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Case Report

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Brown Mc Lean Syndrome: A Case Report

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Abstract

Purpose: *The aim of this study was to report a case of Brown-McLean syndrome (BMS).*

Method: *A complete ophthalmic evaluation and follow up for a 77 year old patient with BMS.*

Result: *BMS is a rare condition with corneal edema involving the peripheral cornea with orange-brown pigment deposition underlying the edematous area. The edema typically starts inferiorly and advances circumferentially to superior cornea. Central cornea remains clear in most patients. It requires no further treatment and annual follow-ups are adequate.*

Conclusion: *Patients with BMS can rarely progress to corneal decompensation; however, they should be periodically monitored and made aware of early clinical symptoms and signs of ocular surface complications.*

Keywords: Brown-McLean syndrome, Peripheral corneal edema, Marginal corneal edema, Corneal decompensation

Introduction

The syndrome was first described by Brown and McLean in 1969. (BMS) is a clinical condition with peripheral corneal edema observed in patients with long-term aphakia after cataract surgery. It was initially termed as 'Peripheral Corneal Edema After Cataract Extraction' [1]. The corneal edema usually starts after a latent period of several years following surgery, and typically affects the peripheral 2-3 mm, usually starting inferiorly and later extending circumferentially in severe cases. Punctate brownish pigmentation and localised guttae are occasionally found on the corneal endothelium underlying the oedematous area. The conjunctiva remains unaffected and no neovascularization is seen in the affected cornea [2,3]. There may be coexisting predisposing factors such as spontaneous lens resorption, lens subluxation or intermittent angle closure [4,5]. Transillumination shows peripheral iris atrophy and is not the result of liberation of iris pigment because the trabecular meshwork is not unduly pigmented in these cases. Although altered aqueous dynamics, iridodonesis, and movement of other tissues or the intraocular lens have been proposed as etiologies, the cause of the syndrome remains unknown. Confocal microscopy findings of the affected area found normal endothelial counts and endothelial cell morphology [6]. Specular microscopy of the central cornea shows normal counts and morphology of endothelial cells. Most patients with BMS are asymptomatic, but some of them may complain of foreign body sensation or even pain, from ruptured bullae [4]. Most patients require no therapy, however, penetrating keratoplasty and peripheral conjunctival flaps have been used for pain control.

Case History

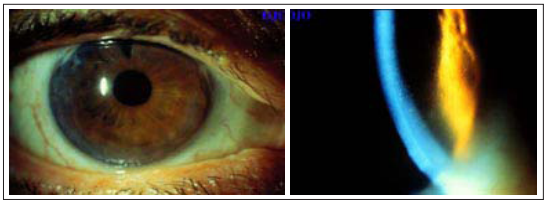
The patient is a 77 yrs old female who came for a follow-up.
POHx: Bilateral intracapsular cataract extraction was done 25 years ago. She is a PMMA contact lens user since 1975.
PMHx: Non-Contributory
SHx: Non-contributory
FHx: Non-contributory

Examination

Vision: OD 20/40, OS 20/30 with contact lenses OU
External exam: Normal
Pupils: Reactive to light, no APD
Motility: Full
Intraocular pressure: OD 14 mm Hg, OS 16 mm Hg

Slit Lamp Examination

OD – The peripheral concentric corneal stromal and epithelial edema extending 2.5-3.5 mm from the limbus, sparing central cornea is seen. Peripheral iridectomy is present superiorly. Aphakia is noted. Anterior chamber is quiet. No keratic precipitate or endothelial pigmentation is seen.



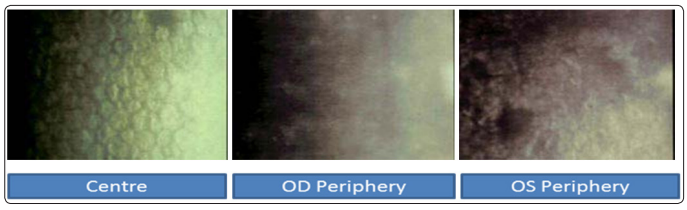
OS – The peripheral concentric corneal stromal and epithelial edema extending 2.0-3.0 mm from the limbus, sparing central cornea is seen. Aphakia is noted. Anterior chamber is quiet. No keratic precipitate or endothelial pigmentation is seen.

Fundus

OD – Normal
OS – Normal

Endothelial Specular Microscopy

Specular microscopy of the center of the cornea is essentially normal whereas peripheral cornea shows polymegathism and decreased number of corneal endothelial cells.

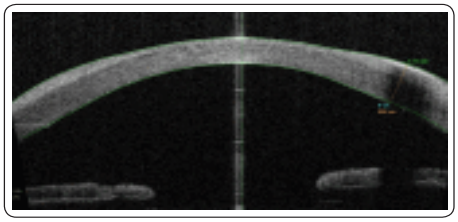


Scheimpflug Imaging

It compared limbus-to-limbus corneal thicknesses. Maximum corneal thickness is 1312 um and the thinnest location is 570 µm located 1.67 mm nasal and 0.89 mm inferior to the corneal apex. The maximal corneal density reached 100% at 4 mm from centre in the right eye as compared to 6 mm in the left eye. The vertical average corneal density was higher along the depth in the right eye than the left eye. The peak in average corneal density was 75% at 0.1 mm from epithelium in the right eye as compared to 48% in the left eye, which corresponds to the anterior stromal layer.

Anterior Segment Optical Coherence Tomography (AS-OCT)

Right eye shows a grey scale image of the cornea. The thickness of the peripheral cornea is 966 µm whereas the central cornea is 556 µm. Hence there is increased peripheral corneal thickness and normal central corneal thickness.



Differential Diagnosis

1. Marginal keratopathy related to collagen vascular disease
2. Fuch's endothelial dystrophy
3. Endothelitis
4. Furrow degeneration
5. Brown-McLean Syndrome

Discussion

Brown-McLean syndrome is a relatively rare clinical condition that involves static peripheral corneal edema, most commonly seen in patients with a significant period of aphakia. The pathophysiology of Brown-McLean syndrome is not fully understood. The normal endothelial cell density and preserved corneal architecture seen on anterior segment optical coherence tomography (AS-OCT) is unlike that seen in cases with post cataract surgery corneal endothelial decompensation. Scheimplflug imaging accurately measures corneal thickness and provides a better assessment than traditional contact-based pachymetry. AS-OCT generates high-quality cross-sectional images of the cornea. There may be a role of Scheimplflug imaging and AS-OCT in objective disease monitoring in cases with Brown-McLean syndrome.

Conclusion

The patients with stable visual acuity over years after cataract surgery require no further treatment. Annual follow-up is adequate. Patients should be educated on symptoms and signs of ocular surface complications. Further ophthalmic surgeries (Anterior stromal puncture, Penetrating keratoplasty, Peripheral conjunctival flaps) can be performed for pain control.

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