

# Torpedo Maculopathy

## Torpedo Maculopathy

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### ABSTRACT

To describe the optical coherence tomography (OCT) and fundus autofluorescence (FAF) properties of Torpedo maculopathy in a 10 year old boy with a complaint of recently developed oddness in his vision. Fundoscopy revealed hypopigmented lesion of the temporal macula in the right eye. Spectral domain OCT showed an abnormally thin retinal pigment epithelium, and relatively thickened retinal layers overlying this area. Choroidal transmission was increased. FAF image revealed isofluorescence of the lesion except for hyperautofluorescent border at the nasal side and hypofluorescent temporal margin. His visual acuity was 10/10 in both eyes (Snellen) with an auto-refraction (under cycloplegia) of +0.50, +0.50 axis 95 in the right eye, +0.75, +0.25 axis 90 in the left eye. Automated perimetry revealed no defect in the visual field defect. He was followed for three months with stable clinical findings.

**Key Words:** Torpedo maculopathy, spectral domain optical coherence tomography, fundus autofluorescence.

### ÖZ

Yakın zamanda başlayan görme bozukluğu şikayeti ile başvuran 10 yaşında erkek çocukta izlenen torpedo makulopatisinin optik koherens tomografi (OKT) ve fundus otofloresans (FAF) görüntüleri tarif edilmiştir. Fundoskopide sağ gözde temporal makulada hipopigmente lezyon gözlenmiştir. Spektral OKT de bu bölgede incelmış retina pigment epiteli ve üzerine göreceli olarak kalınlaşmış üst retina katları saptanmıştır. Koroid geçirgenliği artmış, FAF görüntülemesinde nazalde hiperotofloresan temporalde hipotofloresan sınırlı izotofloresan lezyon gözlenmiştir. Görme keskinliği her iki gözde 10/10 düzeyinde olup, sikloplejili refraksiyonu sağda +0.50, +0.50 95°, solda +0.75, +0.25 90° idi. Görme alanında defekt saptanmayan hasta 3 aylık takiplerinde stabil klinik seyir göstermiştir.

**Anahtar Kelimeler:** Torpido makulopati, spektral domain optik koherens tomografi, fundus otofloresansı.

### INTRODUCTION

Torpedo maculopathy (TM) was first described by Roseman and Gass in 1992 as a single hypopigmented congenital nevi of retinal pigment epithelium (RPE).<sup>1</sup> This congenital asymptomatic flat lesion is characterized by its torpedo-like tip directed toward the foveola, being oval in shape it may include hyperpigmented frayed tail temporally or a rounded margin.<sup>2</sup> It is usually detected on routine eye examination of asymptomatic patients.

In this case report our patient applied because of an oddness in his vision and suspicious purple discoloration of the lower eyelids. He was diagnosed as having torpedo maculopathy with otherwise normal ophthalmic and systemic findings.

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## CASE REPORT

The patient was a 10-year-old boy who complained of a change in his vision and a purple discoloration of both lower eyelids for about a few days. There was no history of trauma, allergy, systemic disease or fever. Ocular motility was normal in all directions. There was orthophoria in primary position. Visual acuities were 10/10 in both eyes without correction. Cycloplegic auto-refraction was +0.50, +0.50 axis 95 in the right eye, +0.75, +0.25 axis 90 in the left eye. There were no signs of ocular allergy. There was a relative unremarkable blue-purple discoloration of both lower eyelids. He was under antidepressant therapy with a selective serotonin reuptake inhibitor (Sertraline HCL). Hematological and biochemical blood tests were found to be within normal limits. Anterior segment revealed normal findings. On funduscopy there was a hypopigmented oval-shaped flat lesion located temporal to the fovea. The lesion was 1 disk diameter (DD) horizontally,  $\frac{1}{2}$  DD vertically and was located  $\frac{1}{4}$  DD infero-temporal to the fovea (Figure 1). The fellow eye was unremarkable.

Fundus autofluorescence using short wavelength resulted in isofluorescence of the lesion except for hyperfluorescent border of the nasal margin and hypofluorescence at the temporal tail area (Figure 2). OCT scans (using Topcon 3D OCT-2000 FA plus) showed abnormally thin retinal pigment epithelium (RPE) hyperreflectivity and an increased signal transmission in the choroid under the lesion. The overlying outer and inner retina were normal except for a relative bulging appearance toward the choroid. (Figure 3) Visual field analysis using Humphrey visual field analyzer was unremarkable.

This pathological condition was diagnosed as Torpedo maculopathy and the patient was followed for 3 months without any clinical and functional change.

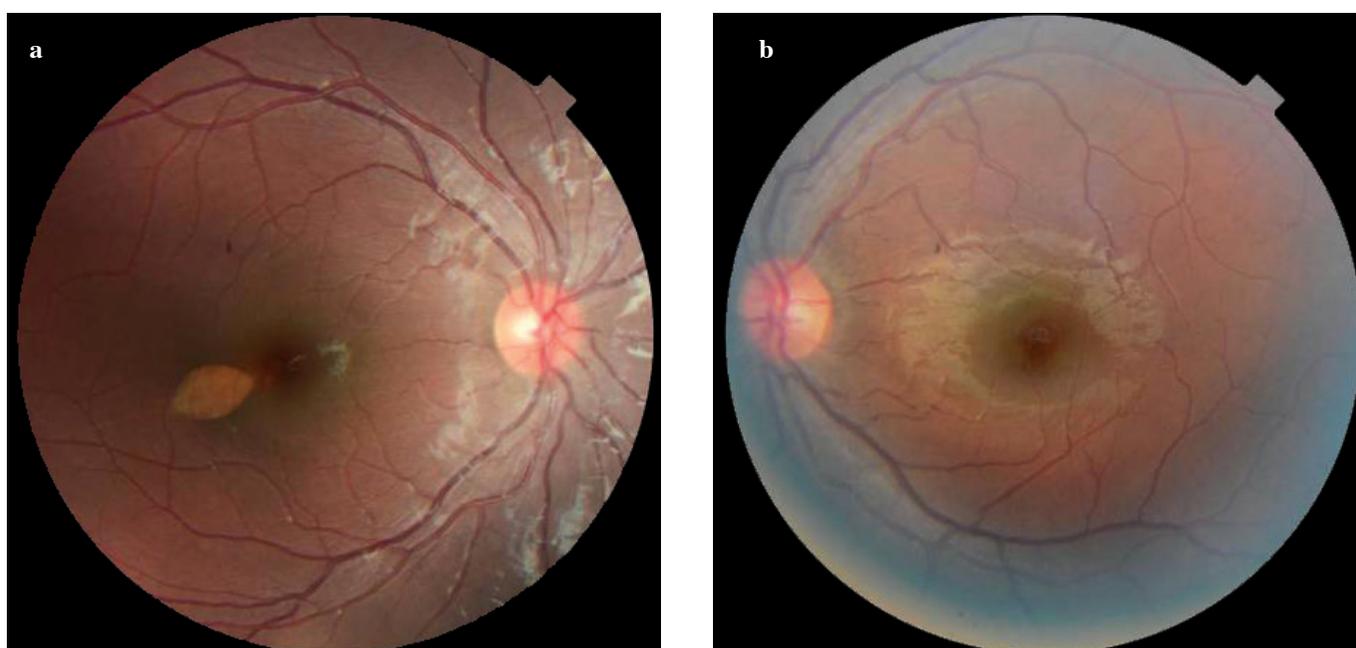
## DISCUSSION

TM is seen as a non-pigmented pointed oval macular lesion with well-defined borders and with a tip toward the foveola and a tail toward the ora serrata. Nasal margin has a sharp point and the temporal margin may either be round,<sup>1,8</sup> as in our case or with frayed tail appearance with linear or spotty pigmentation.<sup>4,8</sup>

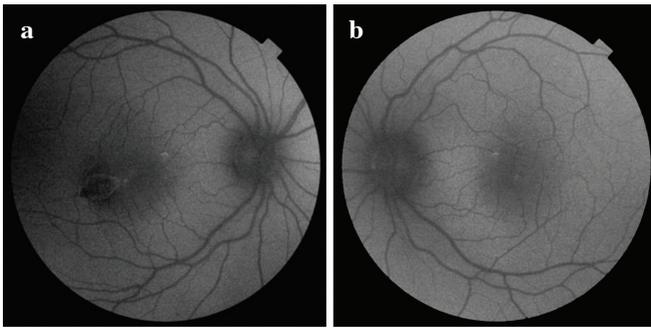
Etiology may be related to the fetal temporal macular bulge that normally occurs at 4-6 months' gestation at the same site,<sup>8</sup> it may represent a persistent defect in the development of RPE in the fetal temporal bulge. Alterations in the choroidal vasculature in the macular area during embryonic development of the eye may be considered as another etiological factor.<sup>7</sup>

In TM, OCT findings include abnormally thin RPE signal, increased signal transmission in the choroid corresponding to the torpedo lesion with no neuroretinal changes.<sup>5,6</sup> Short wave FAF present normal fluorescence of the lesion except for a small hyperfluorescent area at the tail level just as we have observed in our patient.<sup>5</sup> Near infrared NIR-FAF shows hypoautofluorescence at the lesion.<sup>6</sup> Fundus fluorescein is not needed for diagnosis but transmission hyperfluorescence of the lesion was shown in a case series by Golchet et al.<sup>6</sup>

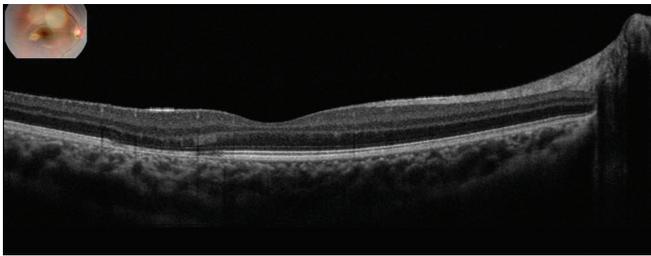
Torpedo maculopathy may be seen as an intraretinal cleft showing fundus excavation<sup>2,9</sup> (recently named Type 2 TM) causing scotoma at the excavation area and decrease of visual acuity if the lesion encroaches upon the central fovea.



**Figure 1a,b:** Fundus photos showing a non-pigmented oval-shaped lesion with clearly defined margins located infero-temporal to the macula with a tip toward the foveola in the right eye (a), and normal fundus features in the left eye (b).



**Figure 2a,b:** Conventional fundus autofluorescence (FAF) image revealed normal autofluorescence of the lesion except for a small hypofluorescent area at the tail pointing to the ora serrata and hyperfluorescence at the part next to foveola in the right eye (a), and normal autofluorescence features in the left eye (b).



**Figure 3:** Optical coherence tomography (OCT) 9mm line section through the lesion revealed normal neuroretinal findings with increased choroidal transmission corresponding to the lesion and decreased retinal pigment epithelial hyperreflectivity. IS/OS and ELM lines can be seen uninterrupted with a relative outer retinal bulging toward the choroid at the site of the lesion.

Accompanying sensory retinal detachment with paracentral nasal defect in a patient with complaint of intermittent floating spots was reported.<sup>10</sup> Buzzonetti et al. reported sensory retinal detachment in a 6-year-old child also reported with reduced multifocal ERG and pattern VEP response.<sup>11</sup> Macular microperimetry showed reduced retinal sensitivity along the pigmented margin of the lesion with normal values over the lesion in another report.<sup>5</sup> We observed no sensorial detachment over the lesion and visual field analysis was unremarkable.

TM has unique clinical and imaging features but in the differential diagnosis melanoma and nevus, congenital or iatrogenic hyperplasia of retinal pigment epithelium (CHRPE) and focal retinal pigmentation due to traumatic or drug induced toxic effects should be considered.<sup>3,4</sup> CHRPE is seen as hyperpigmented lesions with or without accompanying depigmented lacunae or a hypopigmented halo, whereas Gardner lesions may have torpedo-like hypopigmented lesions, they are usually bilateral, multiple and midperipherally located.

In this case report we presented the imaging characteristics of TM in our patient. Being a congenital benign retinal pathology TM rarely threatens vision. There is usually no need for treatment, but yearly ophthalmological examination is recommended.

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