Structural-functional correlation using adaptive optics, visual fields, optical coherence tomography and multifocal electroretinogram in a case of torpedo maculopathy

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We present a case of a 37-year-old gentleman with a rare diagnosis of Torpedo maculopathy (TM). We describe the multimodal imaging features of torpedo maculopathy using adaptive optics, visual fields, OCT and multifocal ERG, and understand the clinical and structural-functional correlation in TM. According to us, this is the first case report to describe the adaptive optics imaging findings in Torpedo maculopathy in English Medical literature.

Key words: Adaptive optics, imaging, OCT, torpedo maculopathy

Roseman and Gass, in 1992, first described torpedo maculopathy (TM) as a rare, congenital anomaly of the retinal pigment epithelium (RPE) characterised by the appearance of a 'torpedo-shaped' lesion located temporal to the fovea.[1]

A diagnosis of TM is made on the basis of its typical fundus appearance described as an oval-shaped solitary hypo-pigmented lesion, resembling a 'bullet' or 'torpedo,' with a wedge-shaped tail extending outward and pointing toward the foveola along the horizontal raphe.[2] Imaging modalities like optical coherence tomography (OCT) have confirmed the hypothesis that TM is a congenital abnormality of the RPE and outer retinal layers. The adaptive optics (AO) imaging

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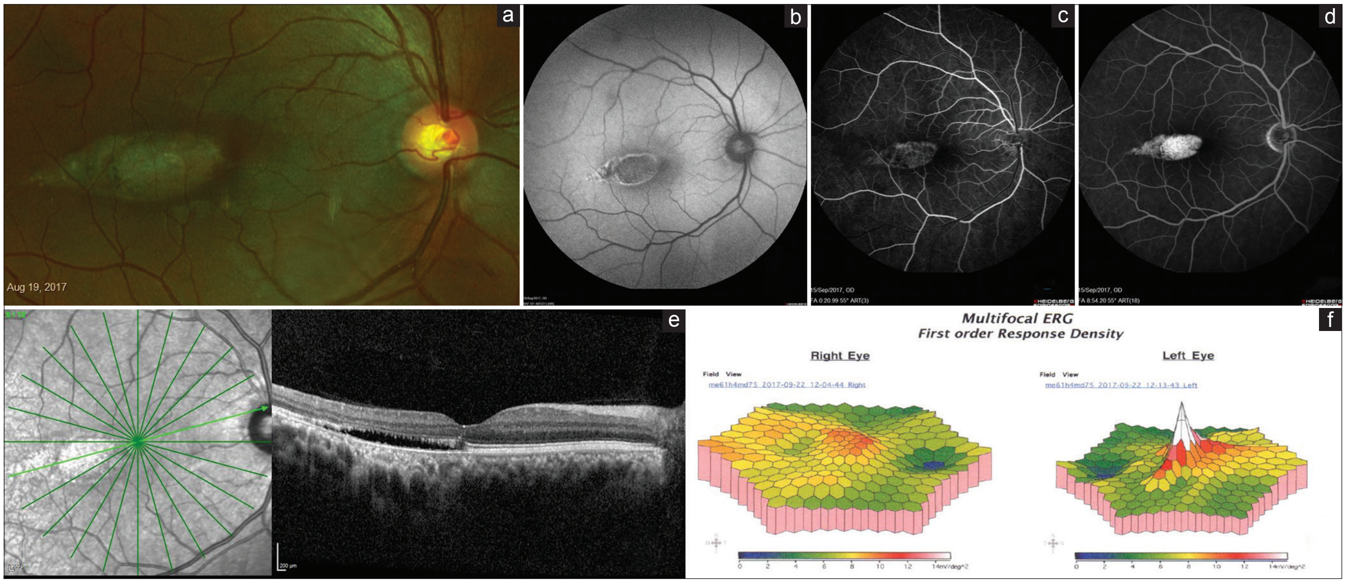

Figure 1: Multimodal imaging in Torpedo maculopathy. (a) Right eye colour fundus showing the classical torpedo-shaped lesion; (b-d) Fundus autofluoroscence and fluorescein angiography images depicting the lesion; (e) OCT image showing the presence of subretinal cleft with loss of ellipsoid and inter-digitation zone with overlying attenuation of the outer nuclear layer. Inner retinal layers appear normal. RPE atrophy with increased choroidal structure visibility is noted; (f) Multifocal electroretinogram showing reduced responses and foveal peak

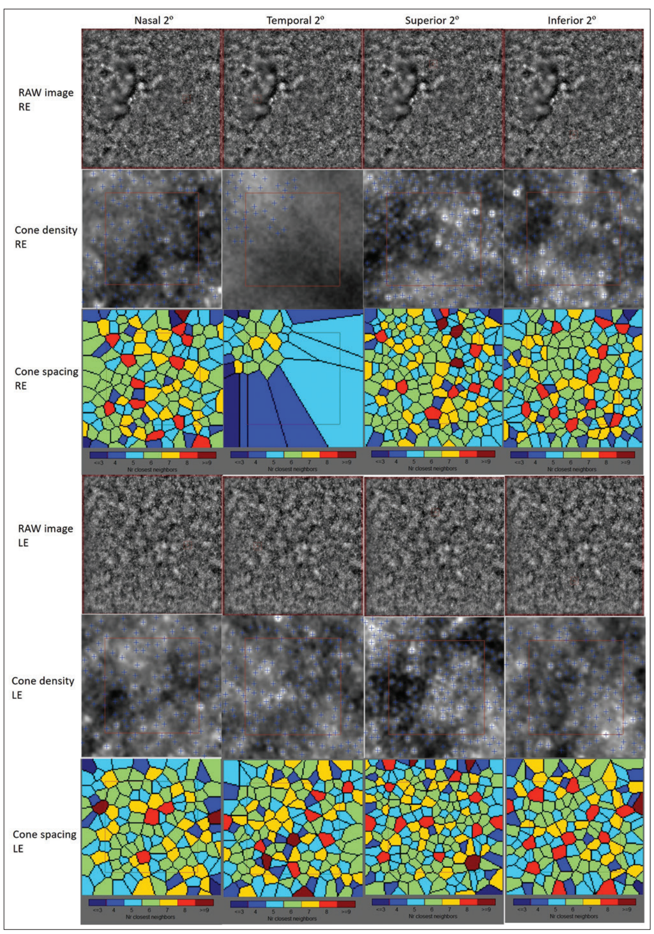

Figure 2: Adaptive optics imaging showing raw images, cone density and cone spacing of both eyes at 2° from the foveal center in all 4 quadrants

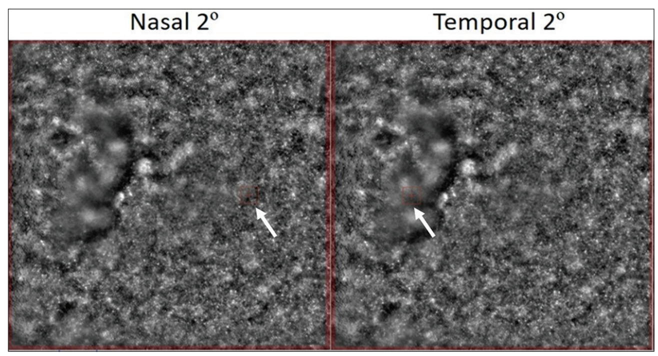

Figure 3: RAW images on Adaptive optics imaging showing the exact site of cone density and spacing analysis (bold white arrow)

technology takes ultra-high-resolution retinal images to study the microscopic structural findings like the configuration and density of photoreceptors.[3] It has found wide applications in retinal pathologies specially inherited retinal diseases. In this report, we describe the microscopic features in the photoreceptors in TM using AO imaging with rtx1 AO imaging camera.

Case Report

A 37-year-old man, diagnosed previously as a 'chorioretinal scar' in the macula, came for follow-up examination at our retina clinic. He had no ocular symptoms. Systemic history was normal. Eye examination revealed vision of 20/20 in both eyes. Anterior segment examination was normal in both eyes. Dilated fundus examination revealed pink and healthy optic nerves with normal physiologic cupping in each eye. Right eye macular examination revealed a classical, torpedo-shaped, hypopigmented chorioretinal lesion. Left

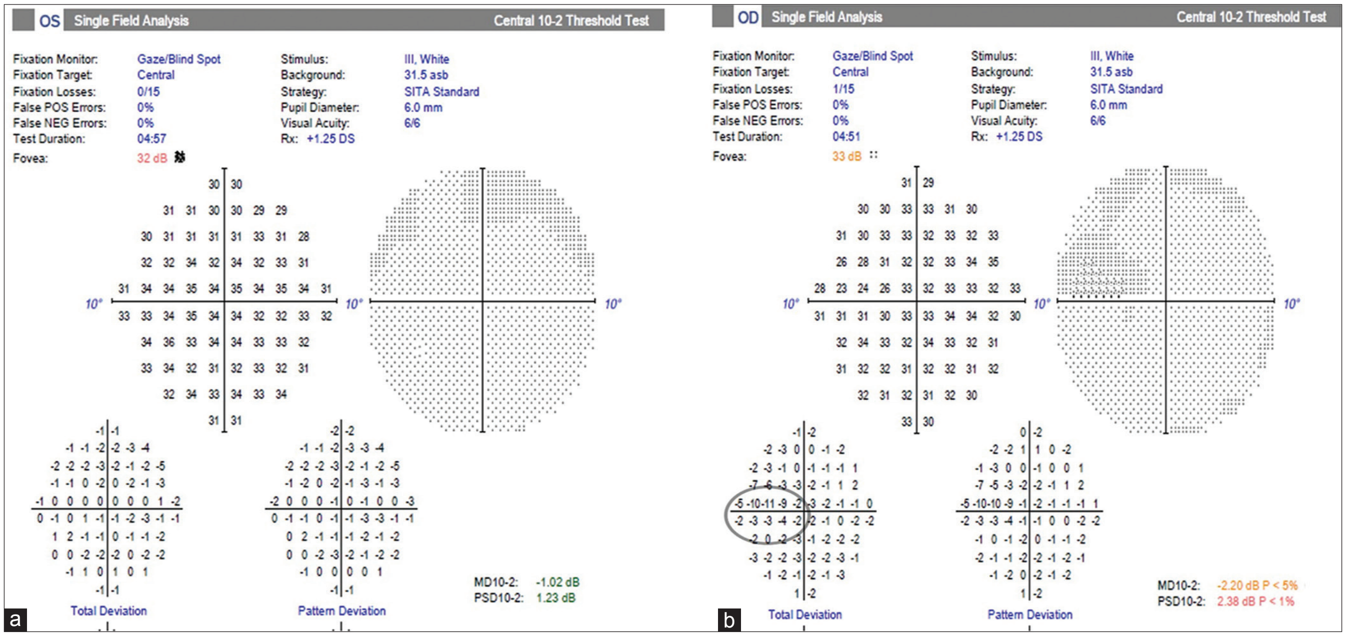

Figure 4: 10-2 Humphrey visual field testing of both eyes. (a and b) Right eye showing reduced retinal sensitivities in the region of TM in right eye compared to other regions in the same eye and left eye (circle with black boundary)

Table 1: Cone density and spacing calculated on adaptive optics imaging

	Right eye		Left eye	
	Cone density (cones/mm^2)	Cone spacing (μm)	Cone density (cones/mm^2)	Cone spacing (μm)
2° temporal	8	173.29	17587	8.35
2° nasal	18509	8.17	19513	7.91
2° superior	22365	7.32	24532	7.06
2° inferior	21999	7.45	17923	8.30

eye macula was unremarkable. Peripheral retina was normal in both eyes. Multimodal imaging of the torpedo lesion was done with fundus autofluoroscence, fundus fluorescein angiography, OCT, multifocal electroretinogram (mfERG) and Humphrey visual fields [Fig. 1]. AO retinal imaging was done using the rtx1 flood illumination-based fundus camera (Imagine Eyes, Orsay, France) and its automated software *AO detect mosaic*™ was used to assess the cone density and spacing across the central retina. Cone density, cone spacing and cone arrangement were analysed with *AO detect mosaic*™ in a region of interest (ROI) of 80 × 80 pixels, corresponding to 62 × 62 μm on the retina for an axial length of 24 mm at 2° from the foveal fixation, in all 4 quadrants in both eyes [Figs. 2 and 3]. Decreased cone photoreceptors with increased spacing between them was noted in the region of TM in comparison to the other areas of both eyes. The values are depicted in Table 1.

Discussion

In this report, we describe the adaptive optics imaging features of torpedo maculopathy using the rtx1 flood illumination-based fundus camera (Imagine Eyes, Orsay, France) and its automated software *AO detect mosaic*™.

TM is often asymptomatic and typically found during routine clinical examination. The classical fundus description is pathognomonic for TM. Despite the typical appearance, the rarity and unknown cause of this disorder may pose a diagnostic challenge. Different imaging modalities have provided insights into the morphological and possible pathophysiological mechanisms associated with this lesion.

Based on OCT, Wong *et al*.[4] have classified the torpedo lesions into 2 types: 1) Type 1 - with attenuation of outer retinal structures and absence outer retinal cavitation; 2) Type 2 - with presence of attenuated outer retinal structures and outer retinal cavitation. These 2 types of OCT findings represent the different levels of involvement of the same disease.

In our case, we found the type 2°CT finding with thinning of the outer retinal layers, presence of subretinal cleft with underlying RPE atrophy and increased choroidal structure visibility. AO retinal imaging was done using the rtx1 flood illumination-based fundus camera (Imagine Eyes, Orsay, France) and analysis was done with its automated software *AO detect mosaic*™. AO imaging involves a deformable mirror which alters its shape corresponding to the wave front of light reflected from the patient's eye. This improves the resolution of image captured by the instrument equipped with the AO technology. The transverse resolution of the AO imaging with the above-mentioned flood illumination camera is 2-4 microns and a central 4° × 4° retinal image was obtained.

In our case, the cone density was reduced and there was increased spacing between the adjacent cone photoreceptors at the lesion site in comparison to the other retinal areas of both eyes. The retinal sensitivities were reduced on 10-2 Humphrey Visual Field test in the region of TM [Fig. 4]. Furthermore, we obtained reduced responses on mfERG at the same location of the torpedo lesion similar to that described by Buzzonetti *et al*.[5] The central 5° ring showed abnormal mfERG waveforms with reduced amplitudes on the temporal side of the fovea compared to the remaining region. This resulted in a blunted foveal peak on mfERG compared to the normal eye. The responses obtained on mfERG are mathematical extractions of individual focal ERGs generated by the electrical activity of the cone photoreceptors at a specific region in the retina. The mfERG responses in TM depends on the extent of photoreceptor loss which can be evaluated with the help of AO imaging. In our case, visual acuity in eye with TM was 20/20 despite having a blunted foveal peak on mfERG. This can be explained by the presence of intact photoreceptors and overlying external limiting membrane at the fovea as seen on OCT.

To conclude, AO imaging is a useful tool in identifying and quantifying the extent of photoreceptor loss secondary to TM. Also, the amount of photoreceptor degeneration has no correlation with visual acuity.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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