Computerised visual field deficits in tears of the retinal pigment epithelium

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Abstract
Retinal Pigment epithelial tears have been well documented as a complication of pigment epithelial detachment in patients with age related macular degeneration. Spontaneous and iatrogenic separation of detached retinal pigment epithelium, with subsequent retraction and exposure of the underlying choriocapillaris and Bruch’s membrane, usually results in poor visual function in the affected areas. However, exact characterisation of the resultant scotomas has not been previously described. We present two patients with spontaneous pigment epithelial tears who underwent Octopus computerised visual field analysis. The density and characteristics of their associated field loss is compared with their clinical and fluorescein angiographic appearance.

Retinal pigment epithelial tears or rips were first described by Hoskin et al in 1981 as a newly recognised complication of age related macular degeneration in patients with associated pigment epithelial detachments. Several authors have since reported on this phenomenon. Associated visual loss is usually abrupt and severe when the foveal or parafoveal area is involved. Casswell and associates noted a 10% incidence of spontaneous tears during routine follow-up examinations in at risk patients. These typically occurred in eyes with late fluorescent pigment epithelial detachments, and usually within a few months of presentation.

To our knowledge the characterisation of scotomata associated with RPE tears has not been considered. For this reason the reproducible quantification of associated scotomata by computerised visual fields was performed to evaluate the visual field loss and compare it with the clinical and fluorescein angiographic appearance.

Patients and methods
Two patients aged 60 and 70, were examined on the Wills Eye Retina Service for age related macular degeneration with associated pigment epithelial detachment. Both were noted to develop spontaneous tears of the retinal pigment epithelium. Neither patient had clinical evidence or a past history compatible with glaucoma, optic neuropathy, or retinal vascular disease.

Computerised visual fields were calculated by the Octopus 201 static perimeter. Basic software programs 31 and 32 were used to define grossly the visual field abnormalities. Higher resolution, with detailed definition of the defects, was obtained with advanced software. Octopus multiple-use user defined programs (UDP) CO-4 and CO-8 were employed. Both patients gave reliable responses to the tests.

Initial testing was carried out with standard 30° radius visual fields with 6° of resolution (programs 31 and 32). These programs were useful in attempting to roughly define the location, size, shape, and contour of the scotomas. Multiple-use UDP programs CO-4 and CO-8 were then used to define closely the visual field loss. Resolution of 1° and 2°, respectively, allowed detailed characterisation of the scotoma. CO-8 is an 8° radius visual field, CO-4 is a 4° radius visual field; both test 57 points, double determining nine of these points.

Case reports

CASE 1
A 70-year-old male was first seen in September 1981 complaining of decreased vision and metamorphopsia in his left eye. His visual acuity was 20/25 in the right eye and 20/400 in the left eye. Funduscopic examination revealed changes consistent with bilateral age related macular degeneration (ARMD) and a subretinal neovascular membrane in the left macula. Subsequent follow-up was uneventful until March of 1986, when a detachment of the retinal pigment epithelium was observed inferotemporal to the fovea in the right eye; visual acuity was 20/30. Six weeks later the patient noted a sudden decrease in vision to 20/80. A pigment epithelial tear, along the temporal edge of the previously recorded pigment epithelial detachment (PED), was noted. Nasal retraction of the pigment epithelium exposed underlying choroid up to the edge of fixation.

Figure 1: Case 1. A fluorescein angiogram at 36 seconds after injection shows early hyperfluorescence in the area of absent retinal pigment epithelium (arrows).
Fluorescein angiography showed a C-shaped area of mottled hyperfluorescence increasing in the late frames and corresponding to the exposed choroidal vasculature. The retracted pigment epithelium resulted in block choroidal fluorescence (Fig 1).

Octopus computerised static perimetry was performed three weeks after diagnosis of the pigment epithelial tear. A standard 30° radius central visual field (program 32) was initially used to outline grossly the extent of the scotoma located superotemporal to fixation (Fig 2). Closer definition of the nasal aspect of the tear, with its retracted pigment epithelium, was accomplished with the UDP CO-8 (Fig 3). Both fields revealed an absolute or near absolute scotoma in areas with absent pigment epithelium. However, decibel loss in the area containing retracted pigment epithelium was minor.

CASE 2

A 60 year-old woman with a history of ARMD was seen in May 1982 with a history for several months of decreased visual acuity and distortion of her vision in the right eye. Visual acuity was 20/40 in the right eye and 20/30 in the left eye. Funduscopic examination revealed a PED of the left eye with scattered drusen. The right eye had numerous drusen and a double pigment epithelial tear. One tear was nasal and the other temporal to fixation. Fluorescein angiography showed retraction of the retinal pigment epithelium centrally on both sides of the fovea. Areas where retinal pigment epithelial was absent showed marked hyperfluorescence (Fig 4).

Octopus computerised static perimetry was performed one year later with a standard 30° radius field, which failed to delineate visual field loss (Fig 5). Higher resolution UDP programs CO-4 and CO-8, however, defined precisely the position of absolute decibel loss corresponding to the clinical position of the two pigment epithelial tears. Normal visual function was maintained centrally between the tears where retracted pigment epithelium was present (Fig 5, 6, 7). Sixty months after initial presentation the patient’s clinical appearance and visual acuity remain unchanged.

Discussion

Tears of the retinal pigment epithelium are a known complication of pigment epithelial...
detachments associated with ARMD. They result in retraction of the pigment epithelial layer towards the PED centre and away from the line of cleavage. The overlying sensory retina remains intact but unsupported by the retinal pigment epithelial monolayer. Angiographically, retracted retinal pigment epithelium blocks choroidal fluorescence, whereas adjacent areas devoid of retinal pigment epithelium are hyperfluorescent.

Although tears occurring in the central macular area have a poor visual prognosis, the nature and density of the resultant scotomas have not been well described. Computerised static perimetry, with resolution up to $1^\circ$ of visual field, allows characterisation of the resultant scotomas when compared with the clinical appearance. In our two patients an absolute scotoma developed where retinal pigment epithelium no longer supported sensory retina. Conversely, adjacent areas containing retracted retinal pigment epithelium continued to function at normal or near normal levels, with minimal or no decibel loss. Although the patient described in our first case had visual field testing within one month after his retinal pigment epithelial tear, our second patient was not tested until one year afterwards. Both had absolute visual field defects implying the permanence of these changes.

The onset of visual dysfunction probably depends on loss of the physical, and therefore metabolic link between photoreceptors and pigment epithelium. Hoskin and colleagues’ report noted one patient whose visual acuity decreased from 20/30 to 20/200 over several days after a tear developed. This drop in vision coincided with the onset of structural changes described in animal models of sensory retinal detachments. Of the 44 eyes with pigment epithelial tears reported by Hoskin and associates only eight retained a normal anatomical relationship between foveal pigment epithelium and photoreceptors. All, as in case 2, retained good visual acuity.

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