Torpedo maculopathy: A primary choroidal capillary abnormality?

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A 26-year-old healthy male patient's fundus revealed findings consistent with torpedo maculopathy. Swept-source optical coherence tomography (OCT) showed a dome-shaped elevation of the retina at the level of ellipsoid zone. On OCT angiography segmented at the level of the choriocapillaris, a cluster of convoluted fine vessels was seen, and further, deeper scans of the larger choroidal vessels showed a slower flow. From these observations along with the embryological correlation of choriocapillaris development, a possibility of an abnormality preventing proper fenestration of the choriocapillaris along the horizontal raphe being responsible for this anomaly is suggested.

Key words: Choriocapillaris, optical coherence tomography angiography, torpedo maculopathy

Torpedo maculopathy is considered to be a benign congenital lesion typically located along the horizontal raphe. It resembles a torpedo (submarine missile) measuring wider horizontally than vertically with the tip of torpedo pointing toward the fovea. It may be associated with satellite lesions located usually temporal to the lesion. It was first described by Roseman and Gross in 1992, before which it was labeled as solitary hypopigmented nevi of retinal pigment epithelium. Here, a case of torpedo maculopathy has been analyzed using swept-source optical coherence tomography (SSOCT) and OCT angiography (OCTA), and the findings have been discussed correlating with the embryology.

Case Report

A 26-year-old female patient presented for routine ophthalmic evaluation. Visual acuity was 20/20 in each eye. Fundus examination of the right eye revealed a fairly well-defined ellipsoid area of variable pigmentation just above the horizontal raphe and temporal to the fovea [Fig. 1a]; left eye fundus was within normal limits. Fundus autofluorescence better defined

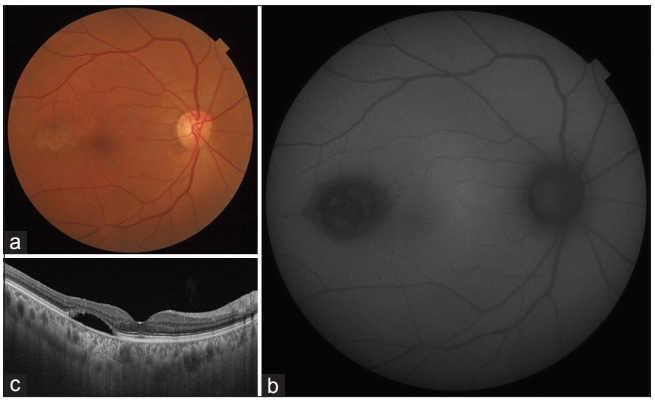

Figure 1: (a) Fundus picture of the right showing a fairly defined ellipsoid area of variable pigmented lesion temporal to the fovea. (b) Autofluorescence of the same autofluorescence better defines the torpedo-like lesion. The autofluorescence pattern is variegated. A central hypofluorescent area with internal focal areas of hyperfluorescent and a rim of hyperfluorescent around the lesion is seen. (c) Swept-source optical coherence tomography revealing anterior lift of the ellipsoid layer forming a cavity with intact inner retinal layers

the torpedo-like lesion. However, the autofluorescence pattern was variegated [Fig. 1b]. SSOCT showed a dome-shaped cavity temporal to the fovea due to an anterior elevation of the ellipsoid zone from an intact retinal pigment epithelium-Bruch's membrane complex; the external limiting membrane and layers anterior to it appeared intact [Fig. 1c]. OCTA did not reveal any abnormality within the superficial or deeper retinal plexus. On segmenting below the RPE, changes were visible in the choriocapillaris layer. A convoluted pattern of fine vessels with some empty spaces between them was visible beneath the area of the separation of neurosensory retina on SSOCT [Fig. 2a]. This was more marked on the nasal and middle portion (red outlined arrow) of the lesion and appeared to slowly merge with the normal pattern on the choriocapillaris on the temporal half (blue outlined arrow). The signal strength by an OCTA depends on erythrocyte movement (flow rate and flow volume) indicating the vessels could be of a larger caliber than the normal choriocapillaris. On segmenting further below the level of choriocapillaris, the larger choroidal vessels with slower flow were seen as dark channels (red arrows). Some of these were masked by a projection artifact of the overlying anomalous vessels (area within yellow arrows) [Fig. 2b].

Discussion

The exact etiology of torpedo maculopathy remains speculative, but based on a consistent location among the reported cases and case series,[1-4] it could be due to (1) retinal pigment epithelial defect during fetal temporal bulge, (2 malformed emissary

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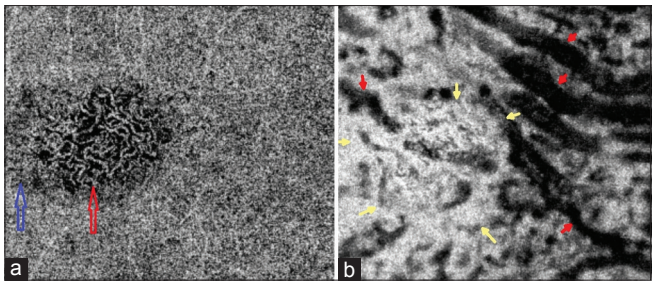

Figure 2: (a) Optical coherence tomography angiography segmented below the retinal pigment epithelium shows a convoluted pattern of fine vessels with some empty spaces between them beneath the area of the separation of neurosensory retina. This is more marked on the nasal and middle portion (red outlined arrow) of the lesion and appears to slowly merge with the normal pattern on the choriocapillaris on the temporal half (blue outlined arrow). (b) On segmenting further below the level of choriocapillaris on optical coherence tomography angiography, the larger choroidal vessels with the slower flow are seen as dark channels (red arrows). Some of these are masked by a projection artifact of the overlying anomalous vessels (area within yellow arrows)

canal of long posterior ciliary artery and the nerve, or (3) defective differentiation of the arcuate nerve fiber bundles along the horizontal raphe. Two types have been described on spectral domain OCT; Type I has an attenuation of the outer nuclear layer, interdigitation zone, and ellipsoid zones in the absence of any cavitation of the outer layers, whereas Type II has outer retinal layer attenuation along with cavitation.[1] Our case resembles Type II due to the presence of a cavity above the RPE.

RPE develops from the outer walls of the optic cup. The reported cases reveal that the RPE could be atrophic, thin, or absent,[3,5-7] leading to hypofluorescence in the central area of the torpedo lesion with a peripheral hyperfluorescent rim on autofluorescence imaging. The cause for peripheral hyperfluorescence is not very clear. In our case, there was a variegated appearance (the central hypointense area had few focal patches of hyperfluorescence) with a surrounding rim of hyperfluorescence. The formation of choriocapillaris has been observed with immunohistochemical markers from 6 to 22 weeks and beyond. The formation of choriocapillaris from an island of progenitor cells is probably responsible for the formation of a lobular pattern which subsequently coalesces to form a network. The inner segment photoreceptors are formed around 22–26 weeks after which further differentiation takes place.[8] Following this, the choriocapillaris gets fenestrated on the RPE/photoreceptor side. Two recent studies of OCTA features in cases of torpedo maculopathy have also documented a loss of choriocapillaris in the area of the lesion.[4,5] Thus, an abnormality in formation of choriocapillaris could be involved in the pathogenesis of torpedo maculopathy. However, since the imaging in these studies was not done on swept-source OCT, the deeper underlying abnormal convoluted vessels and further deeper choroidal vasculature probably could not be analyzed. In one study, the authors mention that the Sattler's layer appears to extend to the cleft.[5] They might be referring to something similar to our description of abnormal convoluted vessels. In our opinion, the convoluted vessels do not seem to be the vessels of the Sattler's layer. Sattler's layer can be clearly seen on OCTA under patches of geographic atrophy and appears to have a much more arborizing pattern.

Conclusion

We cannot conclusively establish the etiology based our single case report. Yet, considering all the above observations, we suggest another possible hypothesis regarding the formation of torpedo maculopathy. We propose that an abnormality preventing proper fenestration of the choriocapillaris along the horizontal raphe may be primarily responsible for this anomaly.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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