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Brown-McLean syndrome secondary keratoconus

Qi Wan[1†], Li Chen[1†], Ran Wei[1], Ying-ping Deng[1*], Ke Ma[1] and Jing Tang[1*]

Abstract

Background Brown-McLean Syndrome is a rare clinical condition typically presenting as localized edema involving a 2–3 mm peripheral area of the cornea, which often has undergone intraocular surgery.

Case report We present a unique case of unilateral Brown-McLean Syndrome (BMS) with concurrent keratoconus in an 18-year-old male patient. The individual sought medical attention due to poor vision in the right eye, which had been unnoticed for a month. Notably, there was no history of ocular surgery or trauma, and the patient did not suffer from any systemic diseases. Clinical examination revealed peripheral corneal edema, extending 2–4 mm from the limbus, exclusively affecting the right eye. Corneal topography indicated the presence of keratoconus, which was an incidental finding during the investigation.

Conclusions This case underscores the importance of considering BMS in the differential diagnosis of peripheral corneal edema even in the absence of ocular surgery or trauma. The potential relationship between BMS and keratoconus is explored, suggesting that long-term edema and degeneration of the peripheral cornea could alter its biomechanical properties, leading to deformation and keratoconus. Thorough clinical evaluation and long-term follow-up are crucial in managing such patients.

Keywords Brown-McLean Syndrome, Peripheral edema, Case report, Keratoconus

Background

The Brown-McLean Syndrome is a rare clinical condition typically presenting as localized edema involving a 2–3 mm peripheral area of the cornea [1]. This syndrome was first described in 1969 by Brown and McLean [2]. Typically, the edema begins inferiorly and spreads peripherally, but the central part of the cornea remains unaffected. In most cases, there are fine brown pigment deposits on the corneal endothelium in the affected area accompanied by a reduction in endothelial cell density [3]. The pathogenesis of Brown-McLean Syndrome has not been fully elucidated, but it may be due to genetic susceptibility and exposure of the eyes to certain specific conditions, such as intraocular surgery, glaucoma, and myotonic dystrophy. It usually takes between 6 and 16 years after surgery for this condition to occur, and there have even been reports of it appearing 34 years post-surgery [4]. These patients often have a history of surgical complications and multiple intraocular surgeries. However, there are a few cases where Brown-McLean Syndrome has occurred in eyes that have not undergone any ophthalmic surgery. For example, Rutzen AR. reported a case of a 50-year-old male with myasthenia gravis who also had Brown-McLean Syndrome, but no predisposing

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factors for peripheral corneal edema were found [5]. Additionally, Jamil AZ and others have described a case of unilateral Brown-McLean Syndrome with bilateral keratoconus, but no history of surgery or trauma, nor any other systemic diseases [6]. This article will report on a case of unilateral Brown-McLean Syndrome with keratoconus without any predisposing factors such as the history of surgery, trauma, and families.

Case report

An 18-year-old Chinese male presented to the ophthalmology outpatient clinic at West China Hospital, Sichuan University after a poor vision in his right eye was discovered during a physical examination one month ago. His condition was insidious with no history of eye redness, eye pain, or increased secretions. There was no history of ocular surgery or trauma. The patient had been myopic since childhood and consistently wore glasses for vision correction with no history of wearing contact lenses. The glasses of the right eye (OD) are myopia −2.00D, astigmatism −1.00D; The glasses of the left eye (OS) are myopia −3.00D, astigmatism −1.50D. His vision had remained stable recently, although he occasionally experienced glare. There was no history of other ocular or systemic diseases. There was no significant family history of eye diseases or other genetic disorders.

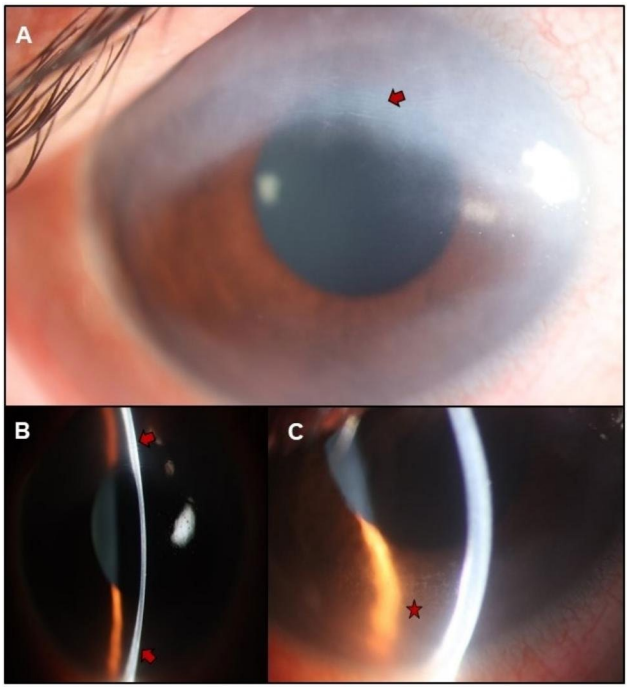

Fig. 1 Digital photograph of the 18-year-old male with Brown-McLean Syndrome. **A**: Photograph of the right cornea showing peripheral corneal edema and central clear cornea. The red arrow shows folds in the descent membrane. **B**: Slit lamp photograph of right cornea. The red arrows reveal the thick corneal stroma. **C**: Slit lamp photograph of right cornea. The red five-star reveals the fine brown pigmentation

Ophthalmic examination revealed that his uncorrected visual acuity was 1.3 (LogMAR) in the right eye and 1 (LogMAR) in the left eye. The best-corrected visual acuity (BCVA) in the right eye was 0.3(LogMAR), with a manifest refraction of -0.5 / -7.00 × 180. The BCVA in the left eye was −0.08 (LogMAR), with a manifest refraction of -4.5 / -1.50 × 180. The corrected intraocular pressure (IOP) was 12.1 mmHg in the right eye and 12.9 mmHg in the left eye. Slit-lamp examination showed no conjunctival hyperemia or edema in the right eye. There was peripheral corneal opacity, with localized edema in the peripheral corneal tissue extending 2–4 mm from the limbus in a 360-degree range. Notable Descemet folds were observed in the superiorly affected area, but the central cornea remained clear (Fig. 1A). Corneal stroma thickening was noted in the affected area (Fig. 1B), and fine brown pigmentation was observed on the endothelium in the inferior affected area (Fig. 1C). The left cornea appeared normal. Both eyes had a central anterior chamber depth of 3.5 CT, with open peripheral anterior chamber angles, normal depth without synechiae, dark brown irises without atrophy or pigment loss, round and equal-sized pupils with prompt light reflexes, clear lenses, and normal vitreous and retinas.

Specular microscopy of the corneal endothelium showed a central endothelial cell density of 2315 cells/mm² in the right eye, with an average cell area of 432 ± 108 μm² and a hexagonal cell proportion of 44% (Fig. 2A). The endothelial cell density in the inferior peripheral cornea of the right eye was 1541 cells/mm², with an average cell area of 649 ± 338 μm² and an unmeasurable hexagonal cell proportion (Fig. 2C). The endothelial cell density in the superior peripheral cornea of the right eye was unmeasurable (Fig. 2D). The central endothelial cell density in the left eye was 3197 cells/mm², with an average cell area of 313 ± 110 μm² and a hexagonal cell proportion of 60% (Fig. 2B). Pentacam HR anterior segment tomography indicated eccentric corneal deformation in the inferior right eye, with a Kmax of 50.2D and a minimum corneal thickness of 542 μm, showing a marked increase in peripheral corneal thickness. The anterior and posterior surface elevations of the cornea in the superior and inferior segments of the right eye were lower relative to the best-fit sphere, while the central surface elevations were significantly increased, suggesting keratoconus in the right eye (Fig. 3A). The left eye had a Kmax of 45.8D and a minimum corneal thickness of 582 μm, with corneal topography indicating a normal morphology (Fig. 3B).Besides, no significant differences were observed in the epithelial mapping images for both eyes (Fig. 4).

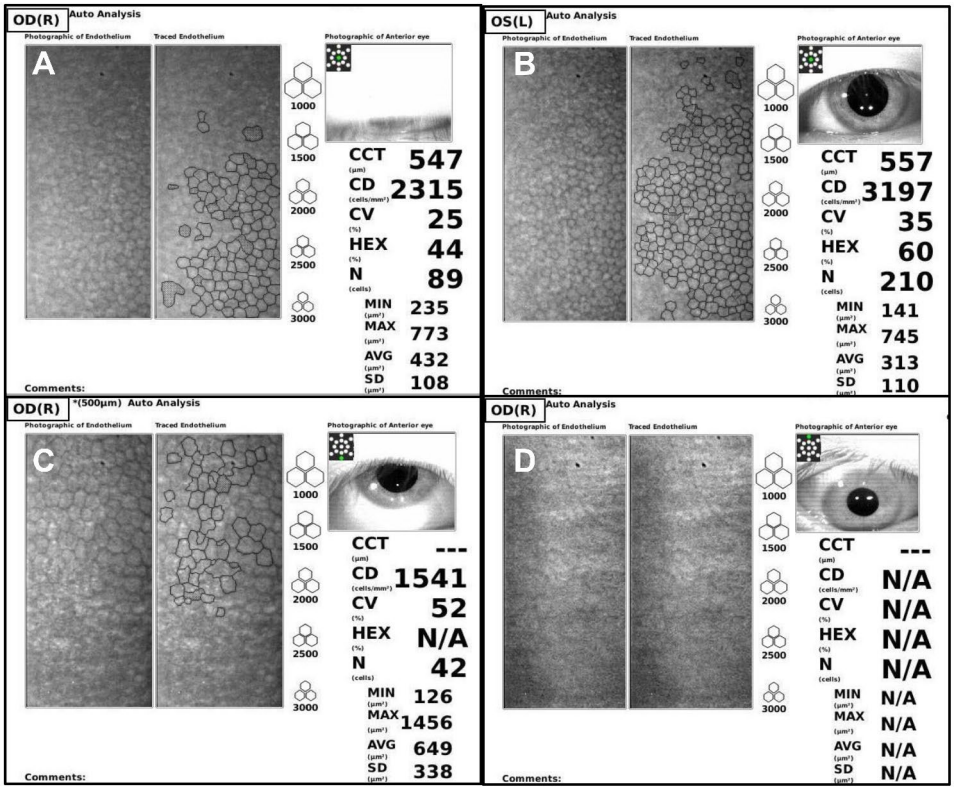

Fig. 2 Non-contact specular microscopy of the corneal endothelium. **A**: The central endothelial cell of the right eye. **B**: The central endothelial cell of the left eye. **C**: The endothelial cell in the inferior peripheral cornea of the right eye. **D**: The endothelial cell in the superior peripheral cornea of the right eye

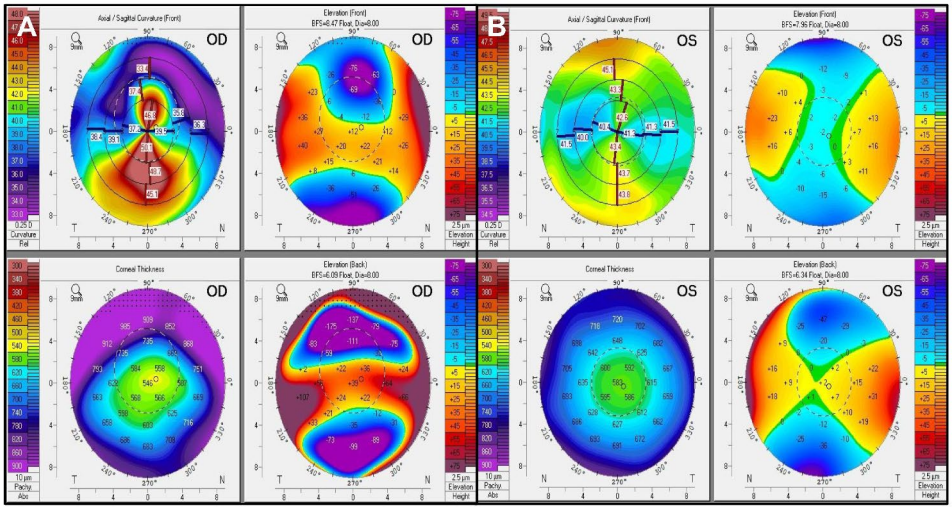

Fig. 3 Pentacam-refractive maps of Case. **A**: The 4 refractive maps of the right eye. **B**: The 4 refractive maps of the left eye

Discussion

Brown-McLean Syndrome is a rare ocular clinical condition, typically characterized by edema in the peripheral 2 to 3 millimeters of the cornea, usually starting inferiorly and spreading peripherally, while the central cornea remains unaffected [7, 8]. Previous literature has mostly reported cases in patients with surgical complications and multiple intraocular surgeries [9, 10]. However, this report details a patient with unilateral Brown-McLean Syndrome without a history of surgery, trauma, and families, suggesting a possible congenital gene mutation or genetic susceptibility. Besides, the patient's personal medical history also lacks apparent triggers. The patient has not experienced any episodes of red eyes, eye pain, or

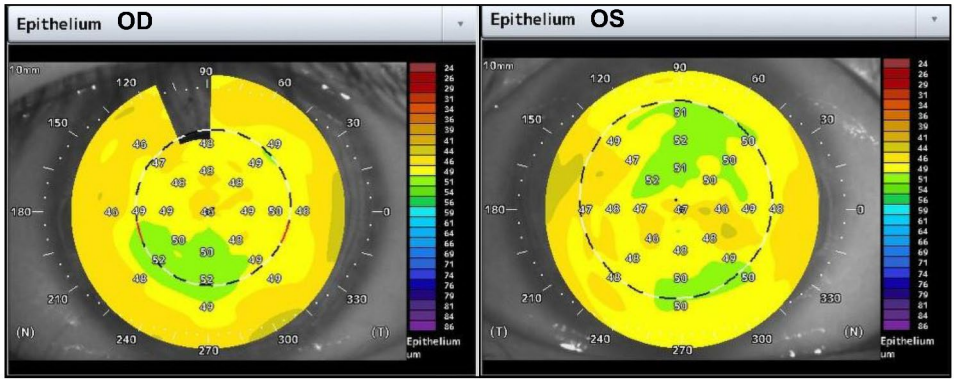

Fig. 4 The epithelial mapping images for both eyes

other ophthalmic conditions, nor does the patient have a history of allergic diseases. This suggests that no known chronic or acute conditions affect the patient's eye health. Additionally, the patient has consistently worn glasses for vision correction but has no history of contact lens wear or physical exercise, which potentially eliminates complications related to corneal contact lens usage and high-intensity exercise [11].

In addition, previous literature has described a case of unilateral Brown-McLean Syndrome combined with bilateral keratoconus [6]. Observing the presence of Brown-McLean Syndrome with keratoconus again in this report suggests a potential causal relationship between the two conditions. It is hypothesized that long-term edema and degeneration of the superior and inferior peripheral cornea could alter the biomechanical properties of the corneal tissue, leading to deformation under the pressure of the upper and lower eyelids, resulting in central protrusion and the formation of keratoconus.

In terms of differential diagnosis, it is important to distinguish this case from Terrien's Marginal Degeneration, corneal endotheliatis, and endotheliopathy. Based on the case's manifestations, Terrien's Marginal Degeneration was ruled out as the patient did not exhibit bilateral peripheral corneal thinning with superficial neovascularization. The localized circumferential edema of the peripheral cornea without any episodes of red eyes, eye pain, or other ophthalmic conditions and anterior chamber reaction also excluded corneal endotheliitis. Furthermore, the patient's central cornea is clear, and specular microscopy shows that the morphology of the corneal endothelial cells is normal. There is no history of systemic disease or infectious diseases, thus ruling out endotheliopathy.

In summary, Brown-McLean Syndrome is a rare clinical condition characterized by edema involving the cornea's peripheral 2–3 mm zone. It may result from a combination of genetic susceptibility and factors such as surgery. Although the patient's central cornea remains clear, allowing for relatively good vision, long-term peripheral corneal edema can lead to refractive errors and potentially secondary keratoconus. This condition requires our heightened attention, and patients should undergo long-term follow-up and observation to ensure appropriate therapeutic measures and management plans are implemented.

Abbreviations

BMS Brown-McLean Syndrome

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Not applicable.

Author contributions

YPD and JT designed the study; QW and LC wrote the initial manuscript; Ran Wei conducted data collection review of the literature. KM revised and edited the paper.

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No funding was received for this study.

Data availability

All the data that substantiates our discoveries can be found within the manuscript.

Declarations

Ethical approval

The ethics committee of West China Hospital of Sichuan University approved the study.

Consent for publication

Written informed consent was obtained from the patient for the publication of this case report.

Competing interests

The authors declare no competing interests.

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