Eye (2007) 21, 249–309

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Sir,

Atypical clinical presentations of Brown–McLean syndrome

Brown–Mclean syndrome (BM syndrome) was first described in 1969 as a rare condition of peripheral corneal oedema with underlying endothelial pigmentation, observed in long term aphakic patients.[1]

The oedema is often observed after a latent period of several years following surgery, and previously has been described as typically starting inferiorly, with progression that may involve the entire circumference in more severe cases.[2]

The syndrome is often asymptomatic, but patients may present with foreign body sensation, or pain due to a ruptured bullae.[2]

The youngest patient previously described with this syndrome was 26 years old and the oldest was 96 years old.[2]

We report two cases of BM syndrome in which one had the unusual presentation of the peripheral corneal oedema starting superiorly, and another of a 12-year-old patient, who shows possible early signs of the syndrome.

Case reports

Case 1

A 79-year-old female was referred to the contact lens clinic in 2001 for a routine follow-up to the right eye. Best-corrected visual acuity (BCVA) was right eye 6/6 and left eye 6/36 due to chronic cystoid macular oedema following extra capsular cataract extraction (ECCE) with posterior chamber intraocular lens (PCIOL) in 1984. The right eye was aphakic due to intracapsular cataract extraction with broad sector iridectomy superiorly (Figure 1). She had a history of severe uveitis in the left eye. Slitlamp examination showed an area of peripheral corneal oedema in the right eye (2.5 mm) with bullae formation covering the entire superior half circumference and no underlying endothelial pigmentation. Inferiorly there was peripheral orange–brown endothelial pigmentation.

After 2 months, the area of corneal oedema showed greater epithelial and stromal thickness and for the first time, endothelial punctate pigmentation was noticed underlying the area of peripheral oedema superiorly, which increased on further review a month later. At this point, the patient was put on 4 months follow-up and has remained to date with no change in her corneal appearance.

Case 2

A 12-year-old girl was referred to the contact lens clinic for assessment in January 2001. She was bilaterally aphakic, having undergone congenital cataract aspiration, right eye (April 1990) and left eye (September 1990). There was no family history of eye problems and the patient had a full-term normal delivery. At the age of 3, she developed a left divergent squint due to ambylopia, for which she had undergone a left squint surgery.

In July 2001, during regular contact lens check up, slitlamp examination revealed discrete, small endothelial brown pigmentation covering the inferior peripheral half of the right cornea (Figure 2). The central cornea had very few corneal guttata, and the diagnosis of BM syndrome was considered to be a possibility. The BCVA was right 6/6 and left 1/60. There was no peripheral corneal oedema in either cornea.

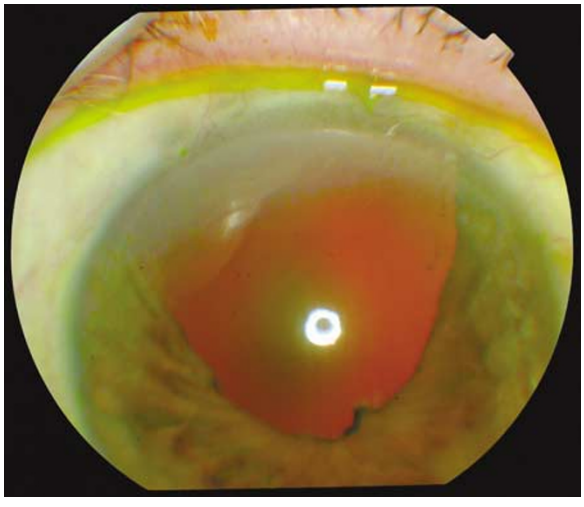

Figure 1 Superior peripheral corneal oedema overlying area of broad sector iridectomy

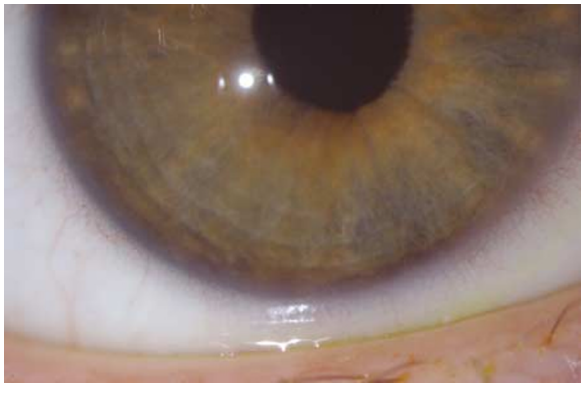

Figure 2 Discrete brown peripheral endothelial pigmentation inferiorly

She was fitted with a soft contact lens to both eyes but the left contact lens was discontinued latter, as there was no visual benefit.

Since the first documentation of the endothelium pigmentation, to date (3.5 years), there has been neither progression to peripheral oedema nor progression of the endothelial pigmentation to cover the superior half of the cornea.

Discussion

Case 1 is the first time that the peripheral oedema has been seen to start superiorly as opposed to the classical description in which the oedema usually begins inferiorly. Initially there was no pigmented endothelium underlying the area of oedema, but it appears at later review. The patient presented in Case 2 was 12 years of age when the typical endothelial pigment was noted, the youngest to present with an aspect of BM syndrome. Previously Gothard *et al*[2] described a 26 years old patient with BM syndrome. Taft *et al*[3] argued that endothelial pigmentation alone is not specific for this syndrome as they observed a similar pattern of pigment dispersion in eyes following ICCE in which there is no sign of peripheral oedema. We would suggest that this syndrome may have a variety of signs that could range from peripheral corneal oedema alone, endothelial pigmentation alone or the combination of both.

In aphakic patients, iridodonesis has been suggested to be a cause of an intermittent abrasion of the endothelium[4,5] and that superior iridectomy could have some role in protecting the superior portion of the cornea from the development of oedema,[2] but we can argue that case 1 had a broad sector iridectomy superiorly yet still had peripheral oedema overlying the area of iridectomy.

It is our belief that BM syndrome is often under diagnosed and that it has variable presentations that should be documented even though it will remain asymptomatic in most patients. Patient should regularly be reviewed by an ocular healthcare professional and educated about the clinical signs and symptoms, this is especially so if the patient is corrected with contact lenses as opposed to spectacles.

References

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Eye (2007) **21,** 249–250. doi:10.1038/sj.eye.6702244; published online 27 January 2006

Sir,

Questionnaire-based research in ophthalmology: questioning the quality

There have been many articles over recent years that have included questionnaires in the study methods, most recently the National Biometry Audit II.[1] Much has been published both in text books and in the peer-reviewed literature concerning best practice in questionnaire design and utilisation,[2–5] and while questionnaires are clearly an invaluable research tool when used appropriately, poorly constructed questionnaires produce meaningless or misleading responses, and lack of rigour in analysis leads to the formation of invalid conclusions.[6]

The National Biometry Audit II utilised a telephone-administered questionnaire, recruiting 94 biometrists from 178 potential interviewees (53%);[1] there would be no reason to assume that the 47% who did not respond were similar to those who did. The risk of systematic bias being introduced is clearly a problem that should have been discussed in the article; nonresponders might well have been found to be less rigorous in their biometry practice than responders, hence the true picture of biometry practice in the UK may be less healthy than that reported.