Mitochondrial myopathy presenting with segmental corneal oedema and retrocorneal membrane

Chronic progressive external ophthalmoplegia (CPEO) is a genetic disorder of mitochondrial DNA characterised by progressive ocular dysmotility and eyelid ptosis.¹ Other ophthalmological manifestations of mitochondrial eye disease include optic atrophy and retinal pigmentary changes. There have also been several reports of corneal opacification associated with Kearns-Sayre syndrome and ophthalmoplegia-plus.²⁻⁵ Corneal decompensation secondary to exposure from lagophthalmos and absent Bell's phenomenon has been reported in CPEO, but few have progressive peripheral corneal oedema.^{6,7} We describe a patient presenting initially with bilateral segmental corneal oedema, in whom the subsequent diagnosis of CPEO followed the corneal changes.

Case report

A 38-year-old black woman with a diagnosis of primary ovarian failure presented with 5 years of bilateral progressive corneal oedema. Initial visual acuity was 20/60 OD (right eye) and 20/40 OS (left eye). Ptosis and diplopia were absent whereas funduscopy was unremarkable. Gradually, the segmental corneal oedema progressed to involve her visual axis OS (fig 1 A,B). Gonioscopy showed a pigmented retrocorneal membrane extending to the peripheral cornea, but not involving the angle structures (fig 1 C,D). Confocal microscopy showed a network of highly reflective bodies corresponding to the pigment spicules seen clinically with endothelial guttae, polymegathism, pleomorphism and reduced endothelial cell density (fig 2 A,B). Bilateral eyelid ptosis was evident within 2 years (fig 1E), with global limitation of extraocular movements. The patient underwent penetrating keratoplasty OS in December 2004. Histopathological examination of the corneal button showed corneal epithelial bullae, stromal oedema and a normal Descemet's membrane. The attenuated, one-cell layered endothelium showed focal loss of cells and phagocytosis of melanin pigment (fig 2C). Additionally, there was a thin retrocorneal membrane, which was negative for cytokeratin (Kermit; fig 2D). In June 2005, the patient underwent muscle biopsy to evaluate the aetiology of her eyelid ptosis and reduced ductional amplitudes. Skeletal muscle analysis showed myopathy (ragged red fibres with intramitochondrial paracrystalline inclusion bodies) consistent with a mitochondrial disorder. Biochemical analysis was remarkable for reduced oxidative metabolism, whereas a known genetic mutation for CPEO was not found.

Comment

Mitochondrial disease presents with a wide variety of ocular, neurological and systemic manifestations. Although corneal endothelial abnormalities have been reported in association with mitochondrial disease, the diagnosis of mitochondrial myopathy is generally manifest before the corneal disease. Moreover, the clinical picture of segmental bilateral oedema in association with a pigmented retrocorneal membrane is unique and previously unreported in CPEO (although pigment deposition

Figure 1 (A,B) Slit-lamp images of the peripheral corneal oedema before penetrating keratoplasty. (C) Gonioscopic and (D) retroilluminated views of the pigmented retrocorneal membrane. (E) Bilateral upper eyelid ptosis. Informed patient consent was received for publication of this figure.

is seen with many types of endothelial dysfunction).⁸ This finding is more characteristic of the Brown-McLean syndrome, which is typically seen after ocular surgery, particularly after intracapsular cataract extraction.⁹ Additionally, Rutzen *et al*¹⁰ described a patient with myotonic dystrophy and peripheral corneal endothelial pigmentary changes without retrocorneal membrane, consistent with the Brown-McLean syndrome. The unique features of peripheral corneal oedema, pigmented retrocorneal membrane and subsequent ocular dysmotility are a unique presentation of CPEO. We recommend that mitochondrial myopathy be considered in the differential diagnosis of young patients with idiopathic corneal oedema.

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Figure 2 Confocal microscopy showing (A) highly reflective spiculated bodies of the posterior cornea, probably corresponding to the retrocorneal membrane and (B) likely pigment of peripheral corneal guttae. Electron microscopy showing (C) degenerated endothelial cells with vacuolar degeneration and phagocytosis of melanin granules (Me), and (D) an endothelial cell (E) overlying a retrocorneal membrane with collagen fibrils (R).

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Importance of multimedia visual information in improving patient understanding of cataract surgery

Cataract surgery is the most common surgical operation carried out by ophthalmologists. Patients undergoing cataract surgery have been found to have a low level of understanding of the operation, and this is increasingly being linked with increasing patient dissatisfaction.¹

Pager's recent article² showed that a simple inexpensive videotape showing patients what

to expect from cataract surgery increases their understanding and satisfaction with the surgery, as well as reduces patient anxiety. We carried out a cross-sectional study to look at whether a multimedia visual computer program improved patients' understanding of cataract surgery as compared to a paper leaflet.

Case report

A total of 42 patients with ophthalmic disorders, with no history of cataract, were allocated to either the computer program or the leaflet group. Understanding of cataract and cataract surgery was assessed by means of a questionnaire relating to what is a cataract and what was involved in the operation.

The results showed that understanding was markedly improved in the computer group as compared with the paper leaflet group. In all, 70% of patients in the computer group had a good understanding of what is a cataract as compared with 59% in the paper leaflet group. Also, 68% of patients in the computer program group had a good understanding of what was involved in the operation as compared with 58% in the paper leaflet group. Mean age in both groups were similar (58.4 v 60.2 years).

Comment

Multimedia visual information is an important tool for teaching; this also involves patients. The results signify that information given via a computer program improves understanding of cataracts and cataract surgery. This has similar

connotations to the findings by Pager in his recent article, which is an important and timely reminder of the need to improve ways of increasing patient understanding and satisfaction in cataract surgery.

This is only a pilot computer program and still requires modifications, but it is thought that using such a computer-based program would be a vital resource for patients in undergoing cataract surgery. Unfortunately, at present, few ophthalmic units have developed new ways of improving patient understanding of cataract surgery. It is hoped that this would act as a template for such development.

In our study, only patients without a history of cataract were included to limit bias due to previous knowledge of cataract. Our study therefore is a good reliable indicator of the use of computer-based information in improving understanding of cataract and cataract surgery in two similar ophthalmic patient populations. We believe that this is mainly due to the improved visual representation of cataract surgery.

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Progressive macular thinning after indirect traumatic optic neuropathy documented by optical coherence tomography

Optical coherence tomography (OCT) is a noninvasive optical imaging technique that provides high-resolution, cross-sectional, in vivo imaging of the human retina from which estimates of retinal layers thickness can be made.¹ OCT is capable of scanning the peripapillary retina, optic nerve head (ONH), and macular region. Previous studies have documented the ability of OCT to image the retinal nerve fibre layer (RNFL) of patients with glaucoma or other optic neuropathies including indirect optic nerve trauma.^{2–4}

Although OCT has for the most part been used to evaluate RNFL thickness, recent software improvements have made it possible to measure macular thickness as well. The purpose of this paper is to report a case of indirect optic nerve injury which illustrates the efficacy of macular thickness measurements in documenting progressive neural damage in a longitudinal study using OCT.

Case report

A 25-year-old woman was admitted to the hospital after injuring her right temple in a car accident. Her visual acuity was hand motion in the right eye (OD) and 20/20 in the left eye,