought care from six different specialties, including ophthalmology and rheumatology, none of which could sufficiently address and manage her ocular symptoms. Slit lamp examination revealed 4+ diffuse superficial punctate keratitis (SPK) in the right eye and 3+ diffuse SPK in the left eye, causing irregular astigmatism and reduced best corrected visual acuity (BCVA). The patient also had corneal stromal scarring in the visual axis of her left eye, further contributing to reduced vision. The patient's BCVA with spectacles was reduced at 20/40 in the right eye and 20/60 in the left eye. Due to the patient's extensive list of allergies and negative side effects to medications, she was unable to pursue many prescription topical or oral medications used to manage dry eyes. Additionally, the patient suffered from ocular cicatricial pemphigoid, neurotrophic keratitis, and recurrent trichiasis. The patient is currently being co-managed by optometry, cornea ophthalmology, and rheumatology.

Entering BCVA (Spectacles)		
	OD	OS
Power	-0.50-1.25x070 ADD +2.50	+0.25-2.75x160 ADD +2.50
DVA	20/40, NI PH	20/60, NI PH

Scleral Lens Fitting Process



Figure 1. Patient unable to keep eyes open without scleral lenses due to pain, dryness, and burning sensation.

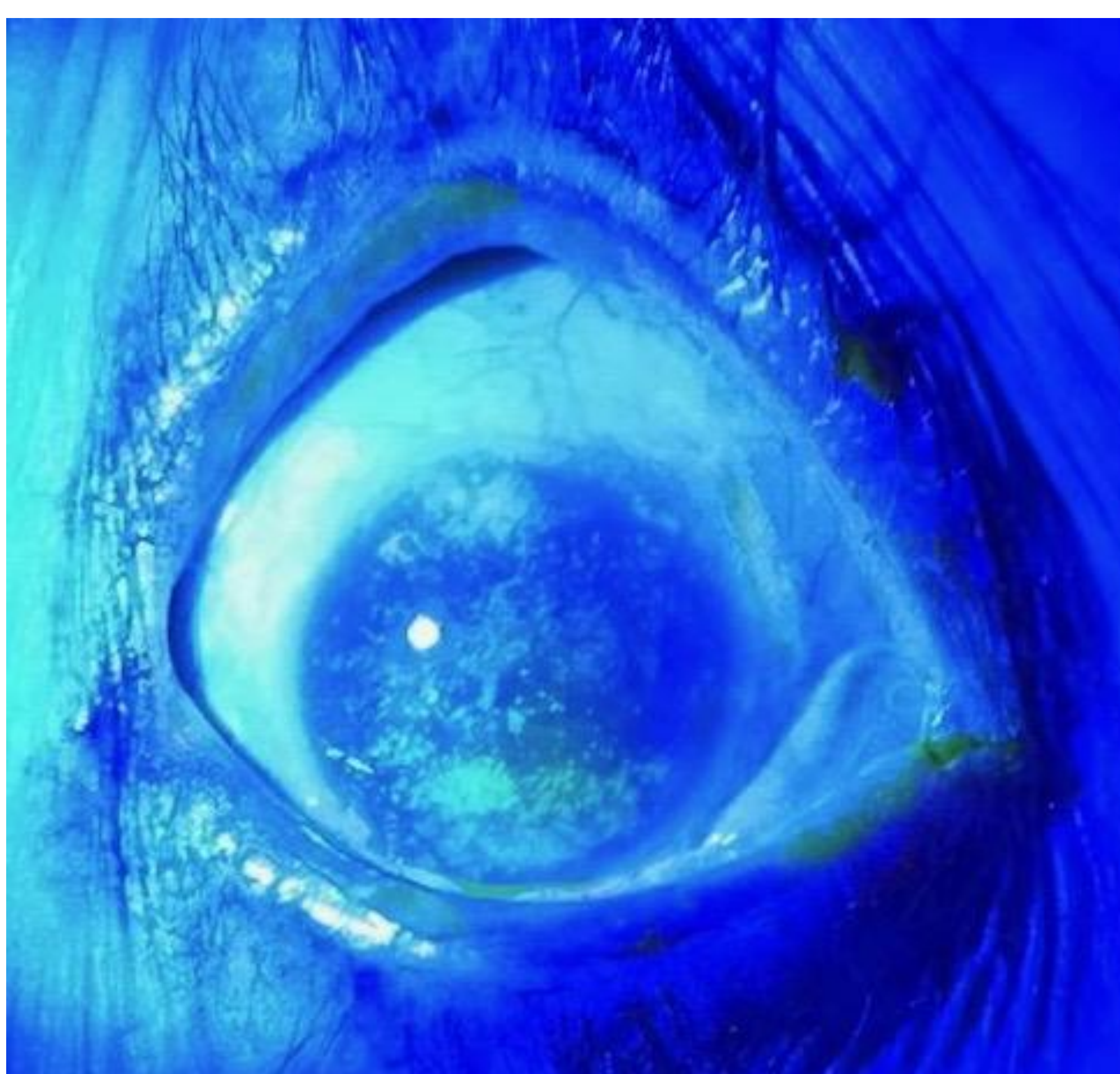


Figure 2. 4+ diffuse SPK OD prior to scleral lens fit.

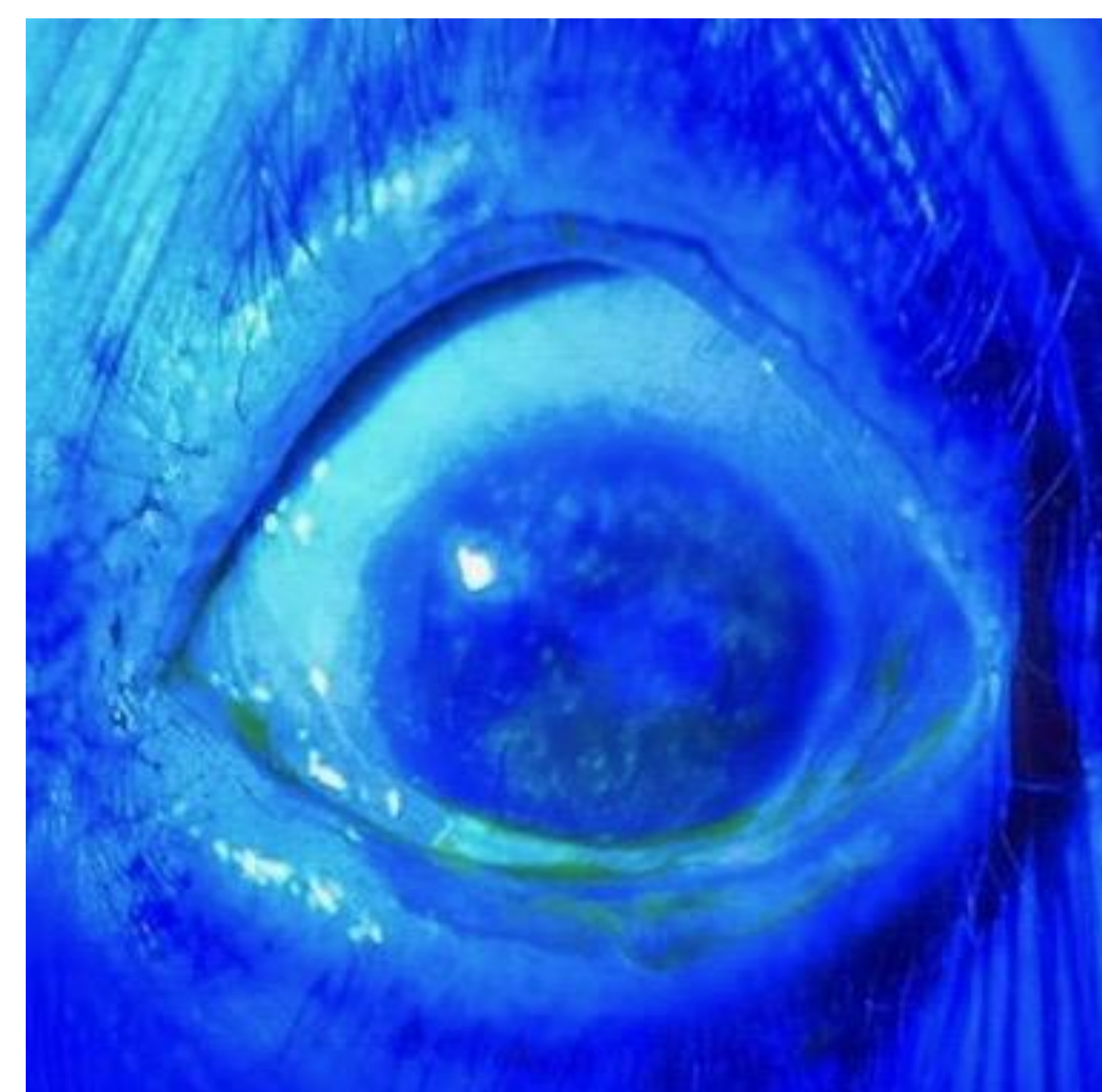


Figure 3. 3+ diffuse SPK OS prior to scleral lens fit.

Finalized Scleral Lens Parameters		
	OD	OS
BC	8.0	8.0
Diameter	15.2	14.9
Power	+0.62	-0.25
Edge	std/steep1	std/steep1
DVA	20/20-1	20/20-1

Discussion

A scleral lens trial provided immediate relief of the patient's ocular signs and symptoms and she was fit in scleral lenses for both eyes. Scleral lenses allowed the patient to keep her eyes open comfortably and protected her neurotrophic cornea from trichiasis. Furthermore, scleral lenses improved the patient's BCVA from 20/40 in the right eye and 20/60 in the left eye to 20/20-1 in both eyes. Ocular cicatricial pemphigoid frequently requires both topical and systemic therapy to maintain ocular integrity.⁵ However, even with immunosuppressive therapy, some patients may still experience severe ocular surface dryness, trichiasis, conjunctival scarring, corneal degradation, and the development of neurotrophic keratitis.^{1-3,5} Neurotrophic keratitis is an ocular disease characterized by a reduction or absence of corneal sensitivity.⁴ Scleral lenses are an effective treatment option for OCP and neurotrophic keratitis, providing therapeutic, protective, and refractive benefits. Scleral lenses provide a post-lens fluid reservoir that constantly lubricates the corneal surface,⁵ thereby alleviating symptoms of ocular surface dryness. Scleral lenses also vault over the cornea and serve as a protective barrier from neurotrophic keratitis and trichiasis.⁵

Conclusions

Ocular cicatricial pemphigoid and neurotrophic keratitis can cause ocular surface dryness, corneal degradation, and vision impairment. Scleral lenses are essential in the management of patients with these conditions and can significantly improve patients' quality of life. This case demonstrates the therapeutic, protective, and refractive benefits that scleral lenses can provide. The patient in this case report is co-managed by cornea ophthalmology and rheumatology so that she can receive both topical and systemic treatment for her conditions. The role of co-management and interprofessional collaboration for ocular cicatricial pemphigoid and neurotrophic keratitis is critical in the management of these diseases and the relief of ocular surface signs and symptoms for these patients.

References

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