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CASE REPORT

Symptomatic extra-macular traumatic pigment epitheliopathy

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To describe the evolution and diagnosis of post-traumatic pigment epitheliopathy (TPE) in a young male following blunt trauma to the globe as well as discuss the histopathological and optical coherence tomography (OCT) features of TPE.

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Clinical Case

A 17-year-old male presented to the ophthalmic casualty clinic after being hit in the right eye by a football. The patient complained of profound blurring of vision from his right eye associated with photophobia and bulbar pain. No symptoms were reported with regards the left eye. The patient and his accompanying relative both excluded any relative past ophthalmic history which could contribute to the loss of visual acuity.

Initial examination revealed a right visual acuity of 6/36 (Snellen) which did not improve on using pinhole. Left eye visual acuity was 6/6 unaided. Ecchymoses of the right superior palpebra was noted on inspection. The right pupil was dilated and non-reactive to light but produced pupillary constriction in the left eye on reverse relative afferent pupillary defect testing. Extra-ocular movements appeared to be unrestricted and there was no globe displacement. Slit-lamp examination of the right eye revealed a diffuse corneal haze, linear epithelial defect and a hyphema was noted in the anterior chamber with active bleeding originating from the supero irido-corneal angle. Intra-ocular pressure in the right eye was 9mmHg. Traumatic mydriasis was noted. No fundal view could be obtained at the time of presentation, but B-scan ultrasonography did not reveal any abnormalities in the fundus. The patient was admitted for strict bed rest and started on topical chloramphenicol ointment, high-frequency dexamethasone eye drops and topical cyclopentolate.

On the second day following admission the corneal haze cleared and a fundal view was obtained. Retinal pallor just superior to the macula could be observed with loss of the foveal reflex (Figure 1). Scattered intra-retinal haemorrhages were observed in the periphery. OCT revealed diffuse retinal oedema across all the retinal layers with a central macular thickness of 440µm. Topical nepafenac was prescribed and the patient was discharged after 4 days once the hyphema resolved. After one week, vision from the right eye improved to 6/18 and the central macular thickness reduced to 223µm.

On the fourth week following presentation the patient started complaining of difficulty fixing on a target when closing his left eye. Repeat examination revealed a visual acuity of $6/9^{+3}$ in the right eye and the pupillary diameter came down to 4mm with a normal photic response. On Amsler grid evaluation the patient could not see the grid inferior to the central target. Fundoscopy revealed an area of mixed hypo-pigmentary and hyper-pigmentary changes superior to the macula which exhibited hypo-autofluorescence (Figure 2). An inferior para-central scotoma, corresponding to the depigmented area on fundoscopy, was confirmed on Humphrey visual field testing (Figure 3). The diagnosis of TPE was made. The patient was advised that the visual field defect should not affect his activities of daily living as it will be compensated for by the binocular field and his left eye was the dominant eye of the two. Yearly follow-up was advised then after.

Figure 1 Fundal photo of the right eye demonstating pigmentary changes superior to the macula.

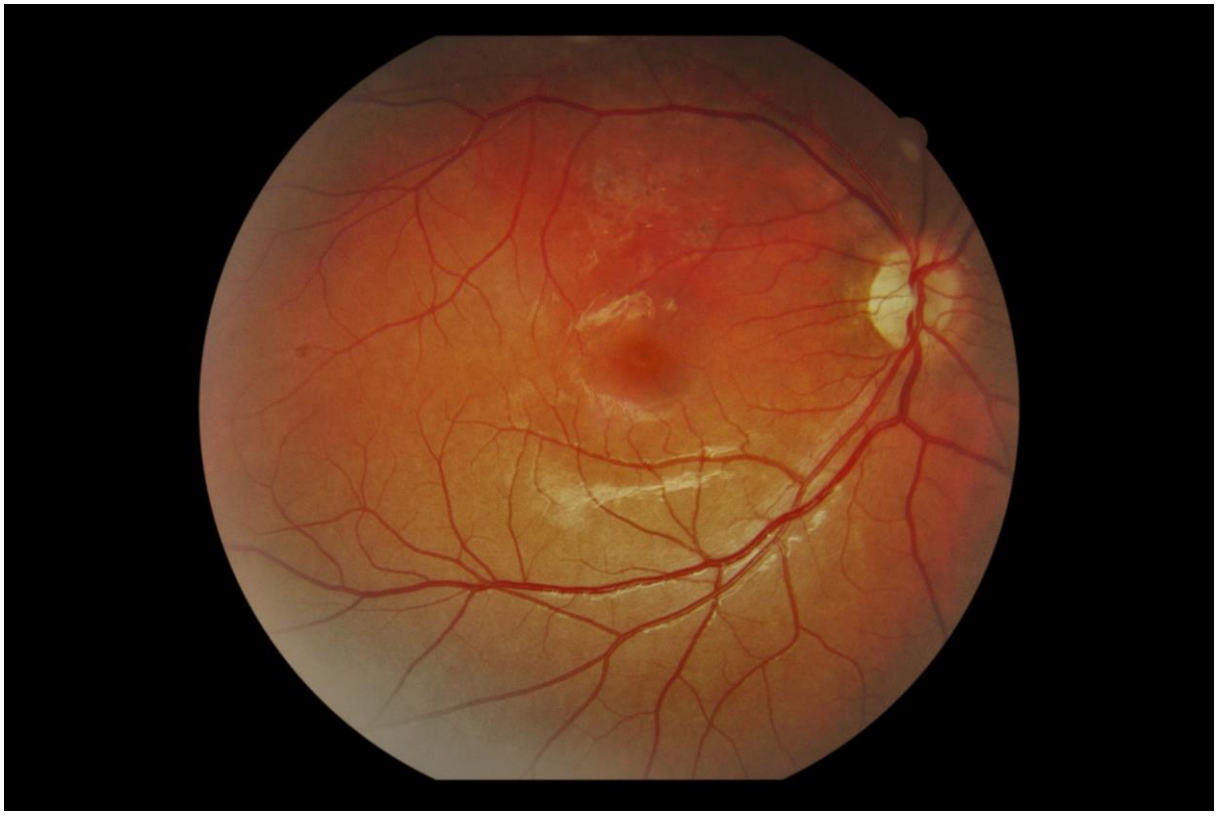


Figure 2 Fundal Auto-Fluorescence photograph of the right eye showing hypo-autofluorescence superior to the macula denoting RPE loss.

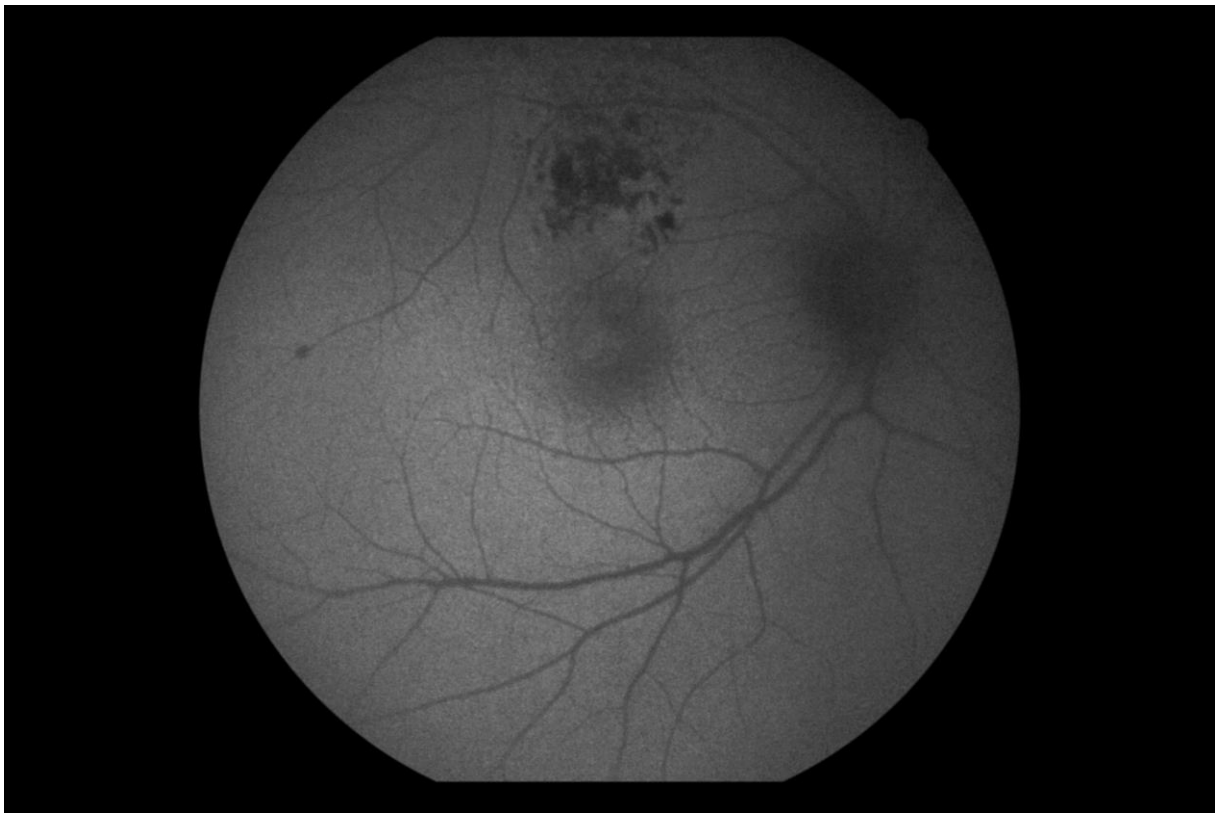
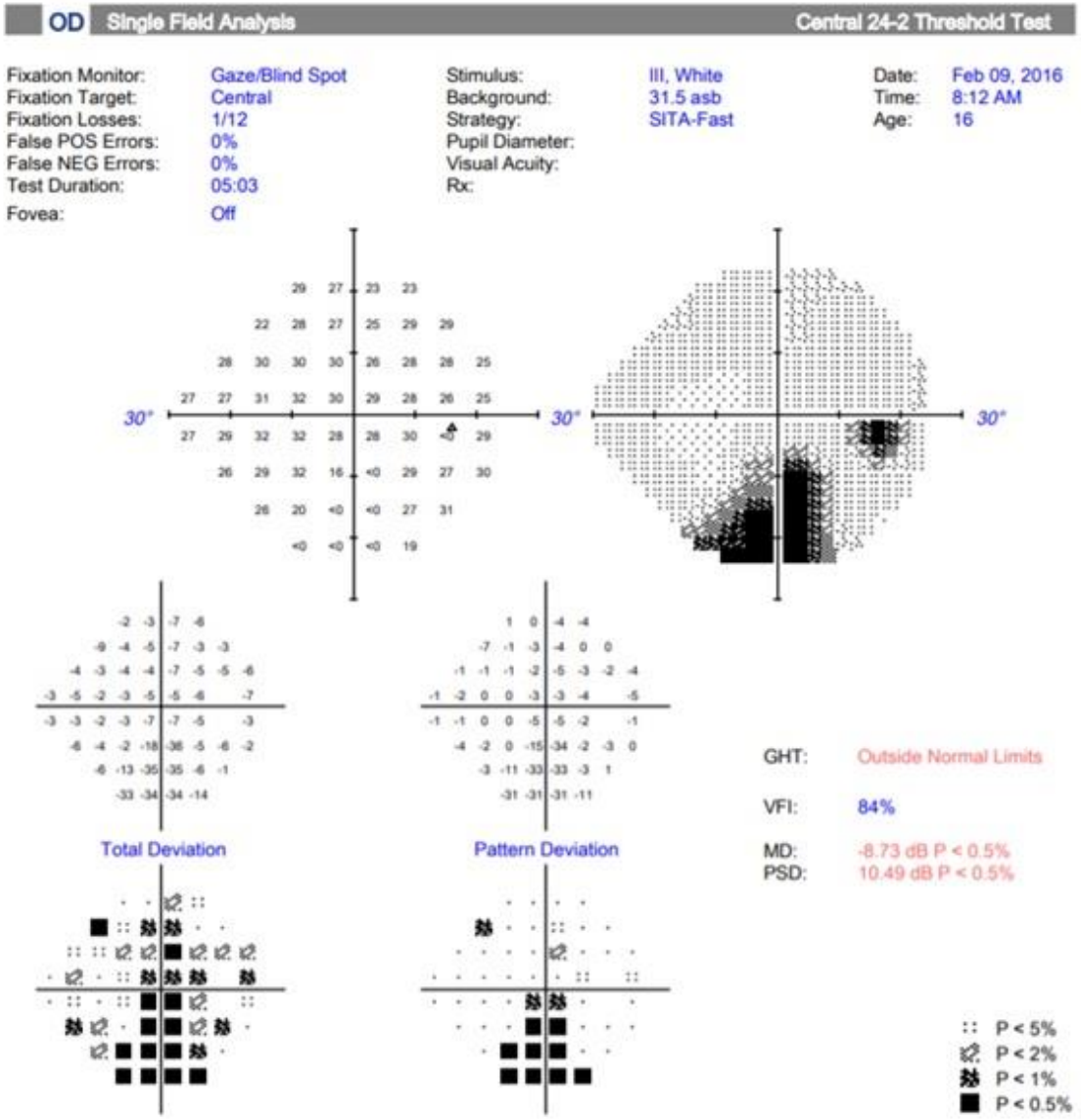


Figure 3 24-2 Humphrey visual field test confirming the inferior para-central scotoma in the right visual field corresponding to the superior-lying are of RPE loss secondary to TPE.



Discussion

Blunt trauma to the eye is characterized by acute globe deformation followed by reformation resulting in stretching of the intraocular contents. Vitreous deformation, caused by the transmission of kinetic energy, results in stretching of the retinal tissues, particularly the outer retinal layers which lack the structural support of Müller cells offered to the inner layers[1], resulting in a condition known as commotio retinae (CR). This condition is clinically characterised by a white-yellow opacification of the affected retinal tissue, as noted on dilated fundoscopy, and can be classified as macular or extra-macular CR with the former having the highest risk of visual loss due to the development of central or para-central scotomata.[2] The development of these scotomata may be due to irreversible damage to the outer retinal tissues resulting in ~~a~~ fragmentation of the photoreceptor outer

segments and retinal pigment epithelium (RPE) apical process loss.[1] Permanent disruption of the aforementioned structures has been coined as traumatic pigment epitheliopathy (TPE), a late complication of CR.[3]

Lavinsky *et al.* described the evolution of TPE in 2 patients following a closed globe injury secondary to blunt trauma which was preceded by CR over the same fundal area 1-3 months before the diagnosis of TPE was made.[3] On fundoscopy, TPE was described as an area containing mixed atrophic and hypertrophic RPE changes, the latter being characterised by scattered loci of hyperpigmentation. The RPE changes noted on fundoscopy corresponded to the patterns seen on fundal autofluorescence imaging demonstrating a high degree of heterogeneity[3] which could be attributed to foci of RPE cell loss (hypo-autofluorescence) and areas of metabolic substrate accumulation (hyper-autofluorescence) from photoreceptors and RPE cell degeneration.[4] The prognosis of TPE depends on the location involved, hence involvement of the macular area carries the highest risk of functional visual deficit due to the formation of a central scotoma although symptomatic para-central scotomata, as in this case, have been documented in the literature.[2] To date there is no effective therapy which can aid in the resolution of this condition.

Histopathological reviews on the formation of CR and its progression in animals[5] and humans[1] have been published in the past. Blight and Hart carried out sequential retinal biopsies in animal subjects following blunt ocular trauma over a period of 2 months.[5] The latter experiment determined that the immediate changes following trauma, including the characteristic opacification of the retinal tissue, was not an ischaemic phenomenon but was due to the formation of intracellular oedema, primarily in the outer retinal layer. The formation of intracellular oedema was accompanied by the fragmentation of the photoreceptor OS layer as well as the loss of apical processes of the RPE cells[5] which would explain the acute symptomatology in macular CR should the posterior pole be involved. Similar changes were noted in the report published by Mansour, Green and Hogge who carried out a histological examination of an enucleated eye obtained from a patient involved in a motor vehicle accident.[1] Examination of retinal biopsies 2 months following trauma showed re-organisation of the photoreceptor OS layer in animal subjects accounting for the symptomatic improvement noted after the resolution of CR.[5] There is no published literature on the histopathology of TPE or its progression from CR, although the findings noted on examination and imaging in this case suggest that when subjected to more severe trauma, loci of RPE cells may be lost (accounting for the hypo-auto-fluorescence noted on fundal autofluorescence photos) with subsequent metabolic damage to the outer retinal layers, which would have also been subjected to mechanical disruption, resulting in permanent visual field defects.

Despite the lack of direct observation of the histological changes that occur in TPE, OCT has allowed researchers to gain more insight on the pathophysiological process leading from CR to TPE in a non-invasive manner.[6] In a case series consisting of 13 patients investigated using spectral-domain OCT during their follow-up following blunt ocular trauma, Souza-Santos et al. noted immediate post-traumatic changes on OCT in TPE patients which differed from the early changes noted in CR patients.[6] Hyper-reflectivity across all the retinal layers,

as opposed to hyper-reflectivity noted below the inner-segment/outer-segment junction in CR patients, was associated with disease progression (p=0.002), namely outer nuclear layer and photoreceptor layer thinning[6] which could be attributed to RPE cell loss and secondary photoreceptor cell death.

To date there is no established treatment to interrupt the evolution of TPE and current management is limited to regular clinical examination and the use of ophthalmic imaging for the purpose of prognostication.

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