• Letter to the Editor •

# Case report and literature review of torpedo maculopathy in four preterm infants

*Bo Chen, Hong Yang*

Department of Ophthalmology, Tongji Hospital, Tongji Medical College, Huazhong University of Science and Technology, Wuhan 430030, Hubei Province, China

**Correspondence to:** Hong Yang. Department of Ophthalmology, Tongji Hospital, Tongji Medical College, Huazhong University of Science and Technology, Wuhan 430030, Hubei Province, China. dr_yangh@aliyun.com

Received: 2025-04-02 Accepted: 2025-07-16

**DOI:10.18240/ijo.2026.02.25**

**Citation:** Chen B, Yang H. Case report and literature review of torpedo maculopathy in four preterm infants. *Int J Ophthalmol* 2026;19(2):414-416

**Dear Editor,**

Torpedo maculopathy (TM), first described by Roseman and Gass in 1992[1], is a rare congenital unilateral retinal pigment epithelium (RPE) abnormality. The term "torpedo maculopathy" was coined by Daily[2] in 1993. TM typically spares the foveal center, is asymptomatic, and is often detected incidentally during routine ophthalmic examinations. Through literature search, we did not identify racial or regional differences in TM. It predominantly affects children, with an estimated prevalence of 2 per 100 000 in individuals under 16 ages[3]. While previous reports have focused on pediatric and adult populations, this study presents four cases of TM in preterm infants.

**Case Presentation** This study was conducted in accordance with the principles of the Declaration of Helsinki. Written informed consent was obtained from the patient's parent. Four preterm infants (2 males, 2 females) diagnosed with TM between 2020 and 2024 at the Department of Ophthalmology, Tongji Hospital, Wuhan, China, were included. All lesions involved the right eye. Gestational ages ranged from 30wk+6 to 36wk+5, birth weights from 1.75 kg to 2.83 kg, and postmenstrual ages (PMA) at examination from 41wk to 60wk+5 (Table 1).

All four cases showed no history of retinopathy of prematurity (ROP) or other retinal disorders. Fundus examinations which conducted by RetCam revealed well-defined, oval-shaped hypopigmented lesions temporal to the macular fovea, measuring 0.5-2 disc diameters (PD) horizontally and 0.3-1 PD vertically. These lesions exhibited a characteristic "torpedo-shaped" configuration: the head was oriented along the horizontal raphe toward the fovea, while the tail extended outward as a wedge-shaped lesion with normal overlying retinal vasculature (Figure 1). Satellite lesions adjacent to the primary lesion were observed in Case 1 (Figure 1A).

Due to the young age of the preterm infants at the time of initial examination, no additional ophthalmic evaluations were performed. In Case 4, a follow-up examination four years later revealed a visual acuity of 0.6 in both eyes, with intraocular pressure (IOP) measuring 18 mm Hg right eye (OD) and 17 mm Hg left eye (OS). Anterior segment examination showed no abnormalities. Fundus photographs of the right eye demonstrated the lesion area has slightly increased compared to the findings four years prior (Figure 2A, 2B). Optical coherence tomography (OCT) of the right eye revealed disruption of the outer retinal inner segment/outer segment (IS/OS) layer without associated outer retinal edema or subretinal fluid, along with atrophy of the choriocapillaris layer (Figure 2C, 2D). According to the OCT-based classification of TM, Case 4 was categorized as TM Type I.

**DISCUSSION**

Since its first description in 1992[1], TM has only been reported in small case series. It is currently considered a rare congenital lesion, typically unilateral, characterized by a torpedo-shaped area of temporal macular hypopigmentation with the apex directed toward the fovea and a tail exhibiting varying degrees of pigment deposition[4]. The pathogenesis of TM remains unclear, with three predominant hypotheses in the literature: 1) a developmental defect of the nerve fiber layer along the horizontal raphe[5]; 2) abnormal choroidal or ciliary body vascular development leading to localized, non-progressive RPE dysfunction[6]; 3) persistent RPE developmental defects secondary to fetal temporal bulge formation[4].

Recent studies have detailed the multimodal imaging features of TM. TM is often incidentally detected during routine examinations. Fundus color photography typically reveals a spindle-shaped hypopigmented or hyperpigmented retinal lesion temporal to the fovea[7], consistent with the findings in

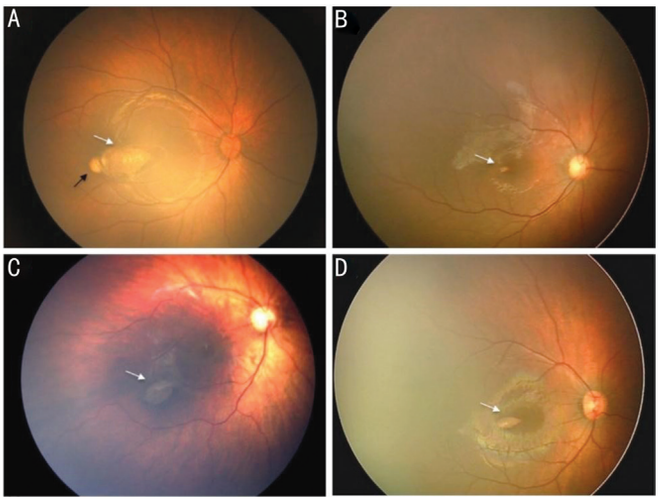


**Figure 1 Wide-field fundus images captured by RetCam of Cases 1-4** White arrows indicate the oval-shaped hypopigmented lesions; Black arrow in A indicates the satellite lesion (Case 1).

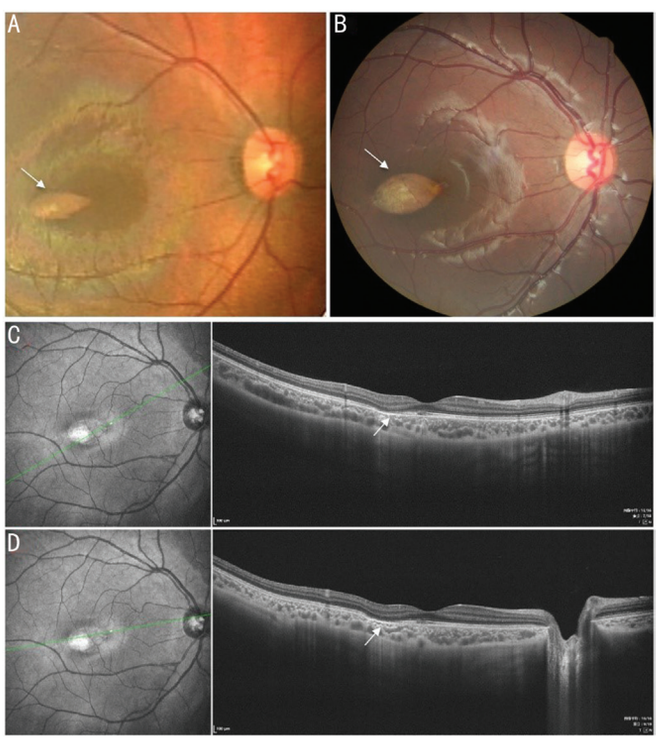


**Figure 2 Fundus and OCT images of Case 4 at different ages** A: Fundus image of Case 4 at age of two months old, white arrow indicates the oval-shaped hypopigmented lesion; B: Follow-up fundus image of Case 4 after four years, white arrow indicates the oval-shaped hypopigmented lesion; C-D: White arrows in optical coherence tomography indicate outer retinal and choriocapillaris atrophy.

the four preterm infants reported in this study. Infrared imaging clearly delineates the lesion as a horizontal oval, aligning with color fundus observations[8]. Autofluorescence typically shows hypoautofluorescence (varying with RPE atrophy severity) with hyperautofluorescent borders (attributed to lipofuscin accumulation due to RPE metabolic dysfunction)[9-10]. Based on OCT features, TM has been classified into four subtypes:

**Table 1 Clinical characteristics of the four cases**

| Case | Sex | Gestational age (wk) | Birth weight (kg) | PMA at exam (wk) |
|---|---|---|---|---|
| 1 | Male | 36+5 | 2.83 | 50+3 |
| 2 | Female | 34+4 | 2.05 | 60+5 |
| 3 | Male | 30+6 | 2.20 | 55+1 |
| 4 | Female | 32+5 | 1.75 | 41 |

PMA: Postmenstrual age.

Type I (disorganized outer retinal layers without cavitation); Type II (outer retinal disorganization with cavitation and neurosensory elevation, often accompanied by inner choroidal excavation), as proposed by Wong *et al*[11] in 2014; Type III (characterized by retinal thinning and hyperreflective spaces in the inner retinal layers superimposed on Type II lesions, but without subretinal clefts), first introduced by Tripathy *et al*[12]; and Type IV (isolated choroidal excavation with intact retinal architecture), recently reported in a case[13]. However, this classification system remains controversial due to uncertainties regarding distinct phenotypic differences among subtypes. In our study, Case 4 was classified as Type I based on OCT findings. OCT angiography (OCTA) demonstrates a 26% flow signal reduction in choroidal capillary within the lesion compared to healthy areas, with generally preserved superficial retinal capillary plexuses and mild involvement of the deep retinal capillary plexus[9]. Fluorescein angiography reveals well-demarcated hyperfluorescence with punctate hypofluorescence and no leakage[8].

TM is clinically diagnosed based on characteristic features but requires differentiation from congenital RPE hypertrophy, Gardner syndrome-associated RPE abnormalities, choroidal and retinal scars and so on. Congenital RPE hypertrophy presents as flat, hyperpigmented (rather than hypopigmented) lesions with round or scalloped margins, typically located in the equatorial or peripheral retina, rarely involving the macula[14]. Gardner syndrome-related RPE abnormalities resemble TM but are smaller, bilateral, irregularly shaped, and randomly distributed[14]. Choroidal and retinal scars are often multifocal, irregular, and may be associated with toxoplasmosis in children[15].

Most TM cases are stable, non-foveal, and asymptomatic, requiring no treatment. Foveal involvement may cause visual decline or metamorphopsia, though no definitive therapies exist. Rare cases with secondary choroidal neovascularization (CNV) may benefit from intravitreal anti-vascular endothelial growth factor (VEGF) agents, though long-term outcomes remain uncertain[16].

Torpedo maculopathy is a rare congenital RPE dysplasia. Although we have reported the first cases of torpedo maculopathy in preterm infants and delineated its clinical

manifestations, this finding does not suggest a causal link between prematurity and disease development. As TM is increasingly identified across diverse populations, its morphological spectrum continues to expand.

**ACKNOWLEDGEMENTS**

**Foundation:** Supported by the National Natural Science Foundation of China (No.82070991).

**Conflicts of Interest: Chen B**, None; **Yang H**, None.

**REFERENCES**

1 Roseman RL, Gass JD. Solitary hypopigmented nevus of the retinal pigment epithelium in the macula. *Arch Ophthalmol* 1992;110(10):1358-1359.

2 Daily MJ. Torpedo maculopathy or paramacular spot syndrome. *Paper presented at the New Dimensions in Retina Symposium* 1993:10-13.

3 Shirley K, O'Neill M, Gamble R, *et al*. Torpedo maculopathy: disease spectrum and associated choroidal neovascularisation in a paediatric population. *Eye* (*Lond*) 2018;32(8):1315-1320.

4 Shields CL, Guzman JM, Shapiro MJ, *et al*. Torpedo maculopathy at the site of the fetal "bulge". *Arch Ophthalmol* 2010;128(4):499-501.

5 Pian DB, Ferrucci S, Anderson SF, *et al*. Paramacular coloboma. *Optom Vis Sci* 2003;80(8):556-563.

6 Teitelbaum BA, Hachey DL, Messner LV. Torpedo maculopathy. *J Am Optom Assoc* 1997;68(6):373-376.

7 Hugo J, Beylerian M, Denion E, *et al*. Multimodal imaging of torpedo maculopathy including adaptive optics. *Eur J Ophthalmol* 2020;30(2):NP27-NP31.

8 Ding YH, Yao BT, Ye H, *et al*. Multimodal imaging of torpedo maculopathy in a Chinese woman: a case report. *BMC Ophthalmol* 2019;19(1):154.

9 Grimaldi G, Scupola A, Sammarco MG, *et al*. Morpho-functional evaluation of torpedo maculopathy with optical coherence tomography angiography and microperimetry. *Am J Ophthalmol Case Rep* 2018;10:165-168.

10 Lopez JM, Miere A, Rabinovic M, *et al*. Torpedo maculopathy: multimodal and retromodal imaging. *Can J Ophthalmol* 2024;59(1):e96-e99.

11 Wong EN, Fraser-Bell S, Hunyor AP, *et al*. Novel optical coherence tomography classification of torpedo maculopathy. *Clin Exp Ophthalmol* 2015;43(4):342-348.

12 Tripathy K, Sarma B, Mazumdar S. Commentary: Inner retinal excavation in torpedo maculopathy and proposed type 3 lesions in optical coherence tomography. *Indian J Ophthalmol* 2018;66(8): 1213-1214.

13 Light JG, Alvin Liu TY. A novel phenotype of torpedo maculopathy on spectral-domain optical coherence tomography. *Am J Ophthalmol Case Rep* 2020;20:100956.

14 Villegas VM, Schwartz SG, Flynn HW Jr, *et al*. Distinguishing torpedo maculopathy from similar lesions of the posterior segment. *Ophthalmic Surg Lasers Imaging Retina* 2014;45(3):222-226.

15 Goh EJH, Putera I, La Distia Nora R, *et al*. Ocular toxoplasmosis. *Ocul Immunol Inflamm* 2023;31(7):1342-1361.

16 Ghezzaz A, Idlefqih W, Chahed S, *et al*. Choroidal neovascularization in torpedo maculopathy treated by aflibercept: long-term follow-up using optical coherence tomography and optical coherence tomography angiography. *Retin Cases Brief Rep* 2023;17(4):433-437.