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# Torpedo Maculopathy

Torpedo maculopathy (TM) is a congenital, benign, and asymptomatic lesion often diagnosed incidentally on routine examination. TM was first reported in 1992 as a flat hypopigmented nevus of retinal pigment epithelium (RPE) in a 12-year-old boy. [1] This classically solitary, oval, torpedo-shaped RPE lesion occurs in the temporal macula with its torpedo-like tip pointed towards the foveola and its tail directed toward the periphery.[1-3] If present, satellite lesions are smaller, flat, and temporally located (Figure 1). [3] The lesion is often stationary; however, enlargement over time has been documented. [2-3] Choroidal neovascularization has been reported, but is responsive to anti-vascular endothelial growth factor therapy.[3] Shirley and colleagues described the largest case series of 8 pediatric patients with TM between the ages of 3 to 15 years of age. [3] Mean age of diagnosis was 8 years with a slight female predominance. The prevalence in the Northern Ireland population was approximately 2 per 100,000 under age 16, though this is likely to be an underestimation as the condition is asymptomatic. [3] There are no definitive systemic associations, though a possible link to tuberous sclerosis and astrocytic hamartoma have been described.[4]

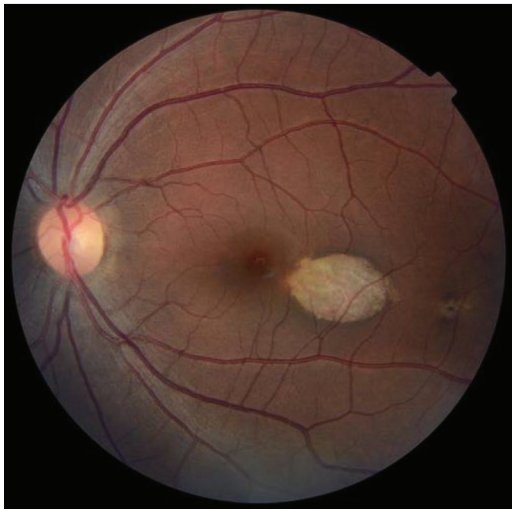

**Figure 1:**
Hypopigmented torpedo shaped lesion at the level of RPE with overlying retinal vessel and satellite site

In the limited case series on TM in the literature, patients have had age-appropriate vision, ranging from fix and follow to 20/20.[2-3] TM lesions typically measure 2-3mm horizontally and 1-1.5mm vertically with a nasal tip 0.2-1mm from foveola (Figure 2). [2] On optical coherence tomography (OCT), there are two subtypes of TM described by Wong and colleagues. Both subtypes have preservation of inner retinal layers, attenuation of outer retinal structures, and increased transmission signal through the choroid.[5] Type 1 lacks any outer retinal cavitation or neurosensory retinal elevation, which is seen in Type 2.[5] It has been postulated that Type 1 lesions transition into Type 2 over several decades; however, Shirley and colleagues' cohort of patients with Type 1 and Type 2 lesions were on average age 8 and 7, respectively.[3,5] Satellite lesions may have normal outer retinal layers or minimal attenuation in comparison to the main lesion.[2]

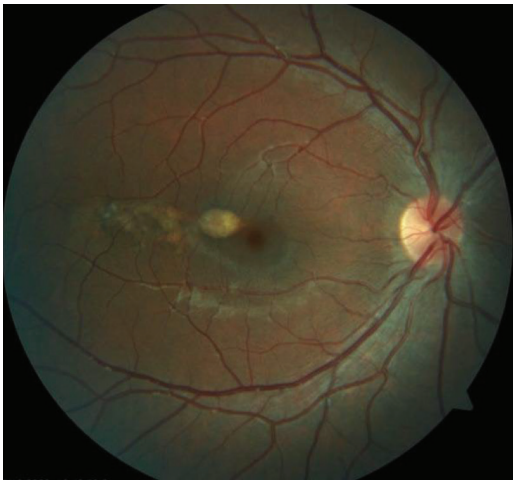

**Figure 2:**
Smaller bullet shaped lesion with adjacent RPE irregularities

Other ancillary imaging can be helpful in characterizing and diagnosing TM. Fundus autofluorescence will often show hyperautofluorescent lesion borders secondary to excessive accumulation of lipofuscin in affected RPE cells, and hypoautofluorescent patches from RPE loss (Figure 3). Fluorescein angiogram shows hyperfluorescence corresponding to the clinical lesion with possible associated hypofluorescence if RPE hyperpigmentation is present. OCT angiography shows normal superficial retinal plexus, but possible attenuation of deep capillary plexus or choriocapillaris[6] Microperimetry may show reduced sensitivity at the site of TM which remains stable with time.[6]

The differential diagnosis for TM includes congenital lesions such as macular coloboma, congenital hypertrophy of RPE (CHRPE), congenital RPE albinotic spots ("polar bear tracks"), chorioretinal scar from infection (i.e. toxoplasmosis), inflammation, or trauma, congenital RPE hamartoma, combined PE-retinal hamartoma, and RPE hyperplasia associated with familial adenomatous polyposis. A detailed medical, including prenatal and neonatal, history can help to narrow the differential. In addition, isolated lesions like CHRPE have a random distribution, rounded appearance, and typically is seen anterior to the equator. Likewise, the RPE abnormalities associated with familial adenomatous polyposis are more randomly distributed in the fundus and tend to be smaller or more irregular in shape.[2-3]

The etiology of TM remains speculative. Leading hypotheses include a defect in the closure of the fetal temporal macular "bulge" normally present during months 4 to 6 of gestation at the same anatomical site, abnormal choroidal development or ciliaryvascular development leading to RPE underdevelopment, and possible sequela of intrauterine infection or inflammation.[2-3,5]

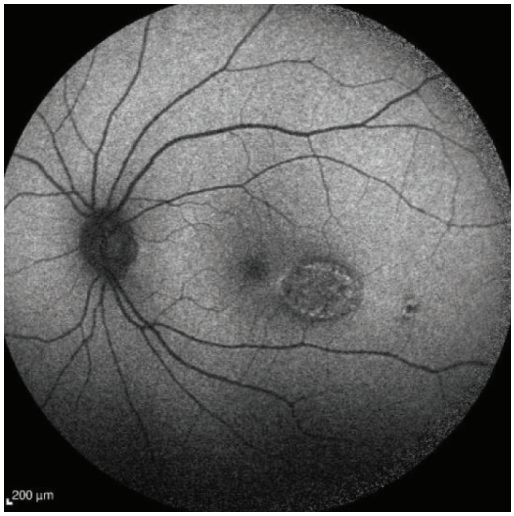


**Figure 3:**
Fundus autofluorescence corresponding to lesion in Figure 1. There is patchy hyperautofluorescence and hypoautofluorescence of both main torpedo and satellite lesions

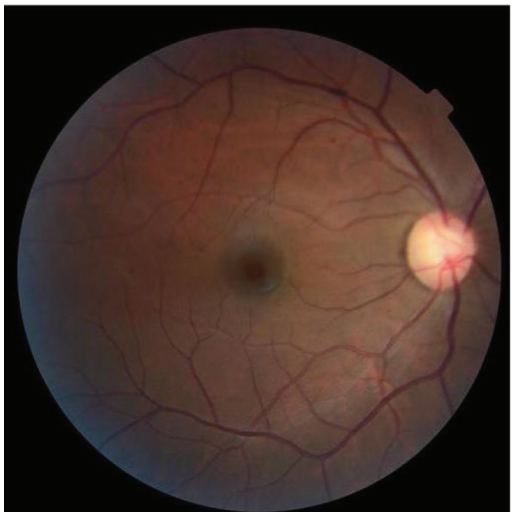

**Figure 4:**
Fundus photo of unaffected right eye (visual acuity 20/20)

Below is a typical presentation of a patient with TM. The patient is a 28-year-old male who presented for retinal consultation secondary to a macular lesion. He denied any ocular history except for new bilateral blurriness which prompted him to see his optometrist. With refraction, he was 20/20 with -1.00 D prescription bilaterally with resolution of ocular complaint. He denied any scotoma or metamorphopsia. On examination, he had unremarkable anterior segments bilaterally. Right fundus exam showed healthy optic nerve, vessels, macula and periphery (Figure 4). Left fundus showed normal optic nerve, vessels, and a flat, torpedo shaped hypopigmented RPE lesion in temporal macula with rounded hyperpigmented tail (Figure 5). There was no subretinal or RPE fluid, hemorrhage, or choroidal neovascular membrane. On OCT, the right eye was normal (Figure 6), but the left eye had a characteristic Type 1 lesion with thinning of outer nuclear layer, attenuation of the interdigitation and ellipsoid zones, and increased posterior hyperreflectivity (Figure 7). There was no choroidal neovascular membrane or subretinal fluid noted. Observation and Amsler grid monitoring were advised for this patient. He will follow up on a semi-annual basis to monitor for CNV activity.

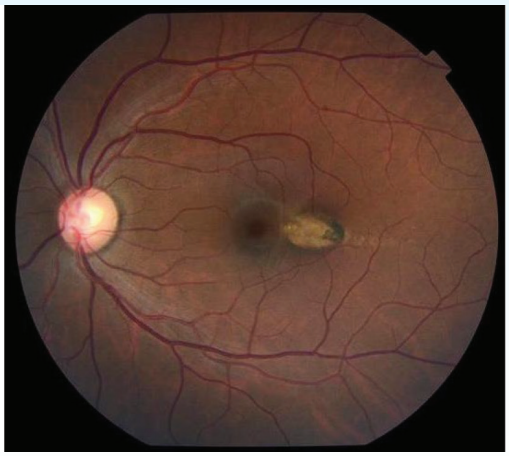

**Figure 5:**
Fundus photo of left eye with torpedo maculopathy (visual acuity 20/20 vision)

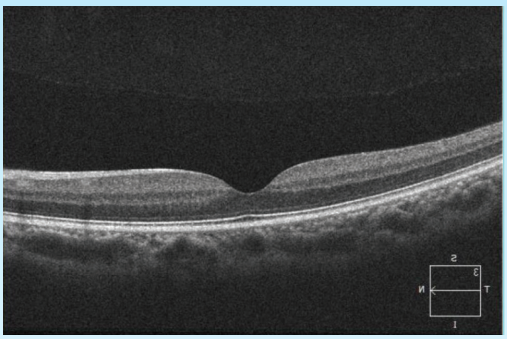

**Figure 6:**
Normal OCT of the right eye including normal outer retinal layers

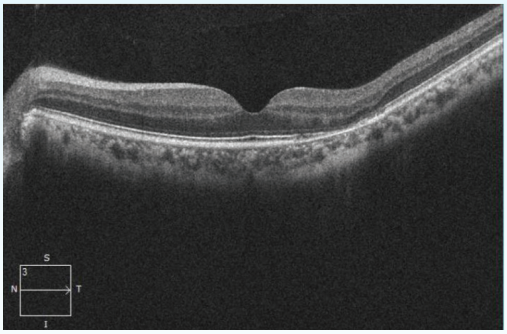

**Figure 7:**
OCT showing Type 1 lesion with thinning of outer nuclear layer, attenuation ofthe interdigitation and ellipsoid zones


***References:***

1. *Roseman RL, Gass JD. Solitary hypopigmented nevus of the retinal pigment epithelium in the macula. Arch Ophthalmol. 1992; 110:1762.*
2. *Shields CL, Guzman JM, Shapiro MJ, Fogel LE, Shields JA. Torpedo maculopathy at the site of the fetal bulge. Arch Ophthalmol. 2010; 128:499-501*
3. *Shirley K, O'Neill M, Gamble R, Ramsey A, McLoone E. Torpedo maculopathy: disease spectrum and associated choroidal neovascularization in a paediatric population. Eye. 2018; 32:1315-20.*
4. *Hansen MS, Larsen M, Hove MN. Optical coherence tomography of torpedo maculopathy in a patient with tuberous sclerosis. Acta Ophthalmol. 2016; 94(7): 736-737.*
5. *Wong E, Fraser-Bell S, Hunyor A, Chen F. Novel optical coherence tomography classification of torpedo maculopathy. Clin Exp Ophthalmol. 2015; 43:342-8*
6. *Grimaldi G, Scupola A, Sammarco MG, Marullo M, Blasi MA. Morpho-functional evaluation of torpedo maculopathy with optical coherence tomography angiography and microperimetry. Am J Ophthal Case Rep. 2018; 10:165-8.*


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# At the **forefront** of **clinical research**

NJRetina continuously conducts clinical trials at key locations. Our clinical research coordinators will be happy to discuss the inclusion/ exclusion criteria or any other aspect of these studies with you or your patients. If you have any questions, please feel free to contact:

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# **Enrolling** Studies:

**Dry AMD**

**Vauxhall**

**GTSCOPE:** A Study of Disease Progression in Genetically Defined Subjects With Geographic Atrophy Secondary to Age-Related Macular Degeneration

**Teaneck**

**Catalina:** A Phase 2 Multicenter, Randomized, Double-Masked, Sham-Controlled Study of the Safety and Efficacy of Intravitreal Injections of NGM621 in Subjects with Geographic Atrophy (GA) Secondary to Age-Related Macular Degeneration (AMD)

**Teaneck and Toms River**

**Gallego:** A phase II, multicenter, randomized, single-masked, sham-controlled study to assess safety, tolerability, and efficacy of intravitreal injections of FHTR2163 in patients with geographic atrophy secondary to age-related macular degeneration (Gallego)

**Wet AMD**

**Edison**

**Pulsar:** Randomized, Double-Masked, Active-Controlled Phase 3 Study of the Efficacy and Safety of High Dose Aflibercept in Patients with Neovascular Age-Related Macular Degeneration

**Diabetic Macular Edema (DME)**

**Teaneck**

**Gleam:** A prospective, randomized, double-masked, active comparator-controlled, multi-center, two- arm, phase 3 study to evaluate the efficacy and safety of intravitreal KSI-301 compared with intravitreal aflibercept in participants with **visual impairment secondary to treatment- naive diabetic macular edema.**

**Teaneck**

**Pagoda:** A Phase III, multicenter, randomized, visual assessor-masked, active-comparator study of the efficacy, safety, and pharmacokinetics of the Port Delivery system with Ranibzumab in patients with diabetic macular edema

**Diabetic Retinopathy**

**Teaneck**

**Pavilion:** A Phase III, Multicenter, Randomized Study of the Efficacy, Safety, and Pharmacokinetics of the Port Delivery System with Ranibizumab in Patients with Diabetic Retinopathy

## Soon to Enroll Studies:

**Diabetic Retinopathy:**

- **Altitude**: A Phase 2, Randomized, Dose-escalation, Observation-controlled Study to Evaluate the Efficacy, Safety, and Tolerability of RGX-314 Gene Therapy Delivered via One or Two Suprachoroidal Space (SCS) Injections in Participants with Diabetic Retinopathy (DR) Without Center Involved-Diabetic Macular Edema (CI-DME) (ALTITUDE) – Teaneck

**Retinal Vein Occlusion**

- **Balaton**: A Phase III, Multicenter, Randomized, Double-Masked, Active Comparator-controlled Study To Evaluate The Efficacy And Safety Of Faricimab In Patients With Macular Edema Secondary To Branch Retinal Vein Occlusion – Toms River