Case Report

Torpedo Maculopathy with Satellite Lesion

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Abstract

Torpedo maculopathy is a rare congenital abnormality of the retinal pigment epithelium (RPE). It is characterized by unilateral, solitary, well circumscribed, hypopigmented, horizontally oval 'torpedo' shaped chorioretinal lesion. It should be considered as differential diagnosis for macular hypopigmented lesions. Considered as solitary, there have been rare case reports of double torpedo and satellite torpedo lesions. Here we report a case of Torpedo maculopathy with satellite lesion with its clinical features and Optical Coherence Tomography(OCT) findings.

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Keywords: *Torpedo Maculopathy, Satellite Lesion, Choroidal Excavation, Optical Coherence Tomography*

Introduction

Torpedo maculopathy is a rare congenital abnormality of the retinal pigment epithelium. The diagnosis of this entity is based on the clinical findings supported by investigations like OCT, Fundus autofluorescence and sometimes associated VF defect. Its first description dates back to 1992 when Roseman and Gass described a solitary, sharply circumscribed, oval, pinkish-white, placoid lesion along the horizontal raphe with a wedge-shaped "tail" at the level of the retinal pigment epithelium in the macula in a 12 year old boy, which was termed as a solitary 'hypo pigmented nevus of the RPE'.[1] This kind of lesion was later termed as 'Torpedo Maculopathy' by Daily because of its peculiar shape.[2] The exact aetiology and pathogenesis of these lesions is still not known. Although considered as rare, asymptomatic, non-progressive and benign; torpedo lesions may be associated with Choroidal Neovascular Membrane (CNVM). Hence they should be kept in mind as the differential diagnosis of well-defined hypo pigmented macular lesions.

Case Report

A 31 year old male came to our Retina clinic for opinion regarding macular scar in the left eye. He was using glasses since 15 years. His BCVA was 6/12 in the right eye and 6/18 in the left eye. Refraction showed -5.50 DS in right eye and -7.0 DS in left eye. Anterior segment was unremarkable in both the eyes. Dilated Fundus evaluation revealed a flat, horizontally oval, well defined hypopigmented chorioretinal lesion in temporal aspect of macula with the narrow fraying tail temporally (Figure 1). The lesion extended till the centre of the macula. The temporal end of the lesion showed hyperpigmentation. There was another smaller hypopigmented satellite lesion temporal to it. Rest of the peripheral retinal examination as well as the retinal examination of the right eye did not reveal any abnormality. OCT of the macula revealed thinning of the outer nuclear layer, attenuation of the ellipsoid zone and RPE with underlying hyper reflectivity of the choroid (Figure 2). The scan over the pigmented part of the lesion showed RPE

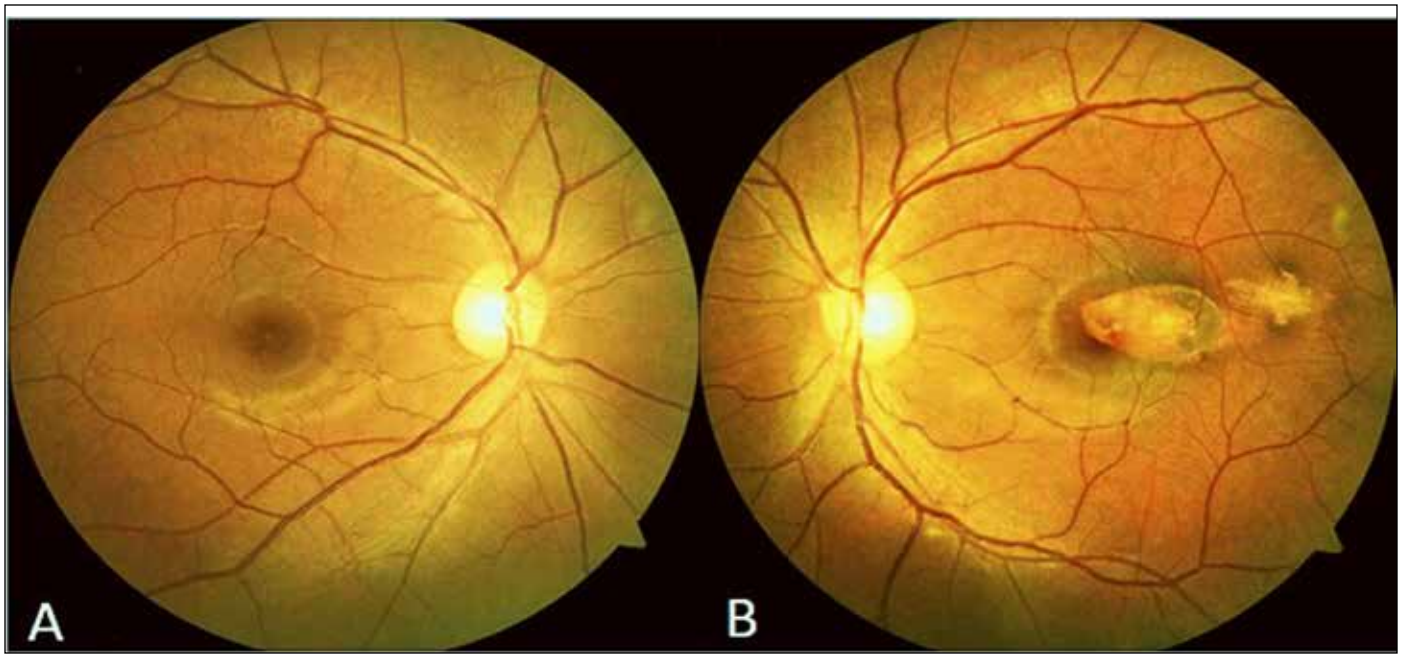
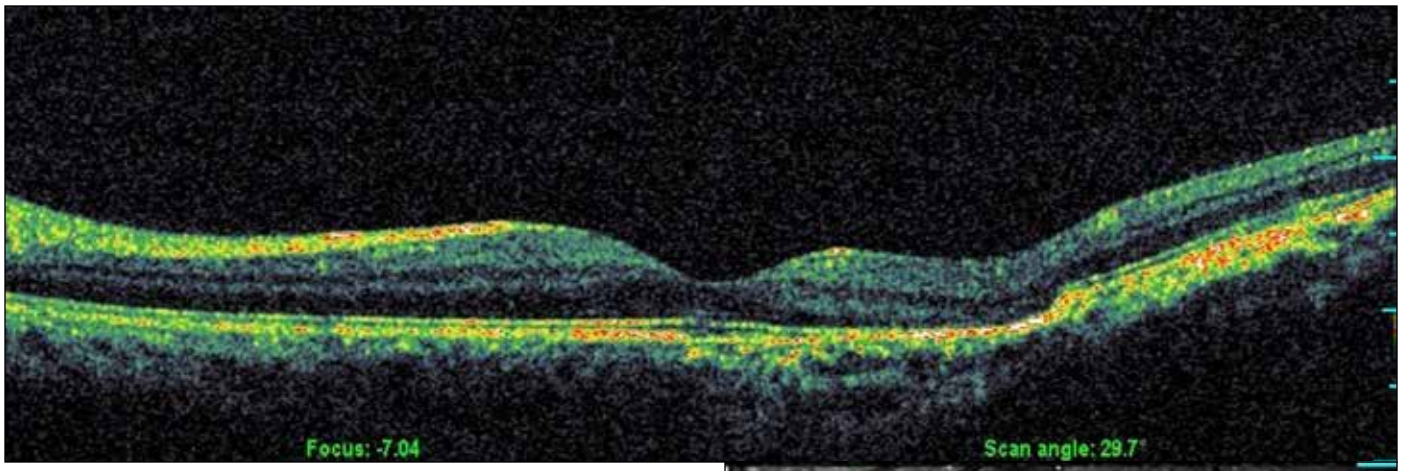


Figure 1: *Right eye fundus image showing normal fundus. B. Left eye fundus image showing Torpedo maculopathy with satellite lesion*



thickening with mild choroidal excavation in addition to the rest of the features. The satellite lesion revealed early disorganization of the ellipsoid zone.

Discussion

Torpedo maculopathy lesions are classically described as asymptomatic unilateral, solitary, hypopigmented lesions. Most of the times they are detected on routine clinical examination. These 'Torpedoes' are usually seen temporal to the macula, but atypical locations have also been reported [3,4]. Irrespective of the location of the lesion, the tip of the lesion always points towards the optic disc. Satellite lesion has been reported by very few authors[4,5].Visual acuity is not affected unless the lesion extends to fovea. Differential diagnosis to be considered are Congenital Hypertrophy of the Retinal Pigment Epithelium(CHRPE), Gardner syndrome, choroidal naevus, chorioretinal scars(toxoplasmosis, trauma), melanoma etc.

The exact aetiology of these lesions is not known. Various hypothesis have been postulated that include persistent defect in the development of RPE in the fetal temporal bulge,[6] abnormal choroidal vasculature,[3,7] developmental defect in the nerve fiber layer at the horizontal raphe,[8] and malformation of the emissary canal of the long posterior ciliary artery and nerve.[9]

Fundus Autofluorescence(FAF) may reveal hypo-autofluorescence, hyperautofluorescent boundaries , or a mixture of hyper and hypo autofluorescence. OCT in this case revealed thinning of the outer nuclear layer, ellipsoid zone, and RPE with underlying hyper reflectivity of the choroid. The area corresponding to the pigmented part of the lesion showed mild choroidal excavation. There was no subretinal cavitation or cleft which has been reported in some cases. Wang et al have classified these lesions into Type 1 and 2 where Type 2 lesions show outer retinal cavitation with or without inner choroidal excavation which is absent in Type 1 lesions[10]. Many times these lesions may show corresponding scotoma on visual field analysis.[10,11] There have been reports of CNVM associated with Torpedo maculopathy.[12,13,14]

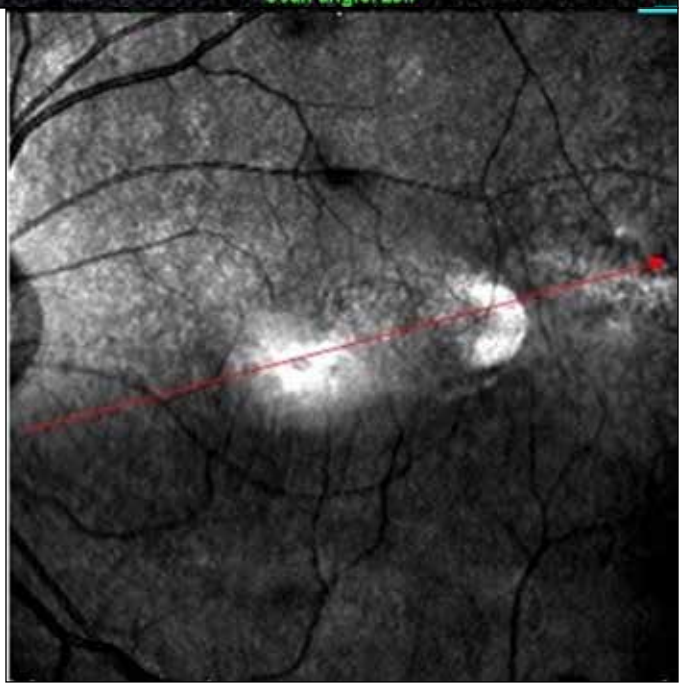


Figure 2: *OCT of the macula showing thinning of the outer nuclear layer, attenuation of the ellipsoid zone and RPE with underlying hyper reflectivity of the choroid. The scan over the pigmented part of the lesion shows RPE thickening with mild choroidal excavation.*

The diagnosis of this condition is mainly clinical with adjuvant investigations like OCT and FAF. Fluorescein Angiography can be done in case a CNVM is suspected. The current case shows similarities in terms of clinical features and imaging findings of Torpedo lesions. This case report however adds to the previously reported rare instances of satellite lesion and foveal involvement of torpedo maculopathy. Although classically described as solitary, our case report adds to the previously reported few cases of satellite lesion associated with Torpedo maculopathy which should be considered while making the clinical diagnosis of Torpedo maculopathy. These lesions should be followed up regularly in view of possibility of development of CNVM.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient has given his

consent for his images and other clinical information to be reported in the journal. The patient understands that his name and initials will not be published and due efforts will be made to conceal his identity, but anonymity cannot be guaranteed.

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