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Short Communication

Torpedo Maculopathy Associated with Refractile Drusen and Dry Age-Related Macular Degeneration: Preserved Retinal Pigment Epithelial Function Around the Torpedo-Like Lesion?

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Torpedo maculopathy (TM), a rare oval-shaped lesion in the temporal macula with its nasal tip toward the fovea, typically shows central hypoautofluorescence with a hyperautofluorescent border suggesting retinal pigment epithelium (RPE) loss and dysfunction, respectively. Some of the optical coherence tomography (OCT) features described include neuroretina thinning, outer retina disruption, RPE atrophy and a subretinal cleft in some cases [1]. Recent multimodal imaging has revealed that intact RPE and Bruch's membrane on swept source OCT in some areas of the lesion [2].

We report an 84 y old Caucasian female with unilateral TM associated with intermediate dry age-related macular degeneration (AMD). To the best of our knowledge, the eldest TM case in the literature and the first with AMD. She was complaining of gradual deterioration in her reading vision and paracentral scotoma in the right eye. Uncorrected visual acuity was 6/9 right, 6/7.5 left. Anterior segments were normal apart from lens nucleosclerosis. Funduscopy showed refractile drusen (material representing a byproduct of defective RPE-photoreceptor metabolism) with patchy areas of chorioretinal atrophy (right>left eye). The refractile drusen in the right macula were distributed within concentric zones across the macula, as recently described by Suzuki et al. [3]. In the left eye, the area around the torpedo lesion did not have refractile drusen or autofluorescence features suggestive of RPE dysfunction. What is more, the OCT scans above and below the torpedo lesion revealed intact structure of the RPE/Bruch's membrane complex and inner choroid (Figures 1and 2).

The aetiology, pathophysiology and natural history of TM remain unknown. Various developmental defects have been proposed at the level of the horizontal raphe nerve fiber layer [4], the RPE within the temporal bulge [5], and the emissary canal of the long posterior ciliary artery and nerve [6]. Here, we present a patient in her 9th decade with intermediate dry AMD where the RPE in the area around the torpedo lesion does not seem to have age-related RPE dysfunction to the same degree as the rest of the macular RPE in that eye or the fellow eye. The pathogenesis of TM may possibly result in choriocapillaris, Bruch's membrane and/or RPE changes around the torpedo lesion that make this area less vulnerable to age-related dysfunction. Longer follow up will shed some light on how the outer retina structure and function evolves around the torpedo lesion.



Figure 1: Right eye macula color fundus photograph (a) showing medium-size refractile drusen in concentric zones around the fovea, mixed with areas of atrophy. The left macula color photograph (b) shows a sharply demarcated temporal oval lesion along the horizontal raphe with the tip pointing towards the fovea, with outer hypopigmentation and inner hyperpigmentation. Medium-sized refractile drusen can be found in the nasal macula and inferiorly but not in the area around the torpedo-like lesion. Right eye autofluorescence image (c) shows а mixture of hypoautofluorescence indicating missing or dead RPE cells, and hyperautofluorescence corresponding with lipofuscin accumulation. Left eye autoflorescence image (d) shows hypoautofluorescence in the inner area of the torpedo-lesion and a mixture of hyper and hypoautofluorescence in the nasal macula corresponding with the area where refractile drusen have developed.

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Figure 2: Right eye Heidelberg spectralis OCT scan images (a) show outer retina and RPE atrophy (arrows) and areas of hyper reflectivity corresponding with refractile drusen (*). Left eye horizontal OCT scans across the fovea (b) show a well preserved photoreceptor layer at the centre of the fovea (double arrow). The torpedo-like lesion shows missing RPE and outer neuroretinal layers (\$). In contrast, the outer neuroretina, RPE/Bruch's membrane complex and inner choroid appear intact above (c) and below (d) the torpedo lesion (^).

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