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Case Report

Atypical Torpedo Maculopathy

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Abstract

Torpedo maculopathy is a rare congenital anomaly of RPE in the temporal macula with unknown etiology. The diagnosis of this lesion is primarily clinical, often an incidental finding as it tends to spare the fovea. The lesion characteristics includes location temporal to fovea, hypopigmentation, horizonally oval shape with certain variable features like intraretinal cleft, fundus excavation, variable hyperpigmentation and associated visual field defects. Here we report a case of nasal macular location of the lesion which represents a rare clinical presentation and is supported by multimodal imaging findings.

Keywords: Torpedo Maculopathy; Atypical; Torpedo; Nasal Location; Maculopathy

Introduction

Torpedo maculopathy is a rare congenital anomaly of RPE in the temporal macula with unknown etiology, often presenting as an incidental finding usually sparing the central fovea [1,2]. It was originally described by Roseman and Gass in 1992 [1] and later termed 'torpedo maculopathy' by Daily in 1993 [3]. Its constant features include location temporal to fovea, hypopigmentation, congenital lesion (s) and horizontally oval shape with certain variable features like intraretinal cleft, fundus excavation, variable hyperpigmentation and associated visual field defects [4]. Wong., *et al.* [5] proposed an OCT based classification of these lesions as Type I with attenuation of outer retinal structures and Type II with additional outer retinal cavitation; recently Type III with inner retinal excavation has also been described [6]. The differential diagnosis includes congenital hypertrophy of retinal pigment epithelium (CHRPE), simple hamartoma of RPE, congenital Toxoplamosis, choroidal nevus as well as other chorioretinal scars.

Case History

A 10-year-old boy was referred to our retina clinic following an incidental finding of chorioretinal scar in his left macula during a routine clinical examination. Best corrected visual acuity was 6/6, N6 in both eyes. The patient had no contributory medical, ocular or family history. Anterior segments of both eyes were quiet and IOP was within normal limits. Slit lamp biomicroscopic examination revealed a well defined, solitary, flat, hypopigmented, obliquely oval lesion in inferonasal macula of left eye. Binocular indirect ophthalmoscopy revealed flat peripheral retina with no other chorioretinal lesions in left eye. Right eye examination was unremarkable (Figure 1-3). The fundus autofluorescence imaging showed hypoautofluorescence with inferior marginal hyperautofluorescence. Optical coherence tomography through the lesion revealed disorganization of outer retinal layers and thinning of RPE (Figure 4). Enface structural OCT at the level of outer retina clearly delineates the extent of involvement (Figure 5) with focal loss of choriocapillaries within the lesion revealed through OCT-A at the level of choriocapillaris (Figure 6).

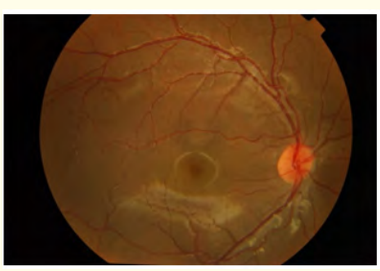

Figure 1: Normal fundus right eye.

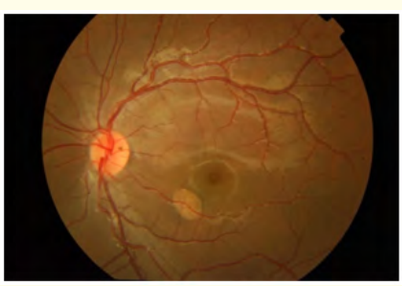

Figure 2: Left eye fundus showing flat hypopigmented pale orange well defined oblique oval lesion in the inferonasal macula.

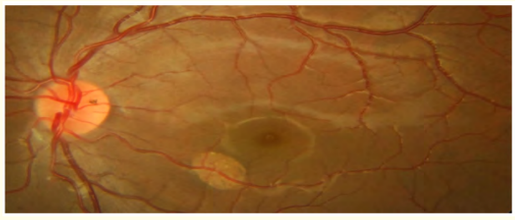

Figure 3: Magnified view of same Torpedo-like lesion.

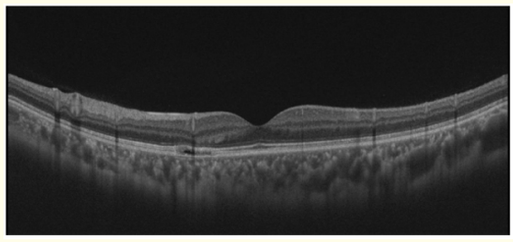

Figure 4: SS-OCT scan shows unaffected inner retinal layers, disorganization of outer plexiform layer(OPL) with reduced thickness of outer nuclear layer(ONL),slightly displaced intact external limiting membrane (ELM) , broadening and attenuation of ellipsoid zone, disrupted interdigitation zone, underlying thinned out intact RPE-Bruch complex with increased choroidal reflectivity.

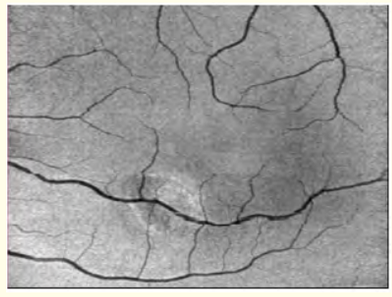

Figure 5: En Face structural OCT at the level of outer retina.

Figure 6: OCT Angiography at the level of choriocapillaries shows focal loss of choriocapillaries within the lesion.

Conclusion

The unusual nasal location represents a rare presentation of Torpedo maculopathy which is supported by multimodal imaging findings.

Acknowledgements

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Conflict of Interest

No financial interest or any conflict of interest exists.

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