Treatment of Visually-Significant Epithelial Basement Membrane Dystrophy with Scleral Contact Lens Versus Superficial Keratectomy

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Abstract

Epithelial Basement Membrane Dystrophy (EBMD) is an anterior corneal condition in which excessive basement membrane is produced, resulting in irregularity of the overlying epithelium. Affected individuals are typically asymptomatic. However, some patients may experience blur, photophobia, diplopia, or acute ocular pain due to corneal erosion[1]. The following examines various approaches to treatment in a case of visually-significant EBMD.

Case Details

Case Presentation:

37 year old white male

Chief Complaint: progressive blur with spectacle correction, worse OD than OS, with glare and monocular diplopia OD

History of Present Illness:
- He was diagnosed with keratoconus OD 4 months prior by an external eye care provider.
- He was fit with a rigid gas permeable (RGP) contact lens the year prior which he reports improved his vision. However, he was ultimately unsuccessful with lens use due to very poor tolerance to lens sensation.

Ocular History: high myopia OU with spectacle correction

Medical History: unremarkable

Relevant Clinical Findings:

Entering VA (cc – spectacles):
- OD: 20/50-1
- OS: 20/25-1

Lensometry:
- OD: -7.25 -0.75 x 005
- OS: -6.75 -1.00 x 173

Manifest Refraction:
- OD: -6.50 -1.00 x 130 VA 20/30-1
- OS: -5.75 -2.75 x 015 VA 20/20

Biomicroscopy:
- Moderate degree of irregularly-shaped gray regions of anterior corneal haze, with negative sodium fluorescein staining of the borders, involving the visual axis OD>OS

Differential Diagnosis

Primary:
- Epithelial Basement Membrane Dystrophy (EBMD)

Others:
- Meesman Corneal Dystrophy
- Reis-Buckler Corneal Dystrophy

Ancillary Testing

Application of Diagnostic Scleral Contact Lens:

	OD
Lens Model	Onefit 2.0
Base Curve	7.5 mm
Diameter	14.9 mm
Edge	Standard
Power	-3.00 sph
Over-Refraction	-5.50 sph, VA 20/20-2

Figure 1.

Improvement of visual acuity with use of a rigid contact lens indicates that corneal irregularity is the cause of the reduced BCVA obtained from refraction.

Imaging

Corneal Topography:

- OD (Figure 2, left): irregular contour without identifiable pattern
- OS (Figure 2, right): irregular contour without identifiable pattern

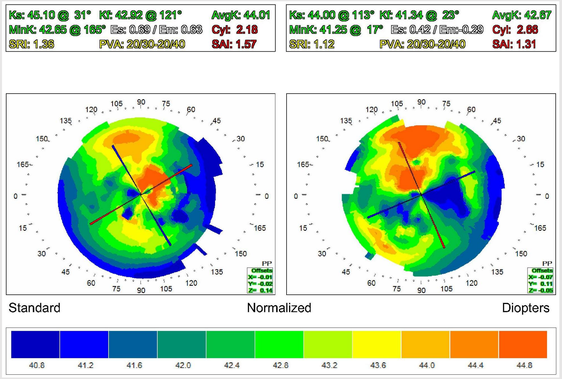

Figure 2.

Corneal Tomography:

- Hyper-reflective, subepithelial lesions, indicating areas of basement membrane thickening, apparent centrally OD in Scheimpflug images

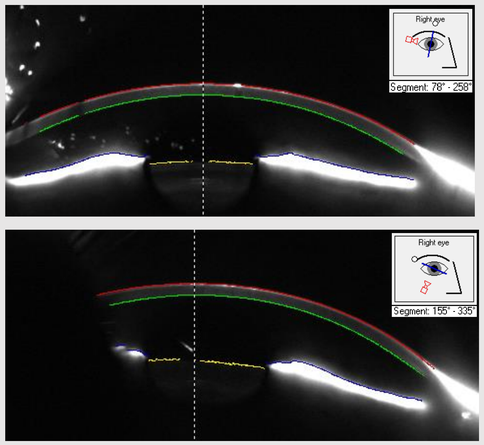

Figure 3.

Diagnosis & Treatment

The diagnosis of EBMD was made based on the clinical appearance of the cornea, which demonstrated characteristic "map" changes found in EBMD[1]. Irregular astigmatism was confirmed by visual acuity improvement with rigid contact lens use and by corneal topography analysis. The previous diagnosis of keratoconus was ruled out using corneal tomography, which did not show the posterior corneal elevation seen in keratoconic corneas[5].

Treatment of EBMD is only indicated in symptomatic patients. Conservative management involves ocular lubrication with artificial tears or punctal occlusion. In more severe cases, corneal debridement or anterior stromal puncture may be effective[1]. Recurrent corneal erosions may result from EBMD and are treated in the acute phase with lubrication and topical antibiotics for infection control. Pain associated with acute erosions can be managed with oral analgesics, topical cycloplegics, or bandage contact lenses[3]. After resolution, the use of topical hypertonic saline solution may be used to aid in the prevention of future recurrences, as they minimize nighttime corneal edema and, thus, reduce friction[1].

In this case, treatment was initiated with a rigid contact lens fitting in order to maximize visual acuity in the setting of irregular corneal astigmatism causing blur and diplopia. A scleral contact lens was chosen due to the potential for good lens comfort and tolerance, given the patient's self-reported history of RGP lens intolerance and failure. Although the patient's left eye was correctable to 20/20 with spectacles, both eyes were fit with contact lenses to avoid anisometropia that would be induced by fitting only the right eye. There was significant difficulty with lens insertion during the fitting process due to patient recoil. He reported severe sensitivity to objects near his eyes.

The patient later presented for dispense of the lenses, with the parameters and fit detailed in Figure 4. After evaluation of the lenses, proposed modifications to the scleral lenses were as follows:
- Flatten base curve by 0.4 mm OU to decrease central clearance;
- Increase edge toricity OU by changing to Flat 1/Steep 1 in order to improve centration and superior limbal clearance;
- Incorporate over-refraction OD.

The patient successfully completed lens insertion and removal training with much difficulty, however he stated that he would prefer surgery to contact lens use if it was a possibility. An intra-departmental referral to a corneal specialist was made and modification of the lenses was deferred pending the consultation.

	OD	OS
Lens Model	Onefit 2.0	Onefit 2.0
Base Curve	8.0 mm	7.8 mm
Diameter	14.9 mm	14.9 mm
Edge	Toric: Standard/Flat 1	Toric: Standard/Flat 1
Other Parameters	Extra Limbal Clearance	Extra Limbal Clearance
Power	-6.62 sph	-3.00 sph
Visual Acuity	20/50	20/20
Over-Refraction	-1.50 sph, VA 20/20	Plano sph
Evaluation of Fit		
Centration	Mild inferior decentration	
Central Clearance	200 um excessive vault	
Limbal Clearance	Adequate N/T/I, none S	
Edges	Aligned 360	
Movement	None	

Figure 4.

The patient was evaluated by the corneal specialist, who proposed superficial keratectomy (SK) as a viable treatment option. Superficial keratectomy is a procedure in which the anterior cornea is manually debrided using a blade to smooth the surface[4]. After discussion of the risks, benefits, and alternatives to SK, the patient elected to proceed with surgery of his right eye. Surgery was performed three weeks later without complication. The patient's right cornea was clear after healing and remained stable over the following two months. The final refractive error of the right eye was -7.00 -3.00 x 176, with a visual acuity of 20/20. The patient reported clear vision and chose to defer surgery of his fellow eye. An updated spectacle prescription was dispensed.

Discussion

Corneal dystrophies encompass a collection of relatively rare, non-inflammatory, heritable disorders in which abnormal substances are deposited in various layers of the cornea. They tend to present in a bilateral, but asymmetric, manner. Corneal dystrophies are sub-categorized by the corneal layers affected. The categories include epithelial/subepithelial, epithelial-stromal, stromal, and endothelial. Dystrophies are in contrast to degenerations, whose etiologies are corneal insult from trauma, infection, age, or other environmental factors[1].

Epithelial basement membrane dystrophy is an epithelial/subepithelial dystrophy and is the most common anterior corneal dystrophy[6]. It is also known as anterior basement membrane dystrophy, map-dot-fingerprint dystrophy, and Cogan's microcystic dystrophy[1]. On slit lamp examination, EBMD is characterized by visible "maps", "fingerprints", and "dots". Map and fingerprint lesions result from an abnormally thick epithelial basement membrane that invaginates into the overlying epithelium in a sheet-like formation. Dot lesions are microcysts that are comprised of either fibrogranular material or degenerative cellular byproducts[1].

EBMD most frequently occurs sporadically. However, some cases demonstrate an autosomal dominant inheritance pattern[2]. It more commonly presents in females greater than 50 years of age[6]. The gene implicated in the development of EBMD is Transforming Grown Factor, Beta Induced (TGFBI). The TGFBI gene is also implicated in most other epithelial corneal dystrophies, including lattice, granular, Reis-Buckler, and Thiel-Behnke corneal dystrophies[1]. The gene encodes the TGFBI protein, whose entire function remains unknown, but does appear to aid in cellular adhesion. It is theorized that a mutation in this gene alters cellular adhesion in a way that allows for extracellular deposition of various substances in the cornea, leading to transparency and refractive index changes. TGFBI is expressed in many tissues throughout the human body. In the human eye, TGFBI is expressed in the cornea, predominantly on epithelial cell surfaces, and in the retinal pigment epithelium[6].

The patient in this case suffered from blur and monocular diplopia as a result of EBMD-induced irregular corneal astigmatism. Rigid contact lens use was optically successful and improved his acuity to 20/20. However, the patient was intolerant to contact lens use due to a phobia of object proximity to his eyes. Thus, he opted for surgical treatment, which was also optically successful. Other reasons that contact lens use might not be ideal for a given patient include poor manual dexterity, financial burden, and inability to adhere to proper lens care and practices.

Conclusion

Epithelial Basement Membrane Dystrophy is a frequently asymptomatic corneal condition. When blur from irregular astigmatism does occur in the setting of EBMD, rigid contact lens correction should be considered as a first-line treatment and as an alternative to surgical intervention. However, treatment plans must be tailored to each individual patient. Contact lens use might not always be the preferred treatment modality for a variety of reasons, despite its less invasive nature.

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