Severe Ocular Surface Disease Propagated by Incomplete Blinking in a Patient with Lamellar Ichthyosis

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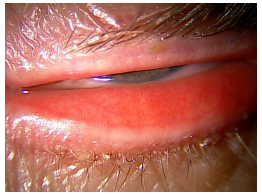
Background

The eyelid margins are responsible for the spread of a thin, stable, and robust tear film across the ocular surface. The motion of the blink and specifically the precise apposition of the upper and lower eyelids is essential for the re-formation of the tear film to ensure an optically clear tissue-air interface.

As described by TFOS DEWS II, tear film instability, further exacerbated by external debris, invokes an immune response within the ocular surface[1]. When these events become chronic in nature, the condition of prolonged reparative processes remains at the ocular surface, leading to sustained inflammation and the pathologies associated with dry eye disease.

The dynamics of the blink mechanism and its role in propagating ocular surface disease has been well-established. During a blink cycle, the meibomian glands are stimulated to release oil and the upper eyelids spread the tear film over the surface of the cornea[3]. It is now known that complete blinks maximize the distribution of tarsal goblet cell mucin and increases the thickness of the lipid layer of the tear film.[2]



Ichthyosis refers to a group of rare heterogeneous dermatological conditions characterized by hyperkeratosis, blistering, and scale formation[4]. Due to the tautness of the skin, individuals afflicted with this condition are often unable to close their eyes fully. The most common ocular manifestation of ichthyosis is cicatricial ectropion and exposure keratopathy.

Case Description

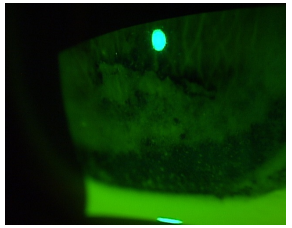


Figure A. Diffuse corneal staining with punctate epithelial erosions and inferior exposure keratopathy

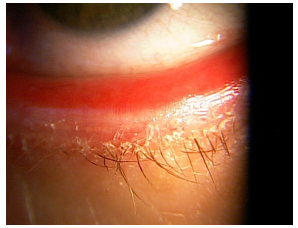


Figure B. Prominent lower lid telangectasia, biofilm, collarettes and seborrheic lash debris

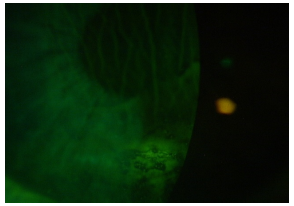


Figure C. Visually non-significant stromal striations and epithelial defect surrounded by heaped basement membrane around margins of the defect

A 39 year old female presented to our dry eye clinic with complaints of dryness, light sensitivity, watering, and 8/10 pain that was progressively getting worse over the past 3 years. Her ocular history was significant for cicatricial ectropion, ulcerative blepharitis and pseudotumor cerebri. She noted that her eyes never closed completely and reported that she had been diagnosed with lamellar ichthyosis. When she presented to our clinic she was taking Restasis twice a day, using preservative-free artificial tears 6-8 times a day and using Aquaphor® and Amlactin® cream around her eyelids.

Table 1. Clinical Findings

Testing	OD	OS
BCVA with spec rx	20/30	20/40
Pupils	PERRL (-) APD	
Extraocular motility	Full and smooth	
Confrontation visual fields	Full to finger counting	
TBUT	4 seconds	4 seconds
TearLab Osmolarity	336	341
Lid margins	3+ demodex and seborrheic blepharitis,3+ telangectatic vessels, 3+ biofilm, (+) lid wiper epitheliopathy OU	
Meibomian gland expression	Turbid expression of nasal glands	No expression possible
Corneal staining	3+ diffuse SPK	3+ diffuse SPK with inferior epithelial erosions
Conjunctival staining	2+ nasal, 2+ temporal	2+ nasal, 2+ temporal
Korb-Blackie light testing	Incomplete lid closure	Incomplete lid closure
Anterior chamber	Deep and quiet	Deep and quiet
Other anterior segment findings:	Visually non-significant stromal striations	Visually non-significant stromal striations
GAT	14 mmHg	13 mmHg
A dilated fundus exam was not performed at this visit.		

Management

We recommended the patient have a microblepharoexfoliation treatment first to clean her eyelash and eyelid margins. An eyelid debridement was performed in office to remove excess keratin and biofilm. Then, the patient was fit with a PROKERA Slim® to heal the cornea in both eyes. The eye was taped shut as much as possible. A thermal pulsation procedure was not recommended at this time due to the patient's widespread gland loss. It was not possible to image her upper eyelids. Warm compresses, copious artificial tears and Lotemax® ointment at night were recommended for at home therapy. The patient will be fit with a scleral lens in the future to protect the cornea.

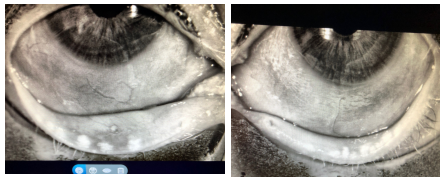


Figure D. Significant meibomian gland dilation and truncation with total gland loss nasally in both eyes

Follow-up

After she had completed her microblepharoexfoliation treatment and PROKERA® in both eyes, she reported that her eyes felt much better. Her BCVA had improved to OD: 20/20 and OS 20/15. She was educated on the importance of eyelid hygiene and was given blink exercises. At a future visit she will be fit with small diameter scleral lenses. She was recommended Ocunox® Vitamin A ointment to use at night to seal her eyelids and improve goblet cell deficiency.

Conclusions

Incomplete lid closure gives rise to severe ocular surface disease. As described by Korb et al., the blink mechanism is responsible for the quality of the tear film and when anatomical restrictions such as ectropion prevent complete blinks, it begets an inflammatory cascade of meibomian gland dysfunction, demodex mite proliferation, biofilm development, and exposure keratopathy. Though this case is associated with a specific dermatological condition, we aim to highlight the importance of productive blinking in controlling the progression of ocular surface disease.

References

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