CASE REPORT

Brown-McLean syndrome

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SUMMARY

An 80-year-old Chinese man with high myopia and a history of right eye cataract extraction and superior peripheral iridectomy developed peripheral corneal oedema after a period of aphakia. The peripheral corneal oedema was static over years and did not progress to involve central cornea hence his visual acuity remained stable. The condition was compatible with the relatively rare Brown-McLean syndrome. Specular microscopy showed normal endothelial cell density in the oedematous peripheral as well as central cornea. While anterior segment optical coherence tomography demonstrated the cross-sectional architecture of cornea, Scheimpflug imaging was used to measure the peripheral corneal thickening and to demonstrate increased corneal density as compared with the contralateral normal eye. These investigations not only help better characterise the rare disease, but also in precisely monitoring any disease progression by periodic measurements.

BACKGROUND

Brown-McLean syndrome (BMS) is a clinical condition with peripheral corneal oedema observed in patients with long-term aphakia after cataract surgery, especially intracapsular cataract extraction. This clinical condition was initially termed as 'Peripheral Corneal Oedema After Cataract Extraction'.¹ The corneal oedema in Brown-McLean syndrome usually starts after a latent period of several years following surgery, and typically affects the peripheral 2–3 mm cornea, 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 neovascularisation is seen in the affected cornea.^{2–3} Although majority of the patients with BMS have history of previous intraocular surgery, the condition could rarely affect virgin eyes.⁴ These patients usually have coexisting predisposing factors such as spontaneous lens resorption, lens subluxation or intermittent angle closure.^{4–5} BMS was once reported in a patient with myotonic dystrophy who had no history of intraocular surgery performed before.⁶

The condition was described prior to the era of confocal microscopy and was thought to be related to diseased corneal endothelium. However, confocal microscopy was able to identify normal healthy endothelium, both in morphology and counts, in the affected cornea of patients with BMS. Hence the pathophysiology of this disease cannot be solely explained by the endothelial layer.

Earlier reports hypothesised that iridodonesis in aphakic patients could result in intermittent peripheral endothelial abrasion, hence a superior iridectomy is protective of the superior cornea.^{3–5} However Almousa *et al*⁷ and Lim *et al* separately reported two patients who presented with peripheral corneal oedema, most marked superiorly and who had had intracapsular cataract extraction with a superior iridectomy performed. Confocal microscopy findings of the affected area found normal endothelial counts and endothelial cell morphology.⁸ These evidence argue the role of endothelium in the pathophysiology of this syndrome, and there exists a spectrum from healthy to decompensated endothelium among patients with clinical features of BMS.

Most patients with BMS are asymptomatic, but some of them may complain of foreign body sensation or even pain, from ruptured bullae.⁴ Since the central cornea is often unaffected, specular microscopy of the central cornea may find normal counts and morphology of endothelial cells.

CASE PRESENTATION

An 80-year-old Chinese patient presented to our eye clinic in June 1989, with history of right eye cataract removal in 1981 and background high myopia bilaterally. His right eye visual acuity was 20/30 while on contact lens for aphakia, and 20/50 for his left eye with moderate cataract. Corneas were clear bilaterally on slit-lamp examination. A superior peripheral iridectomy was evident in the right eye. Intraocular pressures of both eyes were normal. The patient underwent an uneventful extracapsular cataract extraction and implantation of a posterior chamber intraocular lens for his cataract on the left eye in July 1991. Postoperatively his left eye recovered well from the surgery with a best-corrected visual acuity of 20/20 and a clear cornea.

Peripheral corneal oedema of the right eye was first noticed in December 1994, slightly more than 5 years after the initial cataract operation of the right eye. The oedema involved the peripheral 2–3 mm of stroma and epithelium circumferentially along the corneoscleral limbus, sparing the centre of the cornea (figure 1). The anterior chamber was quiet and no keratic precipitate or endothelial pigmentation was documented. The patient was instructed to discontinue contact lens use briefly, but the condition persisted. The intraocular pressure was within normal limits all along. There was no family history of any corneal disease. The left eye remained normal and the left cornea was clear.

A specular microscopy in January 1999 found bilateral normal endothelial cell counts (right eye

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Figure 1 Slit-lamp photograph showing peripheral corneal oedema with relative sparing of central cornea in Brown-McLean syndrome.

temporal 3545; right eye central 3135, left eye central 1772/ mm^2) with normal hexagonal morphology (figure 2A,B). The patient underwent secondary scleral-fixation intraocular lens implantation in January 2001. The intraocular lens subluxed in 2007 requiring removal and exchange for a second scleral-fixation intraocular lens. His right eye intraocular pressure was briefly elevated after the two operations, but subsequently stabilised with topical antiglaucomatous medication. The peripheral corneal oedema remained clinically stable with a visual acuity of 20/30 since 2007 onwards.

Figure 2 Specular microscopy showing preserved corneal endothelial morphology and density in the right (A) and left eye (B) of the patient with Brown-McLean syndrome in the right eye.

INVESTIGATIONS

Scheimpflug imaging (Pentacam, Oculus Inc) compared limbus-to-limbus corneal thicknesses of both eyes. The maximum corneal thickness measured on Scheimpflug imaging was 1312 microns and the thinnest location is 570 μm located at 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.

An anterior segment optical coherence tomography (AS-OCT) images of the right eye showed a grey scale image of the cornea (figure 3). The thickness of the peripheral cornea is measured up to 966 μm whereas the central corneal thickness was 556 μm .

DIFFERENTIAL DIAGNOSIS

Corneal decompensation is a differential diagnosis for corneal oedema after any intraocular operation. In such cases, the condition is due to progressive loss of endothelial pump function, leading to progressive corneal oedema which often involves the entire cornea eventually. Specular microscopy is likely to show reduced cell density of hexagonal endothelial cells and polymegathism.

TREATMENT

Unlike corneal decompensation, Brown-McLean syndrome is usually clinically static and does not affect the central cornea. Hence the visual axis remains clear and the patient enjoys relatively good visual acuity despite refractive errors induced by peripheral corneal oedema. No definite treatment has been described in the literature, although lubricants such as artificial eye drops may help relieve symptoms of irregular ocular surface. Patient should be educated on symptoms of ocular surface complications such as corneal abrasion.

Further ophthalmic surgery can be performed without worsening of the peripheral corneal oedema as in the present case, provided that special attention is given to minimise any potential endothelial trauma.⁶

OUTCOME AND FOLLOW-UP

The patient has a stable visual acuity over the years after his cataract surgery and requires no further treatment. Annual follow-up is adequate and patient is educated on symptoms of ocular surface complications.

DISCUSSION

Confocal microscopy has been used to study the corneal microstructure in patients with Brown-McLean syndrome. With the advent of novel corneal imaging modalities, such as Scheimpflug imaging and AS-OCT, natural history of this syndrome could be better delineated with quantifiable values and accurate measurements.

Prior to the era of AS-OCT any patients with BMS have been followed up for disease progression by confocal microscopy. The present report is the first description of Scheimpflug imaging and AS-OCT in BMS. With its accuracy in obtaining corneal thickness, Scheimpflug imaging proves to be a useful tool in assessing and monitoring the disease with a relatively simple, quick and easily reproducible scan. AS-OCT, on the other hand, generates a cross-sectional image of the cornea in such cases.

Figure 3 High-resolution anterior segment optical coherence tomography scan of the right eye showing increased corneal thickness in the periphery and normal central corneal thickness.

Learning points

- ▶ Brown-McLean syndrome is a relatively rare clinical condition that involves static peripheral corneal oedema, 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 postcataract surgery corneal endothelial decompensation.
- ▶ Scheimpflug 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 Scheimpflug imaging and AS-OCT in objective disease monitoring in cases with Brown-McLean syndrome.

Contributors ALY was involved in the concept and design of the manuscript. KWK, VJ and ALY were involved in writing of the manuscript and approved the final version to be published.

Competing interests None.

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