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# Correspondence

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## Brown-McLean Syndrome in a Patient with Hallermann-Streiff Syndrome

Dear Editor,

Hallermann-Streiff Syndrome (HSS) is a rare genetic condition with multiple systemic involvement chiefly affecting the head and face and includes seven basic signs: dyscephalia and bird-like face, proportionate dwarfism, dental anomalies, atrophy of skin (especially on the nose), hypotrichosis, microphthalmia and congenital cataracts [1,2]. The Brown-McLean Syndrome (BMLS) is a clinical condition with peripheral corneal edema that involves stroma and epithelium with a 2 to 3 mm extension from the limbus to the center of the cornea [3]. We describe a case of HSS presenting with peripheral corneal edema after lensectomy. To our knowledge, this is the first report of HSS with BMLS.

A 31-year-old Caucasian woman with the chief complaint of sever blurred vision since childhood was referred to our hospital for ocular examination. She was at a height of 150 cm with a weight of 40 kg meaning she had proportional dwarfism. She had a miniature bird-like face, parrot beak nose, small mouth, and micrognathia with malformed teeth. She also had hypotrichosis of the scalp and skin atrophy (Fig. 1A).

She was the sixth child of non-consanguineous parents and was born by normal vaginal delivery following 40 weeks of uneventful pregnancy. There was no report of similar disorders in the family.

Her corrected distance visual acuity with a +10.25 -2.00 × 60 glass on the right eye was counting fingers at 30 cm, and was counting fingers at 50 cm for the left eye with a +8.50 sphere. Her right eye was esotropia and she had jerky nystagmus bilaterally. During slit lamp examination, she had bilateral peripheral corneal edema with more severity in the right side. The corneal diameter was 9.5 mm and 9 mm with an axial length of 21.2 and 21.6 mm in the right and left eye respectively (bilateral microcornea). There was golden brown pigmentation on the underlying endothelium of both eyes, but the central cornea was clear bilaterally (Fig. 1B).

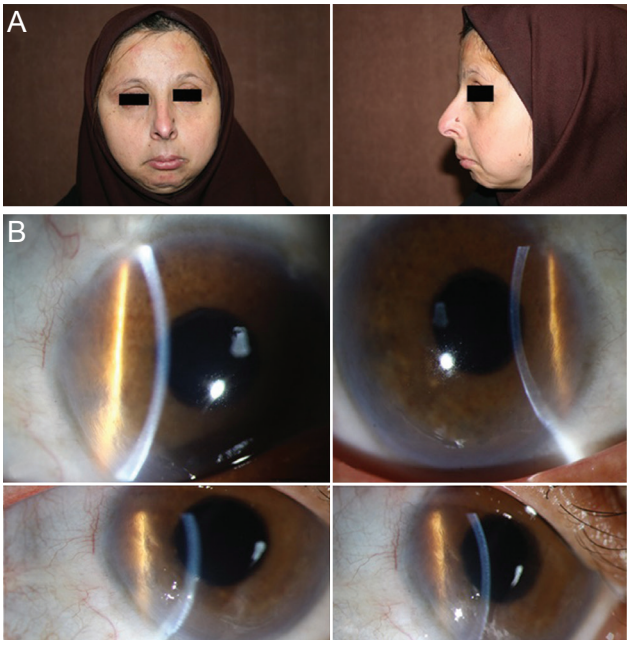


**Fig. 1.** (A) Facial features of Hallermann-Streiff syndrome including dyscephalia, micrognathia, bird-like face, atrophy of nasal skin. (B) Peripheral corneal edema with underlying endothelial pigmentation. Informed consent was received from the patient.

Anterior chambers of both eyes were deep and quiet. Intraocular pressure of both eyes was within the normal limit, and gonioscopy showed open angles in both eyes. She was aphakic in both eyes due to a past history of lensectomy when she was 6 years old. Fundus examination of both eyes showed normal examination.

Our patient had 5 out of the 7 criteria for HSS, but peripheral corneal edema with endothelial pigmentation in

this aphakic patient was related to BMLS. As a result, this patient was diagnosed as an HSS who developed BMLS after lensectomy, performed more than 20 years ago. The time that it takes BMLS to develop is 6 to 34 years after the causative condition, commonly after intracapsular cataract extraction [4].

Although the patient was probably amblyopic in both eyes, she insisted on removing her spectacles. After discussing other possibilities and obtaining an informed consent form, we decided to schedule her for an aphakic Artisan intraocular lens (Ophtec, Groningen, The Netherlands) implantation. Specular microscopy was not possible due to nystagmus.

Aphakic artisan implantation in the right eye was performed cautiously under general anesthesia. The power of the lens was +15 diopter. Postoperative visits of the patient were normal and the cornea had very mild stromal edema which resolved after 2 days. She was satisfied because she stated that the quality of her vision improved, specifically in an almost working condition. After 3 months corrected distance visual acuity was counting fingers at 60 cm and dry refraction in the right eye was: -1 diopter sphere, but the retinoscopic light reflex was not satisfactory. Peripheral corneal edema due to BMLS in this patient had no change overtime and so no more treatment modality was done.

HSS usually involves various ophthalmic abnormalities including: microphthalmia, microcornea, congenital cataracts, nystagmus and some posterior segment findings, such as retinal coloboma, retinal fold and vitreous degeneration [2].

It is suggested that iridodonesis in aphakic patients can intermittently abrade endothelium leading to corneal edema [5]. In our patient, peripheral corneal edema with pigmentation of underlying endothelium in aphakic eye without involvement of the central cornea are compatible with the diagnosis of BMLS. This syndrome is usually asymptomatic but complications such as bullous keratopathy and infectious keratitis may develop [3], so patients with BMLS should be visited periodically and alarming signs should be mentioned to such patients [3].

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## Conflict of Interest

No potential conflict of interest relevant to this article was reported.

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