Multimodal imaging of torpedo-shaped fundus lesions: New insights

Shreyans Jain, Devesh Kumawat, Vinod Kumar

The purpose of this case series is to describe the varied presentation of Torpedo lesions of the fundus and multimodal imaging features in three eyes of three patients. Two patients presented with typical topography, i.e., temporal to the fovea. One patient revealed lesion inferonasal to disc with the head pointing toward the disc. All three patients had an attenuation of outer retinal layers on optical coherence tomography. One patient showed an additional large subretinal cleft. Variable hypoautofluorescence in the area of the torpedo was noted. To conclude torpedo lesions can present at atypical locations, have both retinal and choroidal atrophy and head point toward the optic disc.

Key words: Multimodal imaging, swept-source optical coherence tomography, torpedo maculopathy

The diagnosis of torpedo maculopathy (TM) is clinical and based on typical characteristic features.[1] It is usually a unilateral congenital oval-shaped hypo pigmented chorioretinal lesion present in temporal part of the macula with a pointed tip (head) directed toward fovea.[2] Here, we present the multimodal retinal imaging of three eyes with torpedo-shaped lesions and describe atypical features of disease with an aim to improve the understanding of the disease.

Case Reports

This is a retrospective observational case series of three eyes of three patients who were diagnosed incidentally with torpedo lesions. The study adheres to the institutional guidelines for research and the tenets of declarations of Helsinki.

The medical records were analyzed retrospectively which included demographics, systemic and ocular history, clinical examination details including Snellen visual acuity at presentation, color fundus photographs, optical coherence tomography (OCT) images, and shortwave fundus autofluorescence (SW-FAF). Two patients underwent multimodal imaging using DRI OCT Triton (Topcon Medical Systems, Oakland, NJ, USA) which included color fundus photograph, swept source OCT and SW-FAF. In the third patient, only color fundus photograph and spectral domain OCT (Cirrus HD-OCT 5000, Carl Zeiss Meditec, Dublin, CA) was done.

Access this article online

Quick Response Code:

Website: www.ijo.in

DOI: 10.4103/ijo.IJO_118_18

Dr. Rajendra Prasad Centre for Ophthalmic Sciences, All India Institute of Medical Sciences, New Delhi, India

Correspondence to: Dr. Vinod Kumar, Dr. Rajendra Prasad Centre for Ophthalmic Sciences, All India Institute of Medical Sciences, New Delhi - 110 029, India. E-mail: drvinod_agg@yahoo.com

Manuscript received: 23.01.18; Revision accepted: 10.04.18

Case 1

A 66-year-old male was referred for a "retinal lesion" found during the routine ocular examination. Unaided visual acuity was 20/20 in OU. OD fundus was normal. OS had a torpedo-shaped flat hypopigmented lesion (1 disc diameter (DD) length × 0.75 DD width) in temporal macula with the tip pointing toward fovea or optic disc [Fig. 1a]. On red-free imaging, torpedo shape of the lesion was more evident with sharply delineated border [Fig. 1b].

Swept Source (SS)-OCT (Topcon Inc.,) showed localized thinning of the retina at the site of the lesion with overlying localized posterior vitreous detachment (PVD) [Fig. 1d and e]. While inner retinal layers were thinned out, outer retinal layers were markedly affected. The outer nuclear layer (ONL) was very thin but continuous. Ellipsoid zone (EZ) and interdigitation zone (IZ) were disrupted while RPE was attenuated over the area of the lesion. The external limiting membrane was intact. An area of focal hyper-reflectivity was noted in outer retinal layers at the nasal edge of the lesion. Increased backscatter was noted over the deep choroidal layers (black arrowheads)

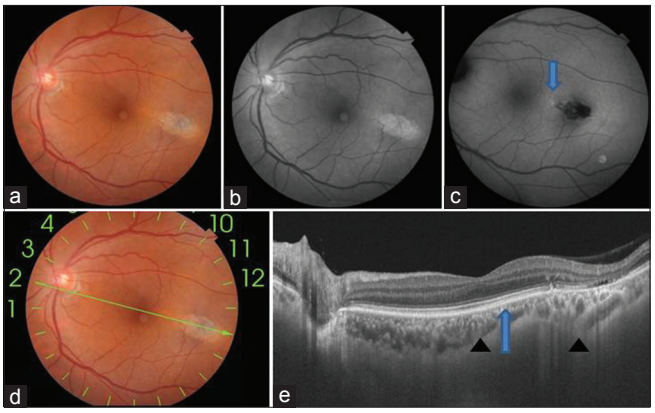

Figure 1: Colour fundus photograph of the left eye (a) of 66-year-old male shows torpedo-shaped hypopigmented lesion with well-delineated margin on red-free imaging (b). Fundus autofluorescence (c) hypoautofluorescence at the level of lesion with hyperautofluorescent margin. At the green arrow in (d) indicating the site of the scan, Swept Source optical coherence tomography (e) showsdisruption of the ellipsoid zone and interdigitation zone with attenuation of RPE. Hyperautofluorescent spot presents nasal to head of the lesion (blue arrow in c) corresponding to a hyper-reflective lesion on optical coherence tomography (blue arrow in e) lipofuscin deposition. Black arrowheads indicate increased backscatter over the deep choroidal layers

Cite this article as: Jain S, Kumawat D, Kumar V. Multimodal imaging of torpedo-shaped fundus lesions: New insights. Indian J Ophthalmol 2018;66:1211-3.

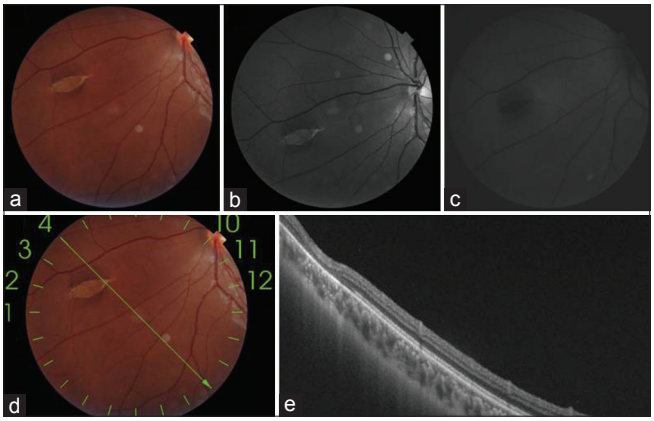

Figure 2: Color fundus photograph of left (a) eye of a 40-year-old male showing torpedo lesion in the inferonasal quadrant with the tip pointing toward the optic disc. Margins of the lesion are better defined on red-free imaging (b). Autofluorescence imaging (c) hypoautofluorescence with hyperautofluorescent margin. At the green arrow (d) indicating the site of the scan, Swept Source-optical coherence tomography (e) thinning of outer retinal layers and choroid

with reduced overall choroidal thickness and prominent large vessel layers.

SW-FAF showed hypoautofluorescence corresponding to the area of the lesion. Mildly increased autofluorescence was noted around the torpedo lesion [Fig. 1c]. There was a small hyperautofluorescent spot seen nasal to head of lesion (blue arrow) which corresponds to a hyper-reflective lesion on OCT [blue arrow in Fig. 1e] images at the level of the outer retina which may be due to local deposition of lipofuscin material. The patient was advised regular follow-up.

Case 2

A 40-year-old male patient presented with difficulty in near vision. Systemic and family history was non-contributory. The BCVA was 20/20 in OU with refractive correction of +0.5 DS/+0.5DC 100° OD and 0.5DS OS. Dilated fundus examination revealed a torpedo-shaped, sharply defined, hypopigmented lesion (1.25DD length × 0.75DD width) inferonasal to disc with the tip pointing toward the optic disc [Fig. 2a]. The lesion did not lie along the horizontal raphe. The lesion was better delineated on red-free fundus imaging [Fig. 2b]. SW-FAF showed hypoautofluorescence with hyperautofluorescent line around the lesion [Fig. 2c]. SS-OCT of the lesion revealed normal inner retinal layers and marked thinning of the outer retinal layer, i.e., EZ, and IZ leading to a focal dipping at retinal surface. The choroidal thickness was reduced at the site of the lesion with preserved large vessel layer [Fig. 2d and e].

Case 3

An 8-year-old girl with Crouzon syndrome was referred for ophthalmic evaluation. Family history was noncontributory. Her BCVA was 20/60 OU with– 2.0 diopters sphere. Anterior segment examination was essentially unremarkable apart from the presence of V pattern exotropia and shallow orbits. She had a large (1.5DD length × 1DD width) flat hypopigmented lesion inferotemporal to fovea suggestive of TM in the OD [Fig. 3a]. The tip of the lesion pointed toward the optic disc. On SD-OCT it showed generalized retinal thinning (marked atrophy of both EZ and IZ.), a subretinal cleft, a shallow choroidal excavation,

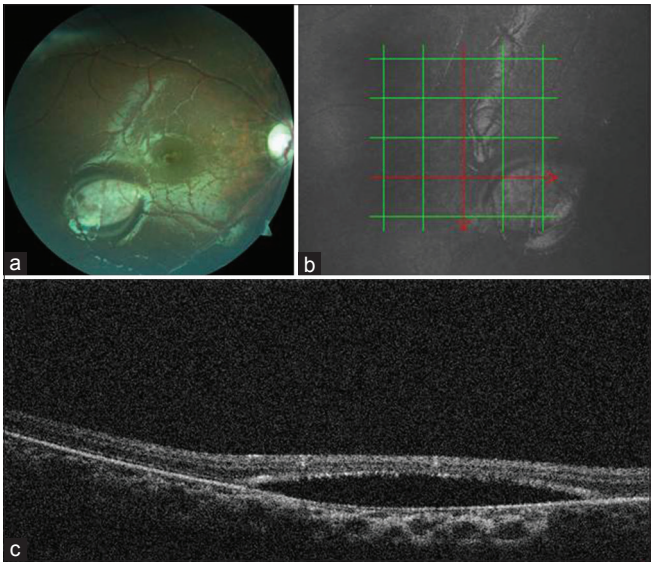

Figure 3: Color fundus photograph of the right eye (a) 8-year-old female child shows torpedo-shaped lesion presents inferotemporal to the fovea. At the horizontal red arrow indicating the site of the scan (b), Spectral domain-optical coherence tomography shows atrophy of ellipsoid zone and interdigitation zone, choroidal excavation, and decreased choroidal thickness (c)

and decreased choroidal thickness with prominent outer choroidal vessels [Fig. 3b and c]. The optic disc was pale in OU. Rest of the fundus in OS was normal.

Discussion

The exact clinical significance of TM is unknown apart from the fact that it may be confused with some forms of congenital hypertrophy of RPE (CHRPE) and may cause field defect in few patients.[2] Various proposed theories include a developmental defect in nerve fiber layer at the horizontal raphe, defect in the development of RPE within the fetal temporal bulge, malformation of emissary canal of the temporal long posterior ciliary artery and nerve and improper fenestration of choriocapillaris along the horizontal raphe.[1,2]

Contrary to the published literature, the lesion was located on the nasal side of the disc in case 2. Furthermore, the lesion was not located along the horizontal meridian. This atypical location of torpedo lesion necessitates its differentiation from CHRPE and RPE lesions of Gardner syndrome. Both CHRPE and RPE lesions of Gardener syndrome are hyperpigmented lesions associated with RPE thickening with decreased choroidal reflectivity on OCT.[3] In our case, however, the lesion was hypopigmented and was associated with increased choroidal backscatter, thus favoring TM like pathology. The occurrence of torpedo lesion on the nasal side challenges all the proposed theories of a defect in the development.

SW-FAF showed hypoautofluorescence at the site of the lesion with a halo of hyperautofluorescence. On the basis of SD-OCT findings, TM has been classified in two types: mild outer retinal disturbance (type 1 torpedo lesion) and outer retinal cavitations (type 2 torpedo lesion).[4] The outer retinal cavitation or subretinal cleft is usually shallow and reported to arise probably due to outer retinal atrophy and shallow choroidal excavation in the absence of angiographic RPE leakage.[1,4,5]

While many OCT features of TM in this case series (ONL thinning, disruption of EZ and IZ, and increased backscatter) are similar to those reported in the literature,[6-8] SS-OCT was able to pick up additional findings. First, thinning of inner retinal layers and overlying localized PVD has never been reported in the literature previously. Second, the subretinal cleft noted in case 3 was too deep to be secondary to outer retinal atrophy alone. We believe inner choroidal atrophy and fluid accumulation secondary to localized RPE defect may have also contributed to it. Third, increased RPE hyper reflectivity was noted in case 1 and 2 in the area of the lesion without any thinning or thickening of RPE. We believe it occurred due to decreased backscatter from overlying attenuated retinal layers. Altered melanin and lipofuscin levels in RPE would also have contributed to this finding. Fourth the outer retinal cavitation was seen in a young child. On the contrary, Wong *et al*. had previously reported type 2 lesions to occur in older age.[4] Thus, the current understanding about this disorder is far near the requisite, and it would be too early to classify them based on the limited published data.

Conclusion

Torpedo lesions can present at an atypical location with both retinal as well as inner choroidal atrophy. The direction of the head is toward the posterior pole, irrespective of the location of the lesion.

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.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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