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Review Article

Distribution patterns of torpedo maculopathy: Further evidence of a congenital retinal nerve fiber layer-driven etiology

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Abstract

With fewer than 100 peer-reviewed cases reported in the world to date, the underlying etiology of torpedo maculopathy has remained elusive. In this literature review, we provide new evidence to better support, reject and unify claims regarding cause, diagnosis, and proper clinical management of this disease. We reviewed 44 case reports and case series, which included 77 patients (after exclusions). We additionally introduced 3 new cases from our clinical practice for a total of 80 cases. Ages at presentation ranged from 6 months old to 73 years old (mean: 24.2 years old). The nasal aspects of torpedo maculopathy lesions pointed toward the optic disc and localized to a kite-shaped region of the temporal macula, correlating with the anatomic junction of the superior arcuate, inferior arcuate, and papillomacular bundles of retinal nerve fiber layer distribution. No patterns were observed among the temporal aspects of the lesions. These findings support a congenital etiology of torpedo maculopathy and a possible influence of the retinal nerve fiber layer in the development of mature retinal pigment epithelium.

Keywords: Congenital, Distribution, Embryology, Hypopigmentation, Nerve fiber layer, Torpedo

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Introduction

In 1993, the term torpedo maculopathy was introduced by Mark J Daily to describe a characteristic torpedo-shaped, presumably congenital lesion of the retinal pigment epithelium (RPE) that generally localized in the temporal macula.[1] A similar lesion was described by Roseman and Gass as early as 1992.[2] Since then, numerous reports have been added to the literature, but sample sizes have been relatively small and disjointed, warranting a more comprehensive review.

Herein we present 3 new cases of torpedo maculopathy in aggregate with 77 previously reported cases to better elucidate etiological cause, diagnostic characteristics, and clinical management recommendations.

Methods

All known peer-reviewed case reports of clinical torpedo maculopathy from October 1992 to October 2017 were obtained from PubMed, Google Scholar, and previously

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Abbreviations: BCVA, best corrected visual acuity, CHRPE, congenital hypertrophy of the retinal pigment epithelium, IOP, intraocular pressure, OCT, optical coherence tomography, RPE, retinal pigment epithelium

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Table 1. Quantitative and qualitative data from all patients reviewed.

	# of cases	Age	Gender	Eye	Nasal aspect	Pigment Present?	Temporal aspect	Pigment Present?	Direction	OCT Cavitation	OCT Excavation
Rigotti et al. [17]	3	Child	?	OS	Temporal parafovea		Temporal macula	Y	Horizontal		
		?	?	?	Temporal to fovea		Temporal macula		Horizontal		
		?	?	?	Temporal to fovea		Temporal macula		Horizontal		
Barbazetto et al. [18]	1	10	F	OD	Temporal perifovea		Temporal periphery		Angled superiorly	Y	Y
Kara et al. [19]	1	0.5	M	OD	Temporal parafovea		Temporal macula	Y	Horizontal		
Shields et al. [20]	2	3	F	OD	Inferotemporal parafovea		Temporal macula	Y	Horizontal		
		11	F	OS	Temporal parafovea		Temporal macula	Y	Angled superiorly		
Golchet et al. [1]	13	33	F	OS	Foveola		Temporal macula	Y	Angled superiorly	Y	Y
		18	M	OS	Foveola		Temporal macula	Y	Angled superiorly	Y	
		1	M	OS	?		?		?		
		7	M	OS	Temporal parafovea		Temporal macula	Y	Horizontal		
		8	F	OD	?		?		?		
		9	M	OD	Temporal macula		Temporal macula		Horizontal		
		13	M	OD	Temporal fovea		Temporal macula	Y	Horizontal		
		14	F	OS	?		?		?		
		20	F	OD	Inferotemporal perifovea		Inferotemporal macula	Y	Angled superiorly		
		24	F	OD	?		?		?		
		25	M	OD	?		?		?		
		33	F	OS	?		?		?		
		68	F	OD	?		?		?		
Roseman and Gass. [2]	1	12	M	OS	Superotemporal parafovea		Temporal macula		Horizontal		
Pian et al. [52]	3	59	M	OS	Inferior parafovea		Temporal macula	Y	Horizontal		
		63	M	OD	Temporal parafovea		Temporal macula	Y	Angled superiorly		
		63	M	OS	Temporal parafovea		Temporal macula	Y	Horizontal		
Trevino et al. [8]	1	25	F	OD	Temporal perifovea		Temporal macula	Y	Horizontal		Y
	0	**22**	**F**	**OD**	**Foveola**	**Y**	**Temporal macula**	**Y**	**Horizontal**	**n/a**	**n/a**
Dolz-Marco et al. [22]	1	7	M	OS	Far temporal macula		Temporal periphery		Horizontal	Y	
Nicolau et al. [23]	1	30	F	OD	Temporal parafovea		Temporal macula	Y	Angled superiorly		
Cullen et al. [24]	1	10	M	OD	Foveola		Temporal macula	Y	Horizontal		
Richez et al. [11]	**0**	**1.5**	**?**	**OU**	**Temporal maculae**	**Y**	**Temporal maculae**	**Y**	**Horizontal**	**n/a**	**n/a**
Suárez et al. [25]	1	4	F	OD	Inferior macula		Inferotemporal macula		Angled superiorly	Y	
Villegas et al. [15]	1	?	?	OD	Temporal perifovea		Temporal macula	Y	Horizontal		
Wong et al. [26]	5	14	F	OD	Temporal fovea		Temporal macula		Angled superiorly	Y	
		15	M	OS	Temporal perifovea		Temporal macula	Y	Angled superiorly		
		37	F	OS	Temporal parafovea		Temporal macula	Y	Horizontal		
		59	M	OD	Temporal macula		Temporal macula	Y	Horizontal	Y	Y
		73	F	OD	Temporal parafovea		Temporal macula	Y	Horizontal	Y	
Giannakaki-Zimmerman et al. [27]	4	8	F	OD	Superotemporal parafovea		Superotemporal macula	Y	Angled inferiorly	Y	
		13	F	OD	Foveola		Temporal macula	Y	Horizontal	Y	
		37	F	OD	Temporal parafovea		Temporal macula	Y	Horizontal	Y	
		32	F	OD	?		?		?		
Rohl et al. [12]	**0**	**10**	**M**	**OS**	**Temporal perifovea**	**Y**	**Temporal macula**	**Y**	**Horizontal**	**n/a**	**n/a**
Makino et al. [13]	**0**	**8**	**M**	**OD**	**Temporal and Inferotemporal macula**	**Y**	**Temporal periphery**	**Y**	**Horizontal**	**n/a**	**n/a**
Güven et al. [28]	1	10	M	OD	Inferotemporal parafovea		Inferotemporal macula		Angled superiorly		
Merle et al. [29]	1	70	?	OS	Temporal parafovea		Temporal macula	Y	Horizontal		
Angioï-Duprez et al. [30]	1	18	F	OS	Foveola		Temporal macula	Y	Horizontal		
Bedar et al. [31]	1	7	M	OD	Temporal perifovea		Temporal macula	Y	Horizontal	Y	Y
Hansen et al. [32]	1	12	M	OS	Temporal perifovea		Temporal macula		Angled superiorly	Y	Y
Su et al. [33]	1	38	F	OD	Temporal perifovea		Temporal macula	Y	Horizontal		

(continued on next page)

Table 1 (*continued*)

	# of cases	Age	Gender	Eye	Nasal aspect	Pigment Present?	Temporal aspect	Pigment Present?	Direction	OCT Cavitation	OCT Excavation
Sanabria et al. [21]	2	12	F	OD	Temporal Parafovea		Temporal macula		Horizontal		
		13	F	OS	Inferotemporal Parafovea		Temporal macula	Y	Angled superiorly	Y	Y
Papastefanou et al. [34]	2	24	M	OS	Temporal parafovea		Temporal macula	Y	Horizontal	Y	Y
		57	F	OS	Temporal parafovea		Temporal macula	Y	Horizontal	Y	Y
Jurjevic et al. [10]	3	20	F	OS	Inferotemporal perifovea		Inferotemporal macula	Y	Angled superiorly	Y	Y
		49	F	OD	Superotemporal parafovea		Superotemporal macula		Horizontal	Y	Y
		34	F	OD	Temporal fovea		Temporal macula	Y	Horizontal		Y
Mercan et al. [35]	1	61	M	OD	Foveola		Temporal macula	Y	Angled superiorly	Y	Y
Schuerch et al. [36]	2	3	F	OD	Superotemporal parafovea		Superotemporal macula	Y	Angled inferiorly		
		36	F	OD	Temporal perifovea		Temporal macula	Y	Angled superiorly		
Pilotto et al. [37]	1	4	F	OD	Temporal fovea		Temporal macula	Y	Horizontal		
Dutra-Medeiros et al. [38]	1	5	F	OD	Temporal fovea		Temporal macula	Y	Horizontal		
Buzzonetti et al. [39]	1	6	M	OS	Nasal fovea		Temporal macula		Horizontal	Y	Y
Tsang et al. [40]	3	18	F	OS	Superior fovea		Temporal macula	Y	Horizontal		
		5	F	OD	Temporal perifovea		Temporal macula	Y	Angled superiorly		
		25	F	OS	Temporal parafovea		Temporal macula		Horizontal		
Shields, Shields. [41]	**0**	**n/a**	**n/a**	**n/a**	**REPEAT CASE**	**n/a**	**n/a**	**n/a**	**n/a**	**n/a**	**n/a**
Bhatt et al. [42]	1	21	M	OS	Foveola		Temporal macula		Horizontal		
Thomas et al. [43]	1	8	F	OS	Foveola		Temporal macula	Y	Horizontal	Y	Y
De Manuel-Triantafilo et al. [44]	2	4	M	OD	Temporal fovea		Temporal macula		Horizontal		
		25	F	OS	Superotemporal perifovea		Superotemporal macula	Y	Angled superiorly	Y	
Teitelbaum et al. [45]	2	?	?	OD	Temporal macula		Temporal macula	?	Horizontal		
		?	?	OS	Temporal perifovea		Temporal macula	Y	Horizontal		
Iaboni et al. [46]	1	21	F	OS	Inferotemporal perifovea		Inferotemporal macula		Angled superiorly	Y	
Hamm et al. [47]	1	60	F	OD	Temporal parafovea		Temporal macula	Y	Angled superiorly	Y	Y
Ali et al. [48]	2	19	F	OD	Superotemporal parafovea		Temporal macula	Y	Angled superiorly	Y	Y
		46	F	OD	Temporal parafovea		Temporal macula	Y	Angled superiorly		
Cherney. [49]	4	?	?	OD	Temporal macula		Temporal macula	Y	Horizontal		
		7	M	OS	Temporal macula		Temporal macula	Y	Angled superiorly		
		22	F	OS	Subfoveal		Temporal parafovea	Y	Horizontal		
		?	?	OD	Temporal parafovea		Temporal macula		Horizontal		
Stoyukhina et al. [50]	1	12	?	OD	Superior parafovea		Superotemporal macula	Y	Horizontal	Y	
de Bats et al. [51]	1	35	M	OD	Superotemporal parafovea		Temporal macula	Y	Horizontal	Y	Y
Patient 1 (This paper)	1	5	F	OS	Superior parafovea		Superotemporal macula	Y	Horizontal	Y	Y
Patient 2 (This paper)	1	54	M	OS	Temporal parafovea		Temporal macula		Angled superiorly		
Patient 3 (This paper)	1	16	F	OD	Inferotemporal parafovea		Inferotemporal macula		Horizontal		

Bold text indicates cases that were excluded.

published reference lists. Search queries included: ''torpedo'', ''torpedo maculopathy'', ''paramacular coloboma'', ''hypopigmented nevus'', and ''albinotic spot''. Macular colobomas and lesions associated with known global syndromes were not included.

These reports were reviewed for age, race, eye affected, shape, location, pigmentation, imaging performed, and exam notes. Four reports were suspected to be misidentified and one report repeated a previously described case. All four cases are included in Table 1 but excluded from the analysis.

Fundus imaging was utilized to gather qualitative information. Preference was given to color fundus photography. After data collection and analysis were complete, the nasal-most points of the lesion origins were plotted on individual reference fundus photographs of a normal left and right eye. This was done using relative measurements between the temporal optic disc margin, the central fovea, and the superior and inferior arcades. Finally, the individual images were aggregated into a single distribution map image of the left and right eyes. This aggregation method helped reduce the bias associated with the manual transfer of this data.

Results

We reviewed 44 previous reports that included 77 distinct cases of torpedo maculopathy after exclusions (4). We additionally introduced 3 new cases for a total of 80 unique cases of torpedo maculopathy (Fig. 1). Age at presentation ranged from 6 months old to 73 years old (mean 24.2 years). Out of 78 cases reporting eye affected, 45 were right eyes and 33 were left eyes. The only bilateral case was excluded due to diagnostic inconstancy with the given diagnosis based on the clinical imaging provided and a failure to rule out salient inflammatory etiologies. No correlations with race were apparent as patients were reported in a wide variety of countries throughout Europe, North America, South America, Africa, and Asia.

There were 67 cases deemed to have sufficient photographic evidence for qualitative review. We generated a right eye and a left eye distribution pattern of these nasal aspects of torpedo maculopathy which revealed a horizontal kite-shaped pattern in both distribution maps (Fig. 2). The temporal aspect distributions did not produce any remarkable pattern. Analysis of the prior case reports revealed that torpedo maculopathy does not always point toward the fovea. However, in all 67 cases, the nasal aspect of the torpedo lesions pointed toward the disc. Relative hyperpigmentation of the temporal lesion aspect was seen in 51 cases, but relative hyperpigmentation of the nasal aspect was not seen.

Optical coherence tomography (OCT) was reported for 51 patients. Of these, 28 cases revealed a cavitation resembling subretinal fluid, and 19 cases reported retinochoroidal excavation. There were 14 cases that reported visual field defects on ancillary testing. Only 1 case reported associated choroidal neovascular membrane requiring treatment with a single series of injections. No clinically significant changes were observed in cases reporting long-term follow-up.

Discussion

Torpedo maculopathy is a rare diagnosis that is ophthalmoscopically characterized as a horizontal, oval-shaped area of hypopigmentation localized to the temporal macula. The etiology of this condition is not well understood as of yet. However, we hope that our findings, in combination with this literature review, may help unify the known clinical features of this rare diagnosis and aid in clinical management.

Our first case was a 5-year-old African American girl who was referred to our center for incidental retinal findings on a routine exam. Best-corrected visual acuity (BCVA) measured 20/50 in the right eye and 20/25 in the left eye. The right eye was undergoing regular patching for refractive amblyopia at the time of the visit. Intraocular pressure (IOP) was normal. A review of the patient's medical history revealed a full-term birth complicated by maternal type 1 diabetes. The patient was evaluated by genetics, who diagnosed diabetic embryopathy in association with patent ductus arteriosus, a ventricular septal defect, and an imperforate anus.

Slit-lamp exam was unremarkable except for ptosis of both upper lids. Fundus exam revealed a unilateral ovoid-shaped area of hypopigmentation localized in the superior and superotemporal macula of the left eye. The lesion originated nasal to the fovea and extended laterally into the superotemporal macula (Fig. 1). An OCT was obtained which revealed underlying cavitation and excavation at the level of the ellipsoid zone, RPE, and choroid, as well as mild intraretinal schisis and a retinal nerve fiber layer defect above the nasal aspect of the lesion (Fig. 1).

Our second case was a 54-year-old Caucasian male who was referred for a concerning lesion in the right eye. BCVA measured 20/40 in the right eye and 20/20 in the left eye. The patient reported a history of pigmentary glaucoma that was treated with peripheral iridotomy in both eyes. IOP was normal. Past medical history was only remarkable for hypertension. Slit-lamp exam was unremarkable. Fundus exam revealed a pigmented mass with orange pigment along the inferotemporal arcade of the right eye and an incidental torpedo-shaped hypopigmented lesion located in the temporal macula of the left eye. The patient was diagnosed with malignant melanoma and treated with brachytherapy of the right eye. The patient had multiple unremarkable Humphrey visual fields of the left eye.

Our third case was a 16-year-old Caucasian girl who presented for an annual eye exam to get new glasses. BCVA measured 20/20 in both eyes. IOP was normal. Past medical history was unremarkable. Her eye exam was normal except for a torpedo-shaped lesion and an additional trailing satellite lesion in the inferotemporal macula of the right eye (Fig. 1). Fundus autofluorescence revealed hypoautofluorescence corresponding to the lesion. An OCT was performed which revealed irregularity at the level of the ellipsoid zone and the RPE (Fig. 1). Confrontation visual fields were unremarkable.

In all three cases, the torpedo lesions were asymptomatic and incidentally discovered on exam. All three cases remained stable at 3-month follow-up. The lesion shape and relative location were variable in all three cases. Patient 2 had numerous normal Humphrey visual field 24-2 tests. The other two patients had no formal visual field testing. Autofluorescence imaging on Patient 3 revealed hypoautofluorescence corresponding to the shape of the lesion and its associated satellite lesion. OCT on Patient 1, revealed a cavitary excavation of the outer retina, RPE, and choroid, which

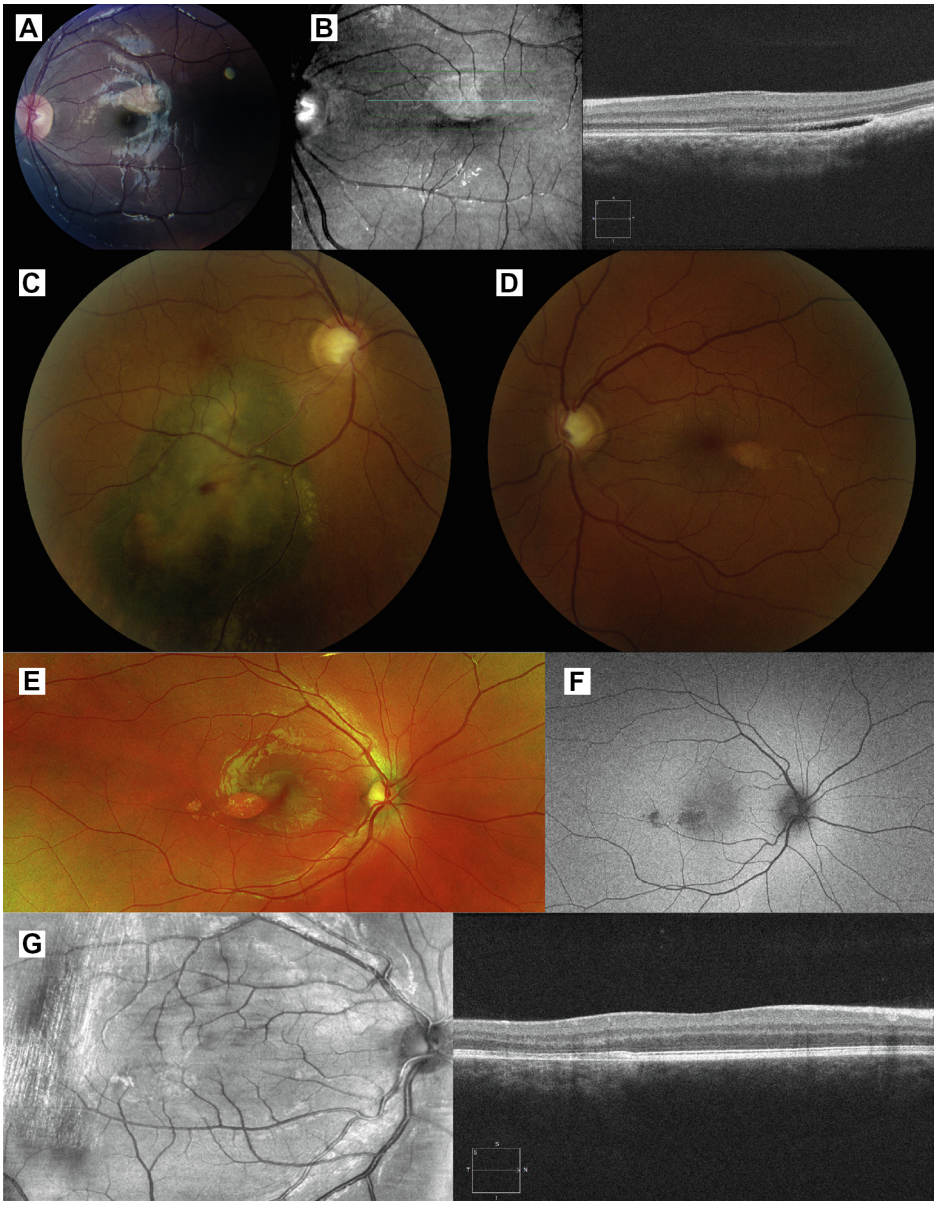

Fig. 1. A. Patient 1 color fundus photo of torpedo maculopathy originating in the superonasal macula of the left eye. B. Patient 1 OCT revealing retinal nerve fiber layer irregularity, mild choroidal excavation, and cavitation resembling subretinal fluid at the level of the RPE and outer retina. C. Patient 2 color fundus photo of choroidal melanoma of the right eye. D. Patient 2 color fundus photo of a small temporal torpedo lesion in the left eye. E. Patient 3 laser scanning pseudo-color image of torpedo maculopathy with a satellite lesion in the right eye. F. Patient 3 autofluorescence image showing hypoautofluorescence corresponding to the torpedo lesion. G. Patient 3 OCT revealing mild disorganization at the level of the ellipsoid zone and RPE.

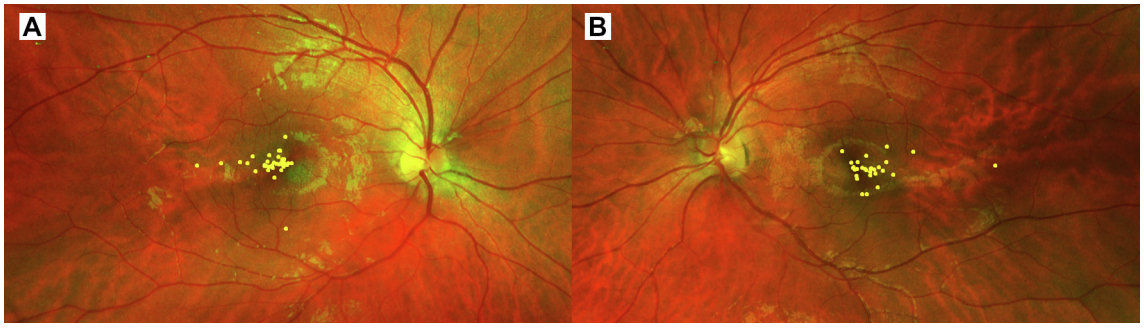

Fig. 2. A. Right eye distribution map of 38 nasal torpedo maculopathy origins transposed on a normal reference eye. B. Left eye distribution map of 29 nasal torpedo maculopathy origins transposed on a normal reference left eye.

resembled subretinal fluid. The lesion observed in Patient 1 also atypically extended into the superonasal macula which allowed for better visualization of a disruption of unknown significance in the overlying retinal nerve fiber layer. This nasal macular location has not been reported previously to our knowledge.

Great advancements in our understanding of ocular embryology have been made with advanced imaging modalities, animal models, and stem cells; however, the specifics of development and differentiation of the RPE and neural retina in human models are not fully understood and are constantly evolving.[3–6] It has long been accepted that the presence of pigmentation in developing RPE indicates differentiation of the RPE cell type. However, recent evidence of continuing polarity interactions between RPE and neural retinal cells during early differentiation suggests that the presence of pigmentation may not signify fully mature RPE.[7] Relatedly, we believe that the distribution patterns of torpedo maculopathy suggest a correlation between the development of retinal nerve fiber layer and the development of mature RPE.

Torpedo maculopathy is presumed to be a congenital lesion of the RPE, which is supported by numerous clinical observations, including presentation in infancy and lesion stability of over long-term follow-up. Torpedo maculopathy lesions characteristically present with decreased or absent autofluorescence which may support not only loss of RPE pigmentation, but also an overall loss of RPE functionality.[8] Additionally, torpedo maculopathy consistently localizes to the temporal macula which may support a congenital etiology with a conserved mechanism.

Review of the literature revealed multiple correlations that further support a congenital etiology. First, every lesion reviewed pointed toward the optic disc. Second, lesion angularity relative to the horizon ranged from perfectly horizontal to superiorly tilted toward the superior aspect of the disc, which is consistent with the slight downwardly-sloped distribution of the horizontal raphe of the retinal nerve fiber layer (Fig. 3).[9] Third, although the temporal aspects of torpedo maculopathy did not produce any recognizable distribution pattern, the nasal aspects localized to a single well-defined kite-shaped region at the junction of the superior arcuate, inferior arcuate and papillomacular bundles of the retinal nerve fiber layer (Fig. 2). According to retinal laser ellipsometry, this kite-shaped region is also associated with relatively low retinal nerve fiber layer density, which may explain the consistent localization of torpedo maculopathy.[9] Based on this data, we suspect a developmental association between the retinal nerve fiber layer and developing RPE.

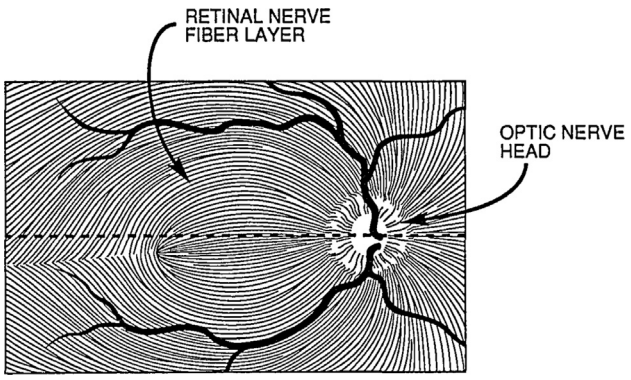

Fig. 3. Schematic of the retinal nerve fiber layer. Reprinted with permission of Robert N. Weinreb, MD.

Fundus autofluorescence and fluorescein angiography maintained a characteristic appearance in all reported cases of torpedo maculopathy. Autofluorescence imaging showed hypoautofluorescence corresponding to the hypopigmented lesion area, and fluorescein angiography revealed a corresponding hyperfluorescent window defect without leakage. These findings were consistent across all lesions reviewed and may indicate a loss of functional RPE.[8]

OCT scans of the torpedo lesions were variable. The outer retina, RPE and choroid were all affected, but to differing degrees along a spectrum. The inner retina was found to be largely preserved with the exception of the retinal nerve fiber layer, which appeared relatively thin directly above the nasal aspect of the lesion in some cases. With that being said, we were unable to measure the thickness of the retinal nerve fiber layer for confirmation. Further studies to better assess the status of the overlying retinal nerve fiber layer may be warranted in the future as technology improves. Some patients may present with mild outer retinal discontinuity of the temporal macula and others may present with outer retinal loss, subretinal cavitation resembling subretinal fluid, and significant choroidal excavation. Overall, only one case report described choroidal neovascularization in association with torpedo maculopathy in our review, and that case was stabilized with a series of three intravitreal bevacizumab injections.[10]

On color imaging, torpedo lesions classically presented in the temporal macula, just temporal to the fovea. Torpedo maculopathy generally spared the foveola, although subfoveal cases were reported. The nasal aspects of the torpedo lesions were generally pointed or rounded and were usually sharply demarcated without hyperpigmentation, possibly indicating a point of origin. The temporal aspects, on the other hand, were non-binary and frequently presented with irregular pigmentation and/or hyperpigmentation. Temporal aspects were rounded, irregular, frayed or non-solitary. In general, the irregular pigmentation and relative hyperpigmentation associated with the temporal aspects were found to be variable.

Four reported cases were found to exhibit nasal hyperpigmentation and irregular borders and all four failed to rule out other prominent differential diagnoses and were therefore excluded from analysis.[8,11–13] Although bilateral cases of torpedo maculopathy are presumed to be possible, the only bilateral case reported in the literature, that was not part of a global syndrome, was one of the excluded reports.[11] This bilateral case presented with decreased visual acuity attributed to the lesions, clear visibility of the choroidal vasculature, circumferential hyperpigmentation and circumferential border irregularity, which is not consistent with torpedo maculopathy. The second case excluded presented as a completely hyperpigmented lesion of the RPE in the macula with circumferential border irregularity and a progressive pseudo-lacuna, which is not consistent with torpedo maculopathy.[12] The third case excluded presented with decreased visual acuity attributed to the lesion, temporal macular dragging, disorganization of the inner retina, circumferential border irregularity, and irregular hyperpigmentation, which is not consistent with torpedo maculopathy.[13] The fourth case excluded presented with decreased visual acuity attributed

to the lesion, clear visibility of the choroidal vasculature, circumferential hyperpigmentation and circumferential border irregularity, which is not consistent with torpedo maculopathy.[8]

Common differential diagnoses to consider when confronted with a macular lesion include toxoplasmosis (or chorioretinal scar from another unknown etiology), toxocariasis, macular coloboma, congenital hypertrophy of the retinal pigment epithelium (CHRPE) and combined hamartoma of the retina and RPE. The major differentiating factors between these lesions and torpedo maculopathy were the presence of 360-degree hyperpigmentation with irregular borders, clear visualization of the choroidal vasculature, reduction of visual acuity attributed to a lesion, macular dragging, and central macular location. The finding of 360-degree hyperpigmentation with irregular borders is most commonly seen in inflammatory conditions with chorioretinal scarring. Clear visualization of the choroid is typically seen in conditions such as chorioretinal scarring and macular colobomas. Reduction in visual acuity attributed to a lesion is commonly seen in conditions that specifically affect the fovea, including chorioretinal scarring, macular coloboma, or macular combined hamartoma of the retina and RPE. Macular dragging has not been associated with torpedo maculopathy, but it can be seen in conditions such as toxocariasis and combined hamartoma of the retina and RPE. Other syndromes associated with torpedo-like lesions include Turcot syndrome,[14] Gardner syndrome,[15] and congenital Zika virus infection.[16]

Conclusion

Torpedo maculopathy is presumed to be a congenital lesion of the RPE that can affect the outer retina and choroid. Distribution mapping analysis of torpedo lesions supports a congenital etiology and suggests a role of the retinal nerve fiber layer in the proper development of mature RPE. Torpedo lesions are generally oval-shaped and sharply demarcated with a longer horizontal length compared to the vertical. They orient toward the optic disc along the horizontal raphe and localize to a kite-shaped region of the temporal macula, correlating with the anatomic junction of the superior arcuate, inferior arcuate, and papillomacular bundles. Torpedo maculopathy is often hypopigmented and sharply demarcated nasally and characteristically variable with pigmentary changes temporally. Most lesions are asymptomatic and rarely involve the foveal center. Patients almost always maintain excellent visual acuity, but visual field defects on formal testing are possible. When presented with a questionable oval-shaped lesion, it is important to consider other differential diagnoses including toxoplasmosis (or chorioretinal scar from another unknown etiology), toxocariasis, macular coloboma, combined hamartoma of the retina and RPE, CHRPE, Turcot syndrome, Gardner syndrome, and Zika virus infection.

Conflict of interest

The authors report no conflict of interest.

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The authors report no relevant financial disclosures or proprietary interests. This report has not been presented previously. Informed consent was obtained for each patient presented. Written permission was obtained from the corresponding author via email to reprint Fig. 3.

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