Brown-McLean Syndrome in a Pediatric Patient

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Abstract
The purpose of this manuscript is to report the case of a 12-year-old patient who presented for routine ophthalmic examination after congenital cataract surgery performed at 2 months of age. The patient was diagnosed with bilateral Brown-McLean syndrome by slit lamp examination. No treatment was required because the patient was asymptomatic and had a clear central cornea. This is the first described case of Brown-McLean syndrome in a pediatric patient, representing the importance of clinical examination in the pediatric age group after cataract surgery because of the risk for patients of developing peripheral edema.

Introduction
Brown-McLean syndrome (BMS) is an infrequently diagnosed condition, defined by peripheral corneal edema that spares the center [1]. Clinically, BMS is most often seen after cataract extraction but has been described after other surgeries as well (e.g., corneal transplantation) [2, 3]. Nonsurgical insults such as lens subluxation [2, 4], angle closure glaucoma [5], myotonic dystrophy [6], and keratoconus [7] have also been described. Here, we report the first case of BMS in a pediatric patient.
Case Report

A 12-year-old boy presented for routine ophthalmic examination. The patient was asymptomatic on presentation. His past ocular history was significant for bilateral congenital cataracts which required bilateral cataract extraction by phacoemulsification with posterior chamber intraocular lens (PCIOL) implantation at 2 months of age. There was no other relevant medical (topical or systemic medication) or surgical history. Despite this intervention, the patient developed bilateral amblyopia and had severe nystagmus when either eye was occluded. At the time of presentation, his visual acuity was hand motion in both eyes. Tonometry and specular microscopy were not possible given the degree of nystagmus. Slit lamp examination of the right eye revealed a clear cornea with a fine pigmented line involving 360 degrees of the peripheral endothelium and a clear PCIOL. The left eye revealed normal conjunctiva, a clear central cornea with 360 degrees of peripheral stromal corneal edema with microbullae without epithelial defect, or vascularization, and a clear PCIOL (fig. 1). The rest of the ocular examination was normal. These findings were consistent with incipient BMS in the right eye and established BMS in the left eye. The patient remained asymptomatic, and no further treatment was started at the time.

Discussion

We herein present the first reported case of BMS in a pediatric patient. BMS, first described in 1969, is a condition defined by the presence of peripheral corneal edema with sparing of the central cornea [1]. In its initial phase, a pigmented line on the peripheral endothelium may be seen. On progression, edema typically starts inferiorly and progresses circumferentially [7]. Our patient's findings were consistent with early BMS in the right and more advanced disease in the left eye. One previous report described a 12-year-old girl with pigmented endothelial precipitates in the inferior peripheral cornea in the setting of congenital cataract removal 11 years earlier. However, the diagnosis of BMS was not certain given the lack of corneal edema [8].

BMS typically occurs, on average, 17 years after surgery, with a widely published range of 1 month to 34 years [3, 4, 6–8]. A recent randomized, controlled study in 114 patients who underwent cataract extraction for congenital cataracts evaluated outcomes after lens implantation versus no lens implantation. Overall, complications were low. With regard to corneal edema, no patients in the aphakia group and 1 patient in the IOL group developed central corneal edema that lasted more than 30 days. No cases of peripheral edema were reported in either group [9].

The pathophysiology, which leads to peripheral corneal edema as opposed to central changes, is still unclear. BMS has been thought to be associated with altered dynamics of the iris and aqueous that can occur after surgery and/or trauma [10]. BMS may have a genetic influence as it was reported in three members of an extended family, with a strong family history of low vision and ruptured tendons [4]. It has also been reported in two pairs of sisters, one pair of which had Marfan syndrome [2].

Interestingly, the condition is typically stable without progressive central corneal decompensation, and, therefore, patients usually remain asymptomatic. Although specular microscopy may be performed to monitor the condition, it may not predict the clinical course, as the central cornea usually remains transparent, even when the central cell density is low [2]. However, transient central edema associated with increased intraocular pressure,
foreign body sensation, infectious keratitis, bullous keratopathy, and corneal endotheliitis in patients with BMS has been reported [11].

As our patient was asymptomatic, at this stage, our plan is to observe him. However, prior reports of patients with BMS described various treatments to alleviate ocular surface discomfort including topical corticosteroids, hypertonic solutions, and/or therapeutic contact lenses [7]. Other reported options include stromal micropuncture of the peripheral cornea and annular amniotic membrane transplantation [12, 13].

In summary, our case highlights that BMS may develop in children as well as in adults. All children who undergo ocular surgery or experience ocular trauma should be monitored on a regular basis for the development of this and other potential ocular complications.

Disclosure Statement

The authors have no conflicts of interest to report.

References

Fig. 1. Slit lamp photograph of the left eye demonstrating peripheral corneal edema.