

Case Report

Microvascular retinal changes in Torpedo Maculopathy

Pushpanjali Badole,* Smitesh Shah,* Amit S. Nene,* Sonal Shah,* Onkar H. Pirdankar¹

ABSTRACT

We are reporting a case of torpedo maculopathy (TM) over a long term follow up with the aid of multimodal imaging of Optical coherence tomography (OCT), Fundus Autofluorescence, En Face OCT, and recent OCT angiography (OCTA) findings. An asymptomatic 18 years old female was clinically found to be having a whitish spindle shaped lesion in the right eye with classic findings of TM. OCT imaging at baseline and at 2 and 4 years follow up did not reveal any change in the retinal structures. A focal excavation of the choroid along with a small subretinal cleft was also seen. Fundus autofluorescence imaging exhibits a central hypofluorescence along with a hyperfluorescent border corresponding to the lesion. OCTA segmentation of the choriocapillaris vasculature showed increased density of the choroidal vasculature suggestive of its role in the pathogenesis of this lesion. Enigma behind the pathogenesis of this rare lesion is still unsolved. Recently introduced imaging techniques can help us understand this lesion and its pathogenesis in detail.

Keywords: Torpedo Maculopathy, OCT angiography, Hypopigmented lesion, Retinal Microvasculature, Multimodal Imaging

Introduction

First described by Roseman and Gass in 1992 as an asymptomatic ‘hypopigmented nevus of the retinal pigment epithelium’, torpedo maculopathy is a rare, congenital and usually benign condition. [1] The classical presentation is of an unilateral, solitary, horizontally oval bullet or torpedo shaped hypopigmented lesion located in the temporal macula with its tip pointing towards the fovea. [2-5] Most patients are asymptomatic and have a normal visual acuity, the lesion being an incidental finding on routine fundus examinations. Intraocular pressures, pupillary reactions and ocular motility are unaffected, however scotomas corresponding to the torpedo lesion are common. [2-3] Paving way to its pathogenesis, it has been reported that torpedo maculopathy is associated with vascular alterations of the choriocapillaris along the lesion. [4] However there are limited reports describing the retinal microvasculature in

torpedo maculopathy. [4-5] Also to the best of our knowledge there is only one study which has described retinal changes over a long term follow up (five years) in patients with torpedo maculopathy. [6] Here we are reporting a case of torpedo maculopathy over a long term follow up along with recent OCT angiography findings.

Case details

Demographics and ocular clinical presentation:

An asymptomatic 18 years old female, during routine ophthalmic examination was found to have a whitish spindle shaped lesion in the fundus of her right eye. There was no significant past medical and ocular history. She denied any history of trauma. Results of Toxoplasmosis titer were negative. There was no history of any systemic illness, long term intake of hydroxychloroquine, and

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trauma. Visual acuity (VA) was 20/20 in both eyes. Slit lamp biomicroscopy and intraocular pressures were normal. Dilated fundus examination revealed an oval flat hypopigmented lesion around 1 disc diameter in size with well-defined margins located temporal to the macula slightly below the horizontal raphe in the right eye. The lesion was elongated with its tip pointing towards the fovea. There was slight hyperpigmentation at the temporal tail of the lesion and at superior and inferior lesion margin (Figure 1A). The findings in the left eye were unremarkable. The patient was followed up at two years (Figure 1B) and four years (Figure 1C) where no significant changes were noted in the shape, size or appearance of the lesion. OCT images were taken at baseline and at 2 and 4 years follow up. We did not find any change in the retinal structures. OCT findings at the latest follow up revealed thinning of the outer nuclear layer, an absence of the external limiting membrane along

with inner segment/outer segment junction. A focal excavation of the choroid along with a small subretinal cleft was also seen. Inner retinal layers presented normally without retinal cavitation, consistent with type I lesion (Figure 2A). Fundus autofluorescence imaging exhibits a central hypofluorescence along with a hyperfluorescent border corresponding to the lesion. The torpedo like structure was very distinct on fundus autofluorescence imaging (Figure 1D). OCTA segmentation of the choriocapillaris vasculature showed a hypo-reflective area (Figure 2B) corresponding to the hyper-pigmented end of the lesion temporally. Increased density of the choroidal vasculature was noted at the nasal end (Figure 2D). Also OCTA revealed decrease in the superficial blood vessels density (Figure 2C) (hyporeflexive area) (Figure 2B) and retinal thickness reduction in the area corresponding to the torpedo lesion.

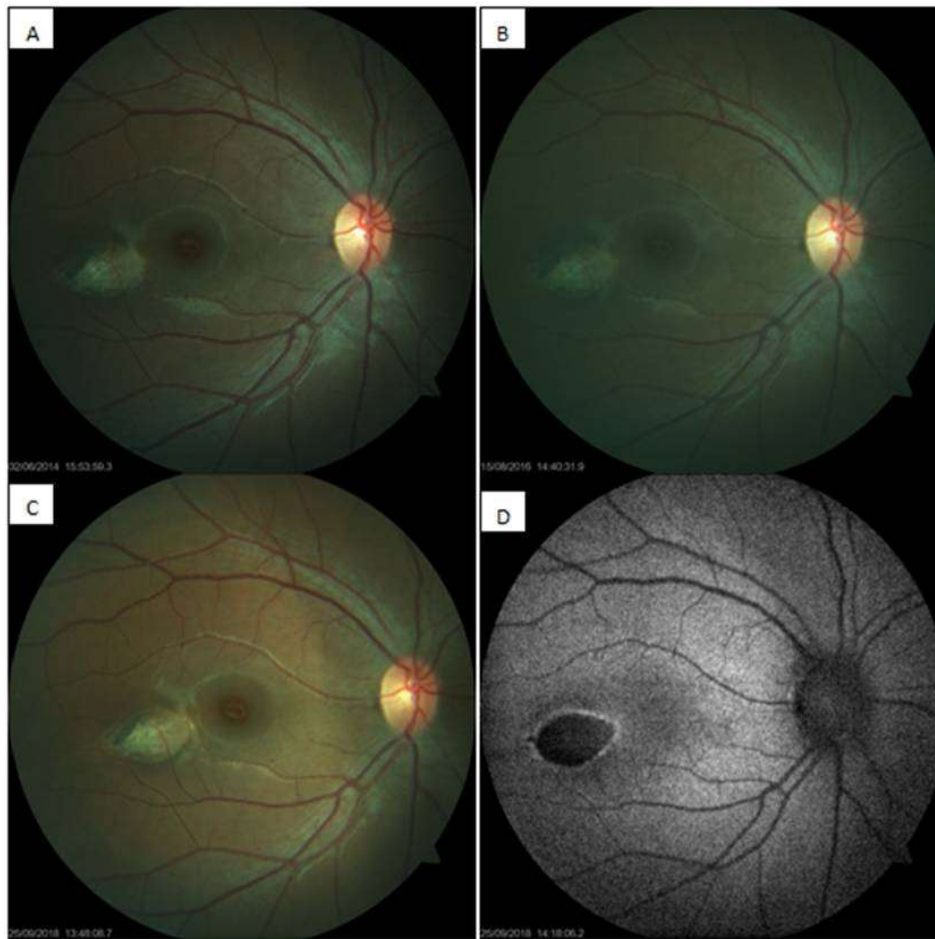


Figure 1: Figure-1 Fundus Image at (1A) Baseline, (1B) 2 years, (1C) 4 years and (1D) fundus autofluorescence

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Discussion

Our patient has the classic lesion of torpedo maculopathy as these lesions are mostly hypopigmented —although hyperpigmented lesions have also been noted previously. [6] Torpedo maculopathy is an asymptomatic condition and the diagnosis is merely based on fundus impression and age of onset. [2] It is typically seen along the horizontal raphe however it can also be located inferior to horizontal raphe as seen in our case or superior to horizontal raphe. [7] Lesion is usually unilateral and single although rare variations of bilateral [8] and double

torpedos [9] have recently been reported. Differential diagnoses to be ruled out are congenital hypertrophy of retinal pigment epithelium (CHRPE), simple hamartoma of the RPE and congenital toxoplasmosis, trauma as well as other chorioretinal scars. Though these lesions remain stable with no risk of visual loss but routine monitoring is advisable as rarely secondary choroidal neovascularization (cnvm) has been reported. [10]

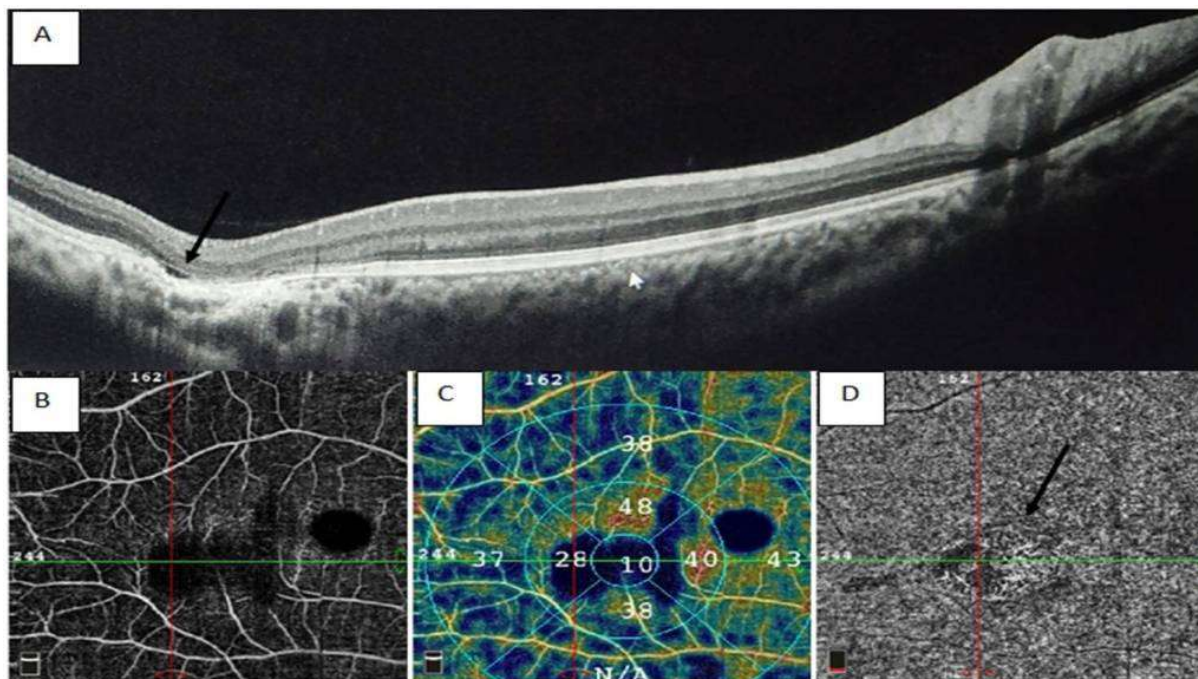


Figure 2: (2A) Focal excavation of the choroid along with a small subretinal cleft (2B) Hyperpigmented end of the torpedo lesion (2C) Reduction in the blood vessels density in the area corresponding to torpedo lesion (2D) Increased density of choroidal vasculature

Wong et al, have classified TM into two types according to the OCT findings. Type 1 has attenuation of outer retinal structures without outer retinal cavitation whereas type 2 has both attenuation and cavitation of outer retina. [11] Presence of outer retinal cavitation differentiates type 2 from type 1. Choroidal excavation and presence of subretinal cleft is a feature reported in recently published literatures [2]. Our case resembles type I since there is absence of retinal cavitation. [11] Recently a newer lesion has been proposed as type 3 which has the typical clinical appearance of TM and OCT features like inner retinal thinning, choroidal excavation but without subretinal

cleft which are features not classical of either type 1 or type 2 TM. [12] Although the torpedo lesion is non progressive, a previous report by Rohl A et al has reported a change in shape and size of torpedo lesion along with progression of a pseudo-lacuna within the lesion. [6] In present case we did not find any change in the retinal structure even after 4 years consistent with TM lesions. Also visual acuity remained the same. Torpedo maculopathy, rarely, can be associated with other ocular co-morbidities such as congenital glaucoma, branch retinal vein occlusion, choroidal nevus and neuro-sensory retinal detachment [3][13] however in our case we did not

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find any other ocular co-morbidity. Torpedo maculopathy is idiopathic in nature however various hypotheses have been proposed and includes developmental defect in the nerve fiber layer [14], abnormal choroidal or ciliary vasculature development [15] persistent developmental defect in the RPE in the fetal temporal bulge [16], congenital outpouching of the choroid [17], degenerative choroidal thinning and consequent separation of the photoreceptors from the RPE[2], and malformation of the emissary canal [3]. OCTA a recently introduced novel modality has opened gates for insights into the pathogenesis of torpedo lesions with the non invasive

Conclusion

A rare condition, torpedo maculopathy may sometimes cause a diagnostic dilemma. Multimodal imaging like OCT angiography may help us solve the mystery regarding the nature of disease and its pathogenesis.

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