Brown-McLean syndrome in an aphakic patient with homocystinuria: The first reported case in Middle East

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Abstract:

Brown-McLean syndrome not reported in association with homocystinuria and generally occurs in patients with long-term aphakia. Presented as peripheral corneal edema with central clear cornea tends to spare vision without the need of any surgical intervention. The purpose of reporting such a rare condition as a first case of Brown-McLean syndrome associated with systemic disease of homocystinuria. Brown-McLean Syndrome in a patient with homocystinuria generally occurs in patients with long-term aphakia after different modalities of surgical intervention. A high clinical suspicion and regular follow-up is warranted for patients with systemic diseases who are aphakic or present with a subluxated lens for early intervention and better prognosis if needed.

Keywords:

Brown-McLean syndrome, homocystinuria, peripheral corneal edema, subluxated lens

INTRODUCTION

Brown-McLean syndrome is a relatively rare clinical condition characterized by a delayed onset of peripheral corneal edema that typically starts inferiorly and progresses circumferentially, sparing the central cornea.^[1] It is most commonly observed in patients with long-term aphakia after cataract surgery, especially intracapsular cataract extraction.^[1] This syndrome has also been described after extracapsular lens extraction, phacoemulsification, lensectomy, pars plana vitrectomy, anterior chamber intraocular lens implantation, and penetrating keratoplasty. In addition, it has been documented in patients with no previous ocular surgery, including in patients with spontaneous lens resorption with iridodonesis, lens subluxation or luxation, the phakic eye with intermittent angle-closure, keratoconus, and myotonic dystrophy.^[1-5]

Homocystinuria is an autosomal recessive disorder belonging to a group of disorders called inborn errors of metabolism.^[2] It is characterized by a deficiency of the enzyme that converts homocysteine to cystathionine

leading to an abnormal accumulation of homocysteine and its metabolites (homocysteine, homocysteine-cysteine complex, and others) in blood and urine.^[2] Ectopia lentis is the ocular hallmark of homocystinuria due to cystathionine-, 3-synthetase deficiency. Lens zonules normally have a high cysteine content, and its deficiency may affect normal zonular development, thereby predisposing to myopia and lens dislocation.^[1-3]

To the best of our knowledge, we present the first reported case in the Middle East of Brown-McLean syndrome in a patient with homocystinuria. The case report followed the ethical standards set by the World Medical Association Declaration of Helsinki. This case report was approved by the Institutional Review Board at King Khaled Eye Specialist Hospital, Riyadh, Saudi Arabia.

CASE REPORT

A 29-year-old Saudi male was referred to King Khalid Eye Specialist Hospital with the diagnosis of the bilateral subluxated lens and pupillary glaucoma in 1995. He was born to consanguineous parents (first cousins) in the Southern region of Saudi Arabia. Extensive

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workup led to the diagnosis of homocystinuria at other general tertiary hospital.

The patient underwent pars plana vitrectomy and lensectomy in the left eye in October 1995, followed by the right eye in February 1996. Postoperative refraction was $+16.00 - 1.00 \times 180^\circ$ in the right eye and $+15.00 - 1.00 \times 180^\circ$ in the left eye.

In 2013, he was diagnosed with chronic retinal detachment in the right eye with a poor prognosis, so no intervention was planned. April 2015, the patient followed up with no light perception (NLP) in the right eye and 20/100 in the left eye. Slit-lamp examination of the aphakic right eye was unremarkable [Figure 1a]. Examination of the left eye revealed peripheral corneal edema, small superior endothelial pigment, large inferior surgical peripheral iridotomy involving the pupil margin and aphakia with vitreous floaters in the anterior chamber superiorly without touching the cornea [Figure 1b-e].

Ultrasound B-scan of the right eye indicated a total funnel retinal detachment with moderate subretinal opacity. The posterior pole was within the normal limits in the left eye.

In 2017, the patient presented with decreased vision secondary to retinal detachment in the left eye after head trauma; his visual acuity was NLP in the right eye and 20/100 in the left eye. The patient underwent scleral buckle and cryotherapy in the left eye.

At follow-up in July 2018, the corrected visual acuity was 20/160 and the peripheral corneal edema with central corneal sparing remained stable in the left eye. The central endothelial cell count was normal in both eyes [Figure 2a] with abnormal cell count peripherally without reading [Figure 2b]. The patient was diagnosed with Brown-McLean Syndrome, and trial of sodium chloride ointment 5% was prescribed at bedtime to the patient. Six months later, the best-corrected visual acuity improved to 20/80, and the peripheral corneal edema almost was the same with flat retina in the left eye.

DISCUSSION

Homocystinuria is an autosomal recessive disorder that usually presents with ophthalmic abnormalities. Clinical ocular features of homocystinuria may manifest as lens dislocation, myopia, and glaucoma as in our case. The major ocular feature is bilateral lens dislocation that has been published in up to 70% of patients at 8 years old and in >95% in the fifth decade of life. Inferior lens dislocation is the most common in patients with homocystinuria, but the position is not diagnostic, as the lens may migrate in any direction. Burke *et al.* reported three patients with inferonasal lens migration like our case and superonasally in two patients.^[2]

Figure 1: (a) Right eye with clear cornea. (b) Left eye, external corneal edema from 2 o'clock till 10 o'clock, with a large surgical peripheral iridotomy inferiorly involving the pupil margin. (c) Left eye with slit view of peripheral corneal edema. (d) Left eye with slit view of clear central cornea with superior endothelial pigment (arrow). (e) Left eye with deep anterior chamber with vitreous floating away from the cornea

Figure 2: (a) Specular microscopy showed normal central endothelial cell density, both eyes. (b) Secular microscopy showed abnormal peripheral endothelial cell density without reading in the left eye

In our case, the patient developed bilateral pupillary block glaucoma secondary to lens dislocation that was treated with bilateral pars plana lensectomy and vitrectomy. He also developed related-complications with aphakia after trauma and finally underwent another surgery in the left eye. On July 2018, we diagnosed Brown-McLean syndrome in the left eye.

The pathogenesis of Brown-McLean syndrome is uncertain. However, it is reasonable to assume a genetic predisposition, as it was reported in 3 members of an extended family who presented with bilateral peripheral corneal edema.^[1,3,5,6] There are other predisposing factors that have been reported as in our case, such as high myopia, dislocation of the lens, and intermittent angle-closure glaucoma. Other possible predisposing factors include chronic uveitis and iridodonesis in aphakic patients that may cause intermittent abrasion of the corneal endothelium leading to peripheral corneal edema.^[1,4,6-9]

Earlier reports hypothesized that superior iridectomy may play a protective role against the development of edema in the superior cornea.^[1,7,9-12] However, in our case, the presence of inferior peripheral edema despite the presence of the large inferior iridectomy did not support this hypothesis.

Furthermore, despite the patient was not on any medication for corneal edema such sodium chloride, although with more surgical intervention for retinal detachment left eye patient keeps stable peripheral corneal edema with almost stable vision, so that makes close follow-up without surgical intervention for edema much valuable especially with such monocular patient.

In conclusion, treating ophthalmologists should consider the possibility of Brown-McLean syndrome in Saudi Arabia as this syndrome generally has no direct effect on central vision since the central cornea is spared. Patients with systemic diseases who present with subluxated lens, such as Marfan syndrome and homocystinuria, or patients who have undergone lens removal should be examined and followed closely.

Patient consent

Written/oral consent to publish the case report was obtained. This report does not contain any personal information that could lead to the identification of the patient.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his

consent for his images and other clinical information to be reported in the journal. The patient understands that name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Suwan Y, Teekhasaene C, Lekhanont K, Supakontanasan W. Brown-McLean syndrome: The role of iridodonesis. *Clin Ophthalmol* 2016;10:671-7.
2. Burke JP, O'Keefe M, Howell R, Naughton ER. Ocular complications in homocystinuria – Early and late treated. *Br J Ophthalmol* 1989;73:427-31.
3. Tourkmani AK, Martinez JD, Berrones D, Juárez-Domínguez BY, Beltrán F, Galor A, *et al.* Brown-mcLean syndrome in a pediatric patient. *Case Rep Ophthalmol* 2015;6:139-42.
4. Mohebbi M, Shadravan M, Pour EK, Ameli K, Badii S. Brown-mcLean syndrome in a patient with hallermann-streiff syndrome. *Korean J Ophthalmol* 2016;30:76-7.
5. Jamil AZ, Rahman FU, Mirza KA, Iqbal W. Brown-mcLean syndrome with keratoconus. *J Coll Physicians Surg Pak* 2012;22:179-81.
6. Vote BJ, Grupcheva CN, Ormonde SE, McGhee CN. *In vivo* confocal microstructural analysis and surgical management of Brown-McLean syndrome associated with spontaneous crystalline lens luxation. *J Cataract Refract Surg* 2003;29:614-8.
7. Gothard TW, Hardten DR, Lane SS, Doughman DJ, Krachmer JH, Holland EJ. Clinical findings in Brown-McLean syndrome. *Am J Ophthalmol* 1993;115:729-37.
8. Rutzen AR, Deen A, Epstein AJ, Maldonado MJ, Hemady RK. Cataract surgery in a patient with Brown-McLean syndrome. *J Cataract Refract Surg* 2001;27:1335-7.
9. Lim LT, Tarafdar S, Collins CE, Ramamurthi S, Ramaesh K. Corneal endothelium in Brown-McLean syndrome: *In-vivo* confocal microscopy finding. *Semin Ophthalmol* 2012;27:6-7.
10. Kam KW, Jhanji V, Young AL. Brown-McLean syndrome. *BMJ Case Rep*. 2013;2013:bcr2013201280. doi:10.1136/bcr-2013-201280.
11. Tuft SJ, Kerr Muir M, Sherrard ES, Buckley RJ. Peripheral corneal oedema following cataract extraction (Brown-McLean syndrome). *Eye (Lond)* 1992;6 (Pt 5):502-5.
12. Almousa R, Johns S, Gibson RA. Atypical clinical presentations of Brown-McLean syndrome. *Eye (Lond)* 2007;21:249-50.