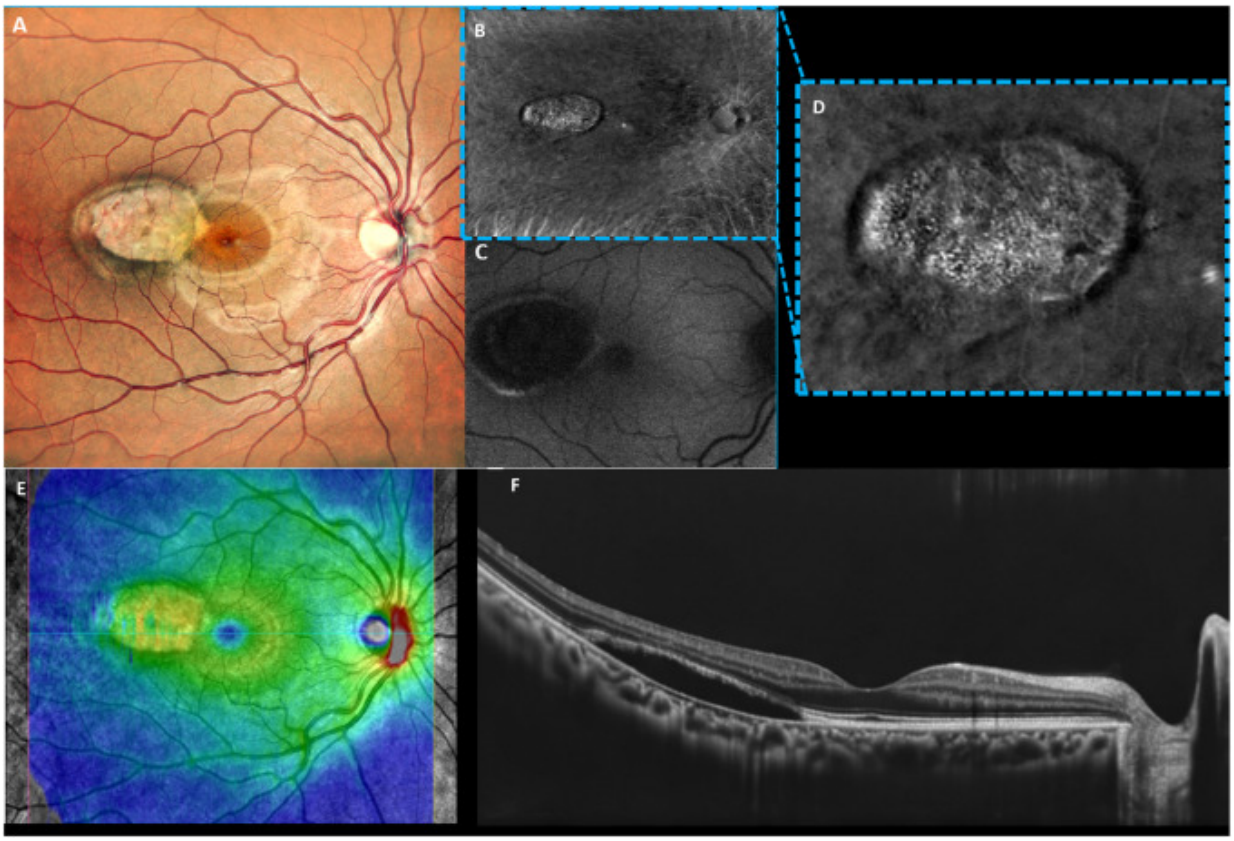
Torpedo maculopathy: multimodal and retromodal imaging



Torpedo maculopathy (TM) was described initially by Rosemann and Gass in 1992 as a torpedo-shaped hypopigmented lesion located in the temporal region of the macula. TM is a rare, benign, and well-defined congenital disease affecting the retinal pigment epithelium (RPE). It typically presents as a solitary, asymptomatic, hypopigmented nevus in the temporal macula, resembling a torpedo-shaped lesion. The name *torpedo maculopathy* was coined by Daily in 1993 because of its distinctive appearance. The lesion was described as congenital, oriented toward the macula, and consistently localized in the temporal region adjacent to the fovea.

In classic fundus examination, TM manifests as an oval-shaped hypopigmented lesion resembling a bullet or torpedo with a wedge-shaped tail extending outward and pointing toward the foveola along the horizontal raphe. TM is likely underestimated because patients are typically asymptomatic, and these lesions are often detected during routine ophthalmologic examinations. Additionally, due to their nonprogressive and generally benign nature, no treatment is required.

TM should be distinguished from other etiologies of hypopigmented retinal lesions based on history and clinical findings. Retinal scars are frequently associated with a history of trauma, infection, or inflammatory condition.

Both optical coherence tomography angiography (OCT-A) and retromode imaging (RMI) are novel non-invasive imaging techniques. While OCT-A provides detailed *en face* structural and flow information on the retinal and choroidal circulation, RMI employs infrared light wavelengths and a confocal scanning laser ophthalmoscope to generate a simulated pseudo-3-dimensional image, providing additional detailed visualization of profound retinal structures.

In this study, we used non-invasive multimodal imaging, including RMI, to describe a case of TM. To the best of our knowledge, this is the first report using RMI in the evaluation of TM.

A 29-year-old female was referred to our ophthalmology department following the discovery of a suspected central serous chorioretinopathy in her right eye during a routine examination. The patient did not present with any other

symptoms and did not experience pain or vision loss in her right eye. There was no history of trauma, and her past medical and ophthalmic records were unremarkable.

The initial best-corrected visual acuity was measured as 20/20 in both eyes. The anterior-segment examination revealed no abnormalities, and the pupils were equal with no signs of afferent pupillary defect. Initial intraocular pressure measurements were 12 mm Hg in the right eye and 14 mm Hg in the left eye.

During the funduscopic examination of the right eye, an oval-shaped, well-defined hypopigmented lesion surrounded by a hyperpigmented area was observed in the temporal sector of the macula. The lesion exhibited an orientation with its tip directed toward the central fovea and its tail extending toward the temporal retina. The lesion appeared elongated in the horizontal axis and revealed normal underlying choroidal details. Conversely, the posterior pole of the left eye appeared normal without any pathologic changes.

To further evaluate the condition, we conducted multimodal imaging using the NIDEK Mirante (Nidek Company, Gamagori, Japan), including multicolor imaging, fundus autofluorescence (FAF), spectral-domain OCT (SD-OCT), and RMI (Fig. 1).

Fig. 1(A) Multicolour photography of the right eye showing an oval hypopigmented lesion surrounded by a hyperpigmented area on the temporal sector of the macula. (B, D) Retromode imaging using left deviation showing a furrowed lesion temporal to the macula and fundus. (C) Autofluorescence shows hypo-autofluorescence with a hyper-autofluorescent border. (E) Under the application of near-infrared and retina mapping, we observed depressed topography of the lesion. (F) Spectral-domain optical coherence tomography showed a neurosensory detachment defect of the outer retina with thinning of the outer nuclear layer and loss of both the ellipsoid and interdigitation zone resulting in a subretinal cleft.

We also performed a swept-source OCT-A using the PLEX Elite 9000 (Carl Zeiss Meditec Inc, Dublin, Calif.).

Multicolour imaging (Fig. 1**A**) of the right eye revealed a spindle-shaped yellowish-white hypopigmented lesion surrounded by a hyperpigmented ring located in the temporal macular area, with its tip pointing toward the central fovea of the macula. FAF (Fig. 1**C**) exhibited a marked hypo-FAF signal at the lesion with a rim of hyper-FAF at the margin.
The SD-OCT (Fig. 1**E, F**) performed by the NIDEK Mirante (Nidek Company) through the lesion showed a normal internal retina, a slightly thinner external retina, and RPE in the temporal macular area, with an overlapping neurosensory detachment and attenuation of the ellipsoid and interdigitation zone area.
Implementation of the RMI technique using leftward deviation in the right eye (Fig. 1**B, D**) revealed a heterogeneously hyperreflective punctate pattern with a centrally depressed oval-shaped crater exhibiting a well-defined contour. This central characteristic of heightened reflectivity corresponds to Sattler's layer, which is visualized due to a lack of the choriocapillaris layer. The encompassing region of diminished reflectivity is linked to the hyperpigmentation surrounding the lesion. Visualization of the choroidal layer through a window defect indicates the absence of fluid beneath the neurosensory detachment.
The swept-source OCT-A device (PLEX Elite 9000; Carl Zeiss Meditec) was used to evaluate the morphologic characteristics of the torpedo lesion. OCT-A 6×6 mm view of the right eye showed a normal superficial and deep vascular complex. OCT-A at the level of the outer retina to choriocapillaris segmentation revealed a signal attenuation corresponding to the neurosensory

detachment. At the level of the choriocapillaris, a central high-flow area corresponding to the visualization of the Sattler layer secondary to the absence of the choriocapillaris was seen, surrounded by an area of signal attenuation in the area of hyperpigmentation (Fig. 2).

Fig. 2Swept-source optical coherence tomography angiography of the right eye showed a normal superficial and deep vascular plexus at the choriocapillaris level unveiling an innermost intense circulation region within the torpedo anomaly, aligning with the observation of the Sattler layer due to the lack of choriocapillaris, surrounded by an area of signal attenuation in the area of hyperpigmentation. SVC, superficial vascular complex; DVC, deep vascular complex; ORCC, outer retinal to choriocapillaris; CC, choriocapillaris.

TM is a rare congenital anomaly usually found in young adults. It is typically located temporally to the fovea, generally does not cause symptoms, and is linked to normal visual acuity. However, in some cases, outer retinal disruption may be observed, and this may result in visual impairment. Other reported complications include choroidal neovascularization, central serous chorioretinopathy, and corresponding visual field scotoma.

In the setting of a multimodal approach, we prefer the following approach:

The fundus color image shows hypopigmented lesions of varying degrees, elongated horizontally, with hyperpigmented RPE changes around them, located in the temporal macula. Patients with separate, smaller lesions called *satellite lesions* also have been described. FAF revealed central hypo-FAF (nonfunctional RPE) with slight hyper-FAF along the outline of the lesion. Based on OCT findings, in 2014, Wong et al. described 2 types of torpedo maculopathies. Type 1 demonstrates attenuation of the outer retinal structures without outer retinal cavitation, whereas

type 2 exhibits both attenuation of the outer retinal structures and outer retinal cavitation. Furthermore, Shirley et al. reported an 8-case series of TM in a pediatric population, and the authors noted that the average age of patients with type 1 and type 2 lesions was 8 and 7 years, respectively, suggesting 2 distinct phenotypic entities that can occur at a young age. In 2018, Tripathy et al. described a type 3 of TM in a case report where the OCT showed a choroidal excavation accompanied by degeneration of the outer retina, and the inner retinal layers had mild disorganization and a subretinal cleft. More recently, Light et al. expanded the classification by describing a type 4 phenotype characterized by a preserved ellipsoid zone, absence of subretinal fluid, and choroidal excavation.

In our case, OCT-A of the choroidal capillary segment revealed an increased density of choroidal vasculature, indicating a thinner RPE and increased choroidal thickness due to enhanced optical signal transmission. However, the superficial and deep layers appeared normal. Choroidal neovascularization is a rare occurrence, and OCT-A can be useful in identifying potential choroidal neovascularization development.

Using RMI, we observed a depression in the lesion with distinct demarcation, providing a heterogeneous image that demonstrates the RPE and choroid layers while showing the absence of subretinal fluid. By employing this technique, we can confirm the absence of subretinal fluid in the space under the neurosensory retina, helping to clarify some theories regarding the suspicion of subretinal fluid in these lesions.

We describe a case of TM in a 29-year-old female without associated pathologies and in whom we used multimodality

imaging, together, for the first time, with a novel RMI technique for this pathology. We recommend a period of long-term follow-up with multimodal imaging, including RMI, for documentation of the stage and to clarify and provide deeper insights into the pathophysiology theories of TM.

Footnotes and Disclosure

Declaration of patient consent: The authors certify that they have obtained all appropriate patient consent forms. In the form, patient provide their consent for images and other clinical information to be reported in this journal. Patients understand that their names and initials will not be published, that due efforts will be made to conceal their identity, but that anonymity cannot be guaranteed.

Conflicts of interest: The authors have no proprietary or commercial interest in any materials discussed in this article.

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Figures

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