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Multifocal Torpedo Maculopathy Complicated by Choroidal Neovascularization

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Abstract

Purpose: To present a pediatric patient with a unique configuration of torpedo maculopathy complicated by macular choroidal neovascularization (CNV). **Methods:** A single case was retrospectively reviewed. **Results:** An 8-year-old male child presented with decreased vision in the left eye and was found to have 2 distinct torpedo maculopathy lesions, a smaller hypopigmented lesion in the temporal parafovea and a larger hyperpigmented comet-shaped lesion in the temporal periphery. Multimodal imaging showed active CNV. The patient received 2 intravitreal injections of ranibizumab with regression of CNV and recovery of visual acuity. **Conclusions:** CNV is a rare complication of torpedo maculopathy that can affect pediatric patients in the absence of choroidal excavation. The presence of a hyperpigmented peripheral lesion exhibiting symmetry across the horizontal raphe lends support to the hypothesis that an alteration in the development and migration of retinal pigment epithelium cells across the fetal bulge results in this disorder.

Keywords

torpedo maculopathy, choroidal neovascularization, retinal pigment epithelium, anti-VEGF agents, pediatric retina

Introduction

Torpedo maculopathy is a rare entity, with fewer than 100 peer-reviewed cases published to date.[1] It is classically characterized as a hypopigmented torpedo-shaped lesion located in the temporal macula. Its etiology remains elusive; however, hypotheses include incomplete differentiation of the arcuate nerve fiber layer bundles, a developmental defect of the retinal pigment epithelium (RPE) within the "fetal bulge," and malformation of the emissary canal of the long posterior ciliary artery and nerve.[2–4] Although traditionally regarded as a benign condition with stable visual outcomes, torpedo maculopathy can occasionally be complicated by the development of choroidal neovascularization (CNV), leading to significant visual impairment and necessitating prompt intervention.

Here, we present what to our knowledge is the youngest case reported to date of a pediatric patient with torpedo maculopathy complicated by CNV. In addition, we describe a unique configuration of the disorder, with 2 distinct lesions, a smaller hypopigmented parafoveal lesion and a larger hyperpigmented comet-shaped lesion in the temporal periphery.

Case Report

An 8-year-old previously healthy White male child was referred for a macular scar in the left eye. He reported a 1-month history of decreased vision in the left eye accompanied by a left inward deviation of the left eye, as noted by his mother. He was born full-term with no significant perinatal history, and a review of systems, medical, social, and family history was unremarkable other than exposure to cats and dogs at home.

The patient's best-corrected visual acuity (BCVA) was 20/20 OD and 20/200 OS. An external examination found a left esotropia with full extraocular motility. The intraocular pressure, pupil examination, and anterior segment examination were normal in both eyes. A dilated examination of the right eye was unremarkable; however, a subfoveal pigmented scar with surrounding subretinal fluid (SRF) was seen in the left eye as well as a temporal parafoveal oval hypopigmented lesion and a larger hyperpigmented comet-shaped lesion in the temporal periphery (Figure 1A). Fluorescein angiography showed leakage from the subfoveal lesion, consistent with active CNV (Figure 1B). Optical coherence tomography (OCT) of the macula showed subfoveal hyperreflective material with adjacent SRF as well as ellipsoid zone attenuation in the area of the hypopigmented lesion (Figure 1, C and D). OCT of the temporal

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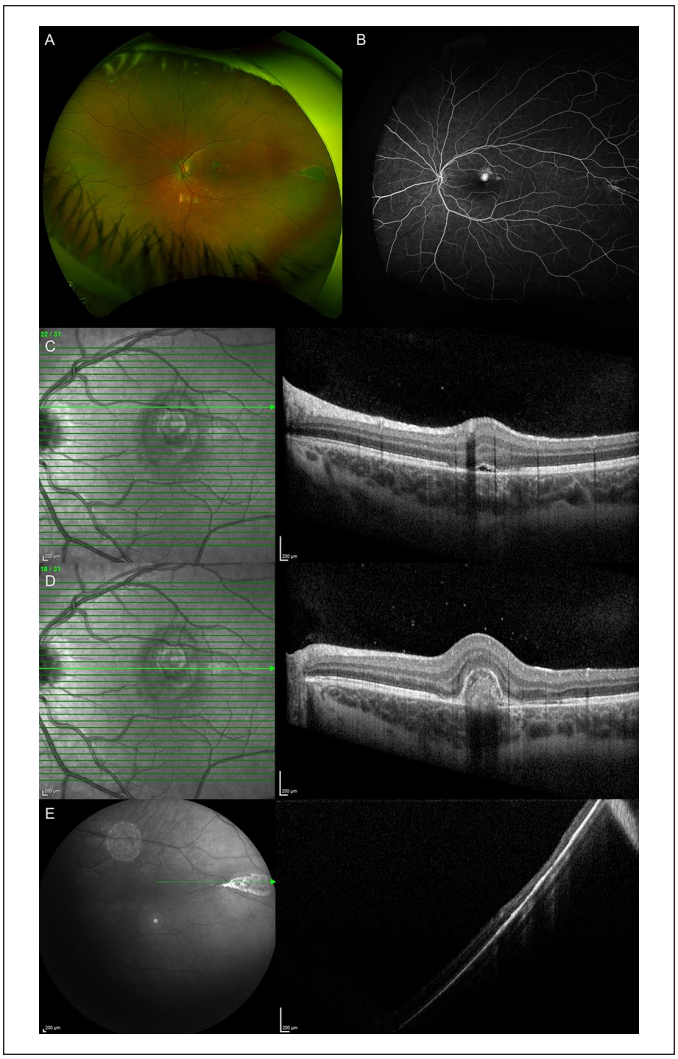

Figure 1. (A) Pseudocolor widefield photograph of the left eye at presentation shows a subfoveal pigmented scar with surrounding subretinal fluid (SRF), a temporal parafoveal oval hypopigmented lesion, and a larger hyperpigmented comet-shaped lesion in the temporal periphery. (B) Late-phase fluorescein angiography shows leakage from the subfoveal lesion, staining of the temporal parafoveal lesion, and blockage from the temporal peripheral lesion. (C and D) Optical coherence tomography (OCT) of the macula shows subfoveal hyperreflective material with adjacent SRF as well as ellipsoid zone attenuation in the area of the hypopigmented lesion. (E) OCT of the temporal peripheral lesion shows outer retinal loss and choroidal hypertransmission.

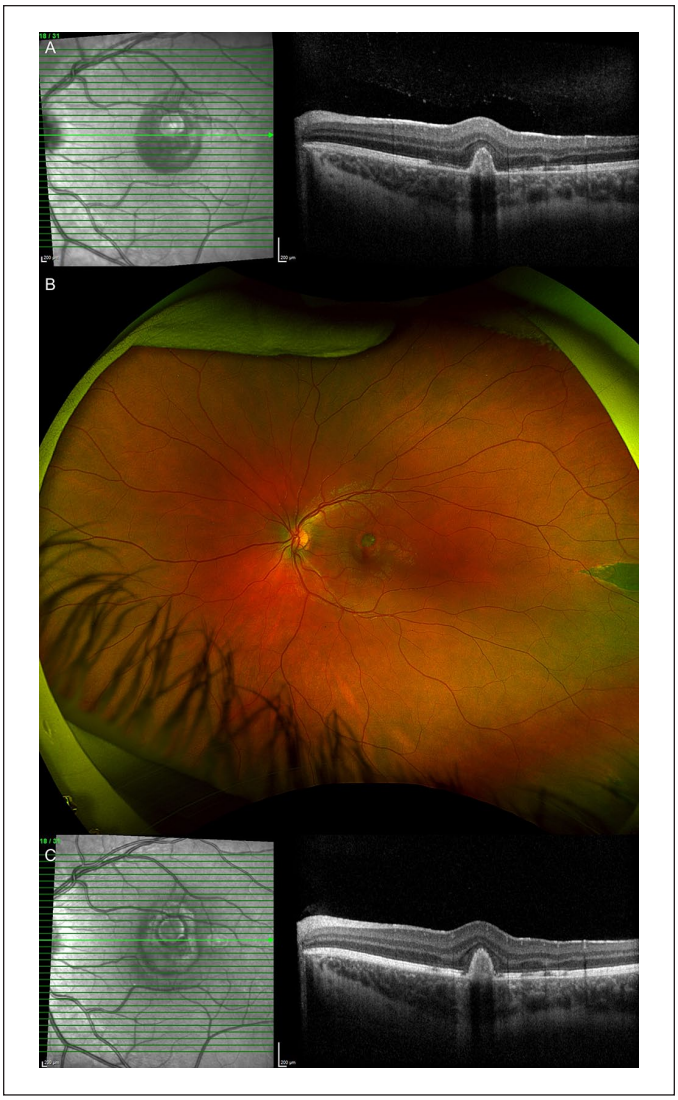

Figure 2. (A) Optical coherence tomography (OCT) of the left macula 1 month after presentation shows decreased subretinal hyperreflective material and subretinal fluid (SRF). (B) Pseudocolor widefield photograph and (C) OCT of the left eye 7 months after presentation shows regression of the choroidal neovascularization, resolution of the SRF, and a stable appearance of torpedo maculopathy lesions.

peripheral lesion showed outer retinal loss and choroidal hypertransmission (Figure 1E).

A systemic workup for toxoplasmosis, syphilis, tuberculosis, and sarcoidosis was unremarkable. Based on the appearance of the temporal lesions, a diagnosis of torpedo maculopathy with CNV was made. The patient had an examination under anesthesia (EUA), and an aqueous sample was sent for *Toxoplasma* polymerase chain reaction, which was negative. Intravitreal (IVT) ranibizumab 0.5 mg was administered to the left eye.

One month later, the patient's BCVA remained 20/20 OD and improved to 20/40 OS. An examination and OCT showed improvement in the CNV with decreased SRF (Figure 2A). Two months after the first treatment, the patient had an additional EUA with IVT ranibizumab 0.5 mg administered to the left eye. At last follow-up 7 months after presentation, the patient's BCVA improved to 20/20 OS with regression of the CNV, resolution of the SRF, and a stable appearance of torpedo maculopathy lesions (Figure 2, B and C).

Conclusions

Torpedo maculopathy was first described by Roseman and Gass[5] as a congenital flat, circumscribed, oval RPE lesion in the temporal macula. Although the underlying pathophysiology is unknown, Shields et al[3] postulated that the consistent temporal

Table 1. Published Cases of Torpedo Maculopathy Complicated by Choroidal Neovascularization.

Study[a]	Age (y)	Sex	Laterality	Presenting VA	Choroidal Excavation	Treatment	Final VA	Follow-up (mo)
Agarwal (2011)[12]	17	Male	OS	20/200	Unknown	Bevacizumab ×3	20/20	Unknown
Jurjevic (2017)[9]	34	Female	OD	20/400	Yes	Anti-VEGF ×4, antitoxoplasmic therapy, steroids	20/20	12
Parodi (2018)[13]	36	Female	OS	20/20	Yes	None	Unknown	Unknown
Shirley (2018)[14]	15	Female	OD	6/6	Yes	Ranibizumab ×1	6/5	2
Soman (2020)[15]	14	Male	OD	6/9	No	Ranibizumab ×2	Unknown	Unknown
Banaee (2020)[16]	19	Female	OS	20/30	Yes	Bevacizumab ×1	Unknown	Unknown
Byer (2021)[17]	63	Female	OD	20/25	Yes	Bevacizumab ×1	Unknown	0.5
Ghezzaz (2023)[18]	74	Female	OS	20/25	Yes	Aflibercept ×14	20/32	30
Current study	8	Male	OS	20/200	No	Ranibizumab ×2	20/20	7

Abbreviations: Anti-VEGF, antivascular endothelial growth factor; VA, visual acuity.
[a]First author.

location and size of torpedo maculopathy lesions represented a development defect of the RPE. Based on histopathologic studies, Streeten[6] observed a bulge in the temporal posterior pole with a high concentration of RPE cells at its apex at 4 months gestation that expanded to a staphylomatous bulge by 6 months and remained a slight residual depression 4 mm temporal to the optic disc at term.

Our case is unique from previously reported cases of torpedo maculopathy in a number of ways and may provide insight into the developmental pathophysiology of the condition. First, our patient presented with 2 distinct lesions located along the horizontal raphe. One was a smaller hypopigmented lesion temporal to the fovea, and the other was a larger hyperpigmented comet-shaped lesion in the temporal periphery. Smaller satellite lesions temporal to torpedo maculopathy lesions have been previously described, and there was a report in the literature of a "double torpedo" configuration of 2 torpedo maculopathy lesions arranged in parallel. However, to our knowledge, our report is the first to describe a large comet-shaped peripheral lesion exhibiting symmetry across the horizontal raphe.[7–10] The horizontal raphe is a demarcation line extending from the macula to the temporal ora serrata, dividing the retina and choroid into separate vascular hemispheres. Its development is thought to be a carefully choreographed process induced in part by the growth of the long posterior ciliary arteries.[11] In our case, the symmetry of both lesions across the horizontal raphe lends support to the hypothesis that an alteration at a specific timepoint of retinal and RPE development results in torpedo maculopathy.

Second, our patient had hypopigmentation of the central lesion and near-uniform hyperpigmentation of the temporal lesion. Some previous torpedo maculopathy lesions have shown hyperpigmentation of their temporal aspect; however, none have shown hyperpigmentation of the entire lesion. Our patient's temporal lesion was also located farther temporally based on a topographic distribution map of previously reported cases.[1] Streeten[6] observed a small dark spot at the apex of the temporal bulge with the highest concentration of RPE cells, a location that was nearer the temporal edge of the bulge, and subsequently expanded to fill the posterior pole. A pause in this process could be consistent with a hyperpigmented temporal lesion and a hypopigmented central lesion, as seen in our case.

Our patient also had macular CNV that was responsive to antivascular endothelial growth factor (anti-VEGF) therapy. To our knowledge, there are 8 previously published cases of torpedo maculopathy complicated by CNV; ours is the youngest patient to date (Table 1).[9,12–18] It has been thought that CNV can develop in torpedo maculopathy as a result of focal alterations of the outer retina, Bruch membrane, and RPE or choroidal ischemia, especially in the setting of focal choroidal excavation.[9,17–19] Unlike most previous cases of torpedo maculopathy with CNV, choroidal excavation was not seen in our patient, highlighting that choroidal excavation is not a prerequisite for the development of CNV in torpedo maculopathy. Similar to some but not all previously reported cases, our patient had an excellent response to relatively few anti-VEGF treatments. In the absence of an established standard of care for CNV in the pediatric population, we chose to treat with ranibizumab, which has been used for CNV in pediatric patients. The MINERVA study found that ranibizumab was safe and efficacious for adult patients and adolescent patients with CNV secondary to uncommon causes, and ranibizumab led to good outcomes in previous cases of torpedo maculopathy with CNV.[14,20–22]

In summary, we present a pediatric patient with a unique configuration of torpedo maculopathy complicated by macular CNV. The incidence of and risk factors for CNV in torpedo maculopathy are unknown, with limited data on its treatment and prognosis. Future studies that highlight this rare complication of an uncommon entity are needed.

Ethical Approval

This case report was conducted in accordance with the Declaration of Helsinki. The collection and evaluation of all protected patient health information were performed in a US Health Insurance Portability and Accountability Act–compliant manner.

Statement of Informed Consent

Written informed consent for publication of patient information and images was not required because no personal identifiable information is included in this report.

Declaration of Conflicting Interests

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