A Case of Simultaneous Keratoconus and Fuch's Dystrophy

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INTRODUCTION

Keratoconus is a corneal dystrophy associated with progressive corneal ectasia and scarring. With no definitive etiology, the corneal ectasia ultimately leads to irregular astigmatism, central anterior scarring, and reduced vision.[1] It is believed that genetics, the environment (eye rubbing, allergies), and the individual's endocrine system all play a role in the onset, progression, and stabilization of keratoconus.[2]
Fuchs dystrophy is a hereditary, progressive disease of the corneal endothelium which results in endothelial cell loss, thickening of Descemet's membrane, corneal edema, and, in late stages, bullous keratopathy.[3] It has a female predilection with an autosomal-dominant inheritance pattern.

CASE

A 38 –year-old Black female with keratoconus presented to the contact lens service complaining of blurry vision, worse in the morning, with her current contact lenses (CLs). She was currently wearing soft CLs as it provided some improvement in visual acuity and had previously failed with corneal gas permeable (GP) lenses due to discomfort. Her last examination was two years prior. Slit lamp exam was remarkable for mild stromal edema with inferior/central thinning; there was also a beaten metal appearance with corneal guttata in both eyes. Dilated fundus examination was unremarkable in both eyes. Various topical medications were started in order to reduce the edema, and corneal GP lenses were trialed to see if improvement in visual acuity could be obtained.

Ocular and Medical History
(+) Keratoconus OU
--Mild corneal thinning OU but no other corneal findings noted then.
--Has been wearing Purevision2 spheres (previously 20/40 OD, 20/50 OS).
*Last medical exam: 2 months prior: denies any medical conditions, medications, or allergies.

Exam Findings

Visual acuity (with soft CLs):	*Manifest Refraction:*
OD: 20/60	OD: -10.50-1.75x080 20/80
OS: 20/200	OS: -13.50-1.25x100 20/100

REFERENCES

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3. Eghrari AO, Riazuddin SA, Gottsch JD. Fuchs Corneal Dystrophy. Prog Mol Biol Transl Sci 2015;134:79-97.
4. Jurkunas U, Azar DT. Potential complications of ocular surgery in patients with coexistent keratoconus and Fuchs' endothelial dystrophy. Ophthalmology 2006; 113: 2187–97.

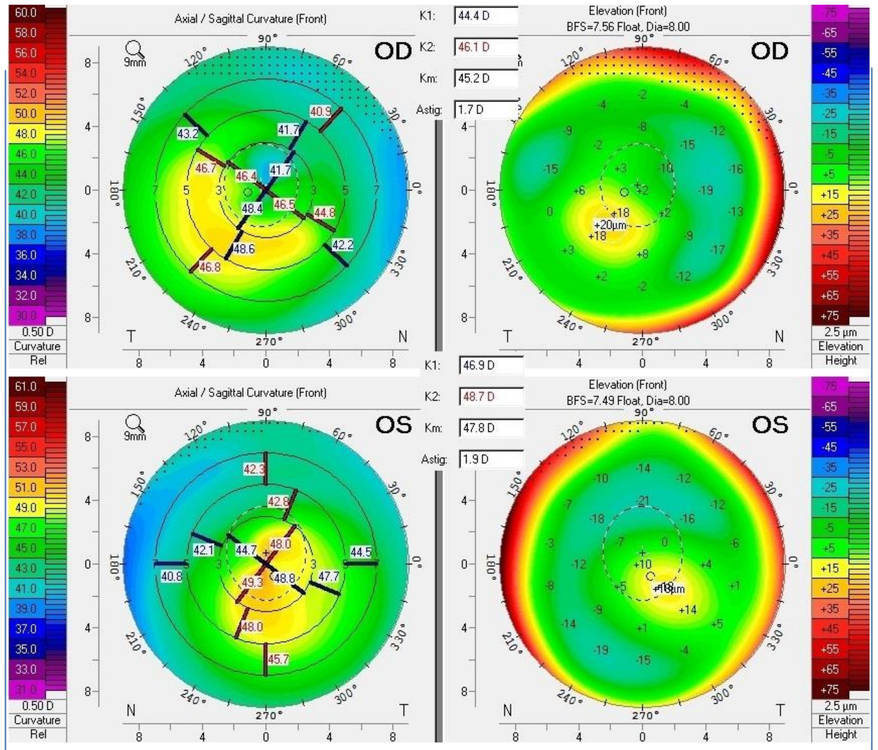

Figure 1. Pentacam topography images

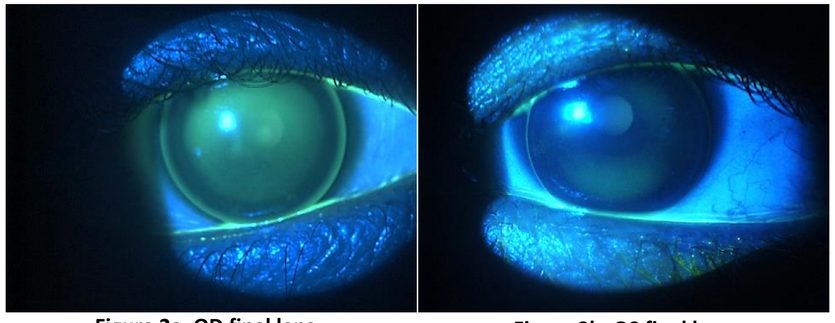

Figure 2a. OD final lens

Figure 2b. OS final lens

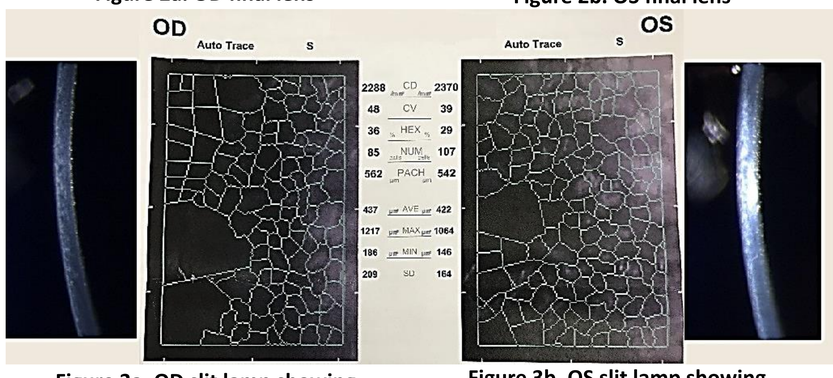

Figure 3a. OD slit lamp showing guttata, with corresponding ECC

Figure 3b. OS slit lamp showing guttata, with corresponding ECC

MANAGEMENT & RESULTS

Visit #1&2: Initial presentation and follow up
Due to presence of guttata and corneal edema (Figures 3), patient was educated to discontinue soft CL wear. Patient was prescribed Muro 128 5% eye drops four-times-a-day in both eyes. She was then examined two weeks later and reports she did not use Muro as it irritated her eyes. She then self-medicated with Systane ointment every night in both eyes with some relief. Was then prescribed FML 0.1% eye drops four-times-a-day in both eyes for two weeks.

Visit #3: Corneal follow up (2 weeks later).
Patient noted that FML provided relief to her discomfort, and there was an objective reduction in the stromal edema as well. At this point, medication was discontinued and Pentacam was performed (Figure 1); diagnostic corneal GP trials were used in-office.
OU: 9.6 diam, 7.3 BC, 8.40 OZ, -3.00DS, PC1: 8.8/0.4, PC2: 10.8/0.2
Fit OU: interpalpebral, centered, 0.50mm movement, central feathery touch, paracentral alignment, midperipheral touch, minimal edge lift.
Over-refraction: OD -5.50sph VA: 20/30, OS -8.50sph VA: 20/40
New lenses ordered with modified peripheral curves and over-refraction.

Visit #4: Dispensing appointment (2 weeks later).
Cornea remained stable. Conforma GP SPH lenses ordered trialed:
OD: 9.6 diam, 7.3 BC, 8.20 OZ, -8.25DS, PC1: 8.3/0.2, PC2: 10.3/0.2, PC3: 12.3/0.3 (heavy blend, lenticularized).
*Fit: adequate (see figure 2a); VA: 20/30, OR NI
OS: 9.6 diam, 7.3 BC, 8.20 OZ, -10.75DS, PC1: 8.3/0.2, PC2: 10.3/0.2, PC3: 12.3/0.3 (heavy blend, lenticularized).
*Fit: adequate (see figure 2b); VA: 20/40, OR NI
Note: minimal to adequate edge lift but corneal health not compromised.

Visit #5: Final appointment (CL wearing time: 6 hours)
Vision and corneal health (with and without CLs) remained stable, and contact lenses showed similar fitting pattern as during the dispense visit. CL Rx was finalized at this visit. A return visit was scheduled for 3 months.

CONCLUSION

This case demonstrates that two corneal diseases can occur at the same time, and specialty contact lenses should be used to determine best corrected visual acuity. Disease severity, and even detection, may be underestimated due to the fact that the corneal thinning caused by keratoconus, and a concurrent increase in corneal thickness caused by Fuchs dystrophy, may combine to falsely normalize pachymetry readings.[4] Close management is needed to ensure adequate corneal health and stable vision, as vision can fluctuate due to either disease.