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Epithelial Bullous Edema in a Scleral Lens Wearer with Marfan Syndrome

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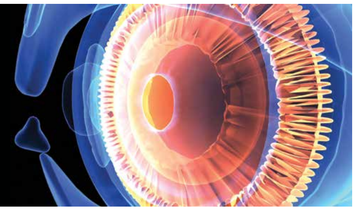

BACKGROUND

Marfan Syndrome (MFS) is a common autosomal dominant disorder that affects the fibrillin gene (FBN1). The glycoprotein fibrillin-1 is encoded by FBN1 and plays a large role in the strength and elasticity of ocular connective tissues.[1] A large spectrum of ocular abnormalities including retinal detachments, increased axial length, high myopia, flat and thin corneas, cilliary hypoplasia, and ectopia lentis are associated with mutated FBN1.[1] Specific to the cornea, fibrillin is predominantly localized to the epithelial basement membrane (BM).[2] A main function of the BM is to anchor epithelium to stroma. In normal eyes, microfibrils containing fibrillin-1 are thought to act as a flexible mechanical anchor at the epithelial-mesenchymal BM interface.[2] Abnormal elastic components are produced from the mutated FBN1 gene which lead to disrupted anchoring processes and corneal thinning and flattening.[1] A lack of adhesive extracellular matrix in the BM has been reported to accelerate epithelial bullae formation without significant endothelial cell dysfunction in cases of bullous keratopathy.[3]

CASE DESCRIPTION

- 27-year old African American female with MFS presented for a scleral lens (ScCL) fit follow-up
- CC: mild blur at distance and near, OU. Has been in ScCL for approx. 2 months
- (+)OHx: two retinal detachments s/p laserpexy and scleral buckle OD, aphakia OU, POAG OU
- Presenting exam reveals: multiple areas of coalesced mid-peripheral epithelial bullae without epithelial breaks or observable stromal edema, OU
- Specular microscopy: endothelial cell count of 3135/mm2 OD and 3106/mm^2 OS
- Central corneal thickness: 695µm OD and 660µm OS
  - *Thicker values established prior to ScCL wear in 2014 (647µm OD and 631µm OS)*

ZenLens Oblate*	Material	Power	CT	Reservoir Depth	Base Curve	Diameter	BCVA
OD	Boston XO2	+7.25DS	480µm	140µm	8.94mm	16.0mm	20/30*, NIPH
OS	Boston XO2	+7.88DS	510µm	189µm	8.94mm	16.0mm	20/25*, NIPH

TABLE 1: *Lens parameters worn by patient. Reservoir depth was measured via iVue anterior segment OCT (Optovue, Freemont, CA), after 8 hours of wear. *Zenlens Oblate (Alden Optical, Lancaster, NY).*

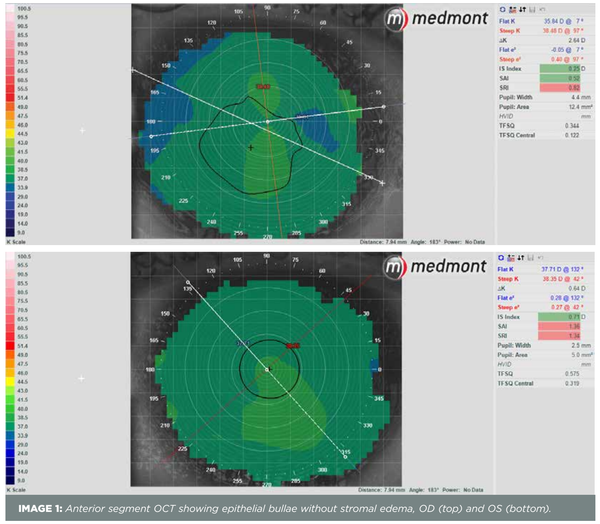

IMAGE 1: *Anterior segment OCT showing epithelial bullae without stromal edema, OD (top) and OS (bottom).*

DIAGNOSIS AND DISCUSSION

This case represents the formation of epithelial bullae with stromal edema in a patient with MFS exposed to hypoxic conditions secondary to ScCL wear. A compromised BM composition from the FBN1 mutation likely resulted in a weakened adhesion of BM to stroma. Despite normal endothelial cell density OU, the patient has a longstanding history of chronic corneal edema. This compounded with hypoxic conditions generated by an excessive ScCL center thickness due to a high hyperopia is thought to be the cause of epithelial bullae formation.

In this case, an oblate design ScCL was selected after considering:

- *Excessively flat corneas creating challenges in obtaining proper fitting relationships in soft, rigid corneal, or hybrid designs in a prolate geometry,*
- *Reduction in oxygen transmissibility (Dk/t) from aphakia,*
- *Need to achieve a well-centered fit to reduce glare from secondary to iris abnormalities.*

Author	Dk	CT	Reservoir Depth
Compañ V et al, 2014	≥ 125	≤ 200µm	≤ 150µm
Michaud L et al, 2012	≥ 150	≤ 250µm	≤ 200µm

TABLE 2: *Guidelines of ScCL parameters to prevent clinically significant corneal edema*[4,5]

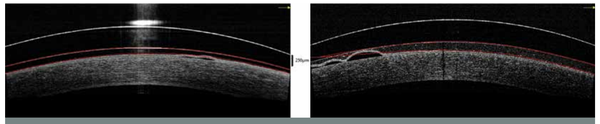

FIGURE 1: *Corneal topography displaying extremely flat corneas of OD 35.84D/38.48D@097 (left) and OS 37.71D/38.35D@042 (right).*

TREATMENT AND RESULTS

- Treatment: 1gtt Muro 128 5% ophthalmic solution QID OU and discontinuation of ScCL wear
- 1-week follow-up: decreased size of epithelial bullae. Muro 128 regimen continued
- 2-week follow-up: disrupted epithelium without staining, no bullae OD, trace bullae OS Muro 128 regimen continued
- A low clearance ScCl or hybrid lens was considered following the resolution of edema

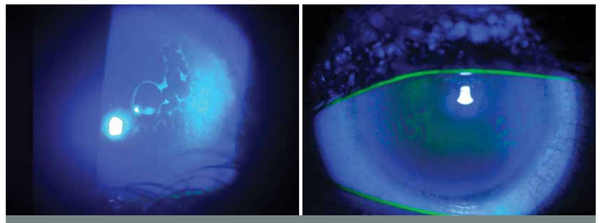

IMAGE 2: *Fluorescein pooling outlining epithelial bullae, OS, at initial presentation (left) and two weeks post Muro 128 treatment (right). Note the decrease in bullae size with surrounding disrupted epithelium.*

CONCLUSION

- Irregular corneas should be monitored closely for adverse effects with ScCL wear
- Patients with compromised corneal integrity should be fit following suggested guidelines regarding proper material Dk, center thickness and central clearance to prevent corneal hypoxia and edema
- Although scleral lenses are currently the most common specialty lens choice, other lens options should always be considered

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