Undiagnosed Limbal Stem Cell Deficiency Contributes to Multiple Contact Lens Failures

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BACKGROUND

Limbal stem cell deficiency (LSCD) is a condition where the cells responsible for regenerating the corneal epithelium are missing or malfunctioning. This results in chronic corneal inflammation, corneal conjunctivalization, and neovascularization, which leads to patient discomfort and chief complaints of pain, red eyes, photophobia, and vision loss[1,3]. LSCD can be genetic, with associations to aniridia for example, or acquired, possibly from inflammatory conditions or even iatrogenic, such as a result of corneal cross linking.

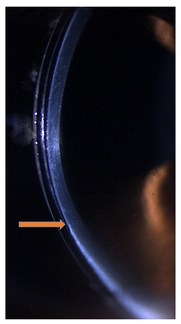
CASE

A 30-year-old female was referred for contact lens fitting due to rapidly progressive keratoconus despite undergoing collagen cross linking procedure in both eyes. Ocular history was positive for chronic redness and severe dryness. Prior to being referred to our clinic, the patient failed with multiple types of contact lenses including soft keratoconic designs, corneal gas permeable lenses, hybrid lenses, and even scleral lenses. The patient presented with bilateral complaints of blurry vision and lens discomfort. The patient was also experiencing severe dryness which was being treated with Xiidra eye drops twice a day and bilateral punctal plugs. In addition, she reported focal redness inferior nasal in the right eye that started few days prior.

The patient was able to tolerate her habitual lenses of unknown parameters on average of 3-4 hours a day due to fogging, dryness, and subjective tiredness. Patient reported good compliance with Boston Simplus care system and filling lenses with Addipak prior to insertion. Assessment revealed a poor fitting lens OD with good vision and the opposite on the left where the OS lens was fitting well but provided poor vision (Table 1, Figure 1 and Figure 2).

	OD	OS
VA	20/20 -1 PHNI	20/100-2 PH 20/25-2
Central	Applanation over cone	1: 1/2
Paracentral	1: ¾	1:1
Mid-peripheral	1: 1/3 with nasal touch	1: ½ nasal, 1:¼ temporal
Limbal	Clearance with nasal touch	Clearance
Edge	Alignment with nasal impingement	Alignment
Other	Scratched lens with deposits in TF	Scratched lens with deposits in TF
Pt comments	**Good vision, uncomfortable**	**Poor vision, comfortable**

Table 1. Habitual scleral lens fit. Parameters of the lens are unknown.



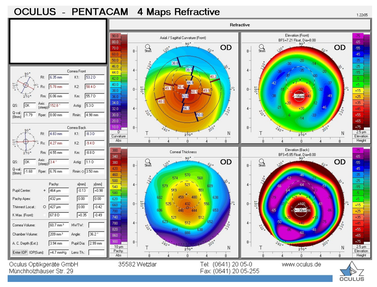


Figure 1. Habitual OD lens seen in optic section. Notice the area of touch inferior to the pupil correlates with her corneal topography.

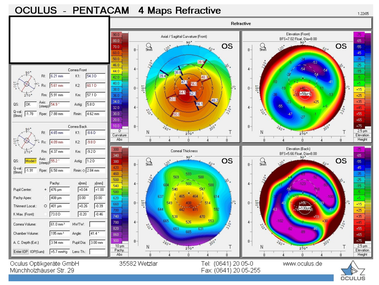


Figure 2. Habitual OS lens seen in optic section. Notice the area of thinned clearance correlated with the corneal topography.

Slit lamp exam showed scleral lens impingement on a nasal fluid filled conjunctival cyst OD, which resulted in irritation and ocular hyperemia (Figure 3A); removal of the lens showed persistent imprint of lens edge (Figure 3B, C).

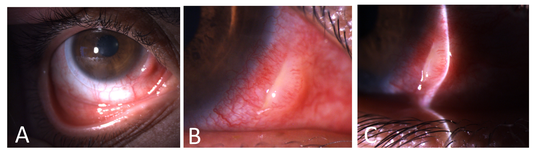


Figure 3. A) OD with lens impingement on nasal conjunctival cyst. B) Nasal conjunctiva remains indented after lens removal. C) Optic section view of clear elevated fluid filled conjunctival cyst.

Staining with sodium fluorescein (NaFl) OD revealed poor surface wettability, an inferior area of pannus along with limbal neovascularization and haze, greatest superior and inferior (Figure 4A, B). Similar findings were seen OS, along with foreign body tracks due to trapped debris under the habitual scleral lens (Figure 4C, D).

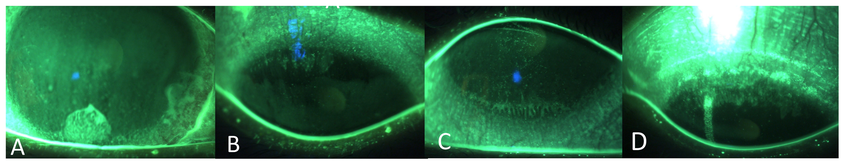


Figure 4. Cornea NaFl staining in different gazes after lens removal. A) OD in primary view. B) OD in down gaze, viewing superior cornea. C) OS in up gaze, viewing inferior cornea. D) OS in down gaze, viewing superior cornea.

ASSESSMENT AND PLAN

The patient was diagnosed with limbal stem cell disease (LSCD) based on biomicroscopy findings and subjective symptoms[3]. She was advised to discontinue habitual contact lens wear due to poor fit OD and poor vision OS and to continue with copious amounts of topical lubrication, including Xiidra eye drops and preservative free artificial tears.

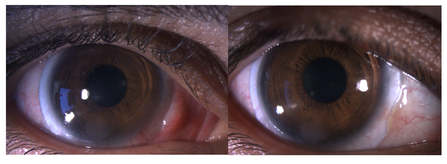


Figure 5. Eye with habitual lens on the left and with newly fit notched scleral lens on the right. Notice the grade 3+ nasal hyperemia and elevated cyst compared to the trace hyperemia with significant cyst improvement seen after a period of discontinued lens wear.

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DISCUSSION

Due to the patient's extensive history of ocular surface disease and contact lens failures likely due to undiagnosed LSCD, there were many considerations for the new lens design (Table 2 and Figure 5).

Consideration	Lens Selected
Hyper dK material	Optimum Infinite (dK: 200)
Lens coating	HydraPeg
Toric/Quadrant Specific Design	OD: quadrant specific OS: toric periphery
Notching or recessing	OD: notch nasal
Care system	ClearCare
Filling solution	Nutrifill

Table 2. Considerations for the new scleral lens design.

Patient was successfully refit into new scleral lenses with improved fit and vision. Afte two weeks post new lens dispense, there was significant improvement in patient's symptoms and corneal health. However, as expected, signs of LSCD persist.

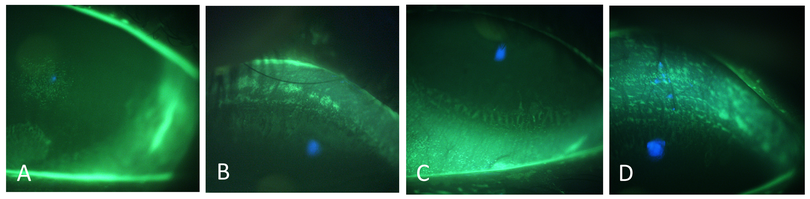

Figure 5. Cornea staining two weeks after new lens dispense. A) OD in primary view. B) OD in down gaze, viewing superior cornea. C) OS in up gaze, viewing inferior cornea. D) OS in down gaze, viewing superior cornea.

LSCD can be hereditary, acquired, or idiopathic with common triggers such as poor fitting contact lenses[4], ocular surgeries involving the limbus, and even radiation. In the case of our patient, undiagnosed LSCD caused poor vision, comfort, and failures with multiple lens types. In fact, the poorly fit habitual contact lenses may have contributed and even acerbated underlying LSCD.

General LSCD signs seen on slit lamp exam include centrally extending hazy epithelium that first starts in the superior limbus, punctate epithelial staining, tear film dysfunction due to goblet cell loss, neovascularization, persistent epithelial defects, fibrovascular pannus, keratinization, and ultimately conjunctivalization of the cornea[3]. Associated symptoms include photophobia and pain, along with irritation, blurry vision, and contact lens intolerance being the most common[2].

Treatment is aimed towards optimization of the ocular surface as quickly as possible by utilizing topical medications, punctal occlusion, and even scleral lens[5]. In fact, early treatment can lead to significant reduction of signs and symptoms[4]. If disease progresses, surgical management options such as mechanical epithelial debridement, ipsilateral limbal translocation, conjunctival limbal autograft, or Keratoprosthesis can be considered.

CONCLUSIONS

Limbal stem cell deficiency leads to ocular surface disease and other complications that make it difficult to achieve a successful contact lens fit. While scleral lenses can be great for managing complicated cases of ocular surface disease due to their unmatched customizability, it is important to treat the underlying issue first. Long term management requires extreme care to make sure the underlying problem is not exacerbated and to prevent further issues from arising.