Unusual Torpedo Maculopathy

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INTRODUCTION

Torpedo maculopathy is a congenital abnormality of the Retinal Pigment Epithelium (RPE), clinically manifesting as an asymptomatic well-circumscribed oval-shaped region of chorioretinal hypopigmentation located temporal to the macula with a torpedo-like tip directed toward the foveola [1,2]. Torpedo lesions are typically unilateral, and may be associated with varying degrees of hyperpigmentation. We describe a case of torpedo maculopathy in an 8-year-old Japanese boy.

CASE REPORTS

An 8-year-old Japanese boy was referred to our hospital because of right fundus abnormality. He had no perinatal problems and his development was normal. He had exotropia in his right eye and he was unable to maintain his eyes in a central position by convergence. Best corrected visual acuity was 0.03 and 1.2 in his right and left eyes, respectively. Fundus examination revealed a sharply demarcated oval, hypopigmented lesion with irregular hyperpigmentation, measuring 1.5 × 4.0 disc diameters (DD), located in the temporal macula in the right eye (Figure 1A). Additionally, a similar lesion (0.5 × 1.0 DD) located above this lesion, dragged disc, and discolorations at the macula were also observed. The fellow eye was unremarkable (Figure 1B). Optical coherence tomography (OCT: RS-3000; Nidek, Gamagori, Japan) revealed inner retinal hyperreflectivity with reduced thickness (Figure 1C, arrows) and disruption of the photoreceptor and RPE (Figure 1C, arrowheads).

DISCUSSION

Torpedo lesion is usually solitary, small, and benign lesion associated with normal visual acuity and normal visual fields [1,2]. However, in our case, torpedo lesion was relatively large and was associated with decreased visual acuity.Bedar et al. [3] described

Figure 1 Right (A) and left (B) fundus photographs of an 8-year-old boy with torpedo maculopathy. The right eye had a sharply demarcated oval hypopigmented lesion with irregular hyperpigmentation and dragged discolored in the temporal macula. Optical coherence tomography (C) revealed inner retinal hyper reflectivity with reduced thickness (arrows) and disruption of the photoreceptor and retinal pigment epithelium (arrowheads).
a case of torpedo maculopathy with elliptically configured yellowish alternations with irregular hyperpigmentation, measuring 1.5×3.0DD. Their fund us changes were very similar to those in our case. Moreover, two torpedo lesions were observed in our case. This condition is extremely rare and unusual.

OCT images from eyes with torpedo maculopathy show thin RPE with increased signal transmission in the choroid as well as a "cleft" in the outer retina associated with loss of photoreceptors, irregular edges of the residual photoreceptors, and thinning of the outer retinal layer. Our OCT findings including reduced retinal thickness, disruption of the photoreceptor, and RPE with a "cleft" formation, were similar to those in previous reports. Moreover, we speculate that hyper reflectivity in the inner retina results from alternation of retinal micro-architecture.

The cause of torpedo maculopathy is unknown. Golchet et al. [1] speculated that the lesion results from a disturbance in the normal development of the choroid or ciliary vasculature. Shields et al. [2] have suggested that the uniform location and size of the lesion indicates a persistent defect in the development of the RPE in the fetal temporal bulge.

Torpedo maculopathy typically remains unchanged over time; however, long-term follow-up will be necessary to further understand this condition.

REFERENCES