Torpedo maculopathy with multifocal central serous chorioretinopathy: A rare case report

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We present a very rare case of torpedo maculopathy (TM) with multifocal central serous chorioretinopathy. A 26-year-old male presented with painless loss of vision in the right eye of 2 months duration. Clinical examination showed a torpedo-shaped lesion temporal to fovea and subretinal fluid in foveal center. Fluorescein angiography showed multifocal leaks. Optical coherence tomography showed an optically clear space/neurosensory detachment at the site of lesion. Patient underwent focal laser to the leaks. TM is a rare congenital disorder detected accidentally during routine fundus examination. It is usually unilateral and does not affect vision.

Key words: Central serous chorioretinopathy, multifocal, neurosensory detachment, optical coherence tomography, torpedo maculopathy

Torpedo maculopathy (TM) is a very rare congenital defect of the retinal pigment epithelium. The diagnosis of the disorder is mostly clinical. It is characterized by a torpedo-shaped hypopigmented patch located temporal to the fovea.^[1] The lesion is usually unilateral and is rarely associated with vision loss. The condition has been variously reported in English literature.^[1-9] We report a very rare case of TM associated with multifocal central serous chorioretinopathy (CSCR). To the best of our knowledge, this is the first case description of the disorder from India.

Case Report

A 26-year-old healthy male presented with gradual, painless loss of vision in the right eye (RE) of 2-month duration. There was no history of any trauma or ophthalmic surgery. Patient had no systemic complaints and was not on any systemic or topical medications. Best-corrected visual acuity at initial presentation was 20/32, N12 in RE and 20/20, N6 in the left

Access this article online	
Quick Response Code:	Website:
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	DOI: 10.4103/ijo.IJO_812_17

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Manuscript received: 01.09.17; Revision accepted: 25.10.17

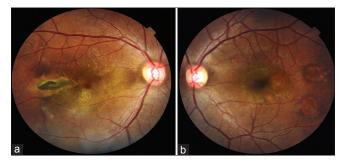


Figure 1: (a) Color fundus photograph of the right eye showing a torpedo-shaped lesion temporal to the fovea. Subretinal fluid seen in the foveal area and inferior to it with few yellowish subretinal deposits. (b) Color fundus photograph of the left eye showing multiple pigment epithelial detachments temporal to the fovea

eye (LE). Anterior segment examination including intraocular pressure was normal in both eyes.

Fundus examination of the RE showed an elongated oval "torpedo-"shaped lesion located temporal to fovea [Fig. 1a]. The lesion was hyperpigmented with hypopigmented margins and sharp nasal margin toward fovea. Subretinal fluid (SRF) was noted involving the foveal center and inferior to the fovea. Few yellowish subretinal deposits were present within the SRF. Funduscopy of LE showed multiple pigment epithelial detachments (PEDs) temporal to foveal center [Fig. 1b].

Fundus fluorescein angiography (FFA) of RE showed inkblot pattern of leaks, temporal to the optic disc, and inferior to foveal center which kept on increasing in size and intensity as the angiography progressed. The torpedo lesion was hypofluroscent in the center and showed staining at its borders [Fig. 2]. FFA of LE showed numerous PED temporal to the fovea. Spectral domain optical coherence tomography (OCT) in RE showed an optically clear area at the site of the torpedo lesion. Few small intraretinal cystic spaces were noted in the inner retina overlying the lesion. There was mild disorganization of the inner retina. Large neurosensory detachment (NSD) involving the foveal center and a PED nasal to the foveal center was seen [Fig. 3a]. Few hyperreflective structures were noted on the under surface of the NSD which could be the subretinal deposits/fibrin as seen during fundoscopy [Fig. 3b]. OCT of LE showed normal foveal dip with PED temporal to it.

A diagnosis of TM with CSCR in RE was made. The patient underwent focal laser to the leakage points. We are now awaiting further follow-up of the patient.

Discussion

TM is a rare congenital, usually unilateral, torpedo-shaped hypopigmented lesion noted temporal to the fovea.^[1] The

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Cite this article as: Panigrahi PK, Minj A, Satapathy J. Torpedo maculopathy with multifocal central serous chorioretinopathy: A rare case report. Indian J Ophthalmol 2018;66:330-1.

Figure 2: (a) Early phase of fundus fluorescein angiography shows two very small leak points (white arrows). The torpedo lesion shows hypofluorescence in the central part with hyperfluorescence at the margins. (b and c) Increase in both size and intensity of the leak points with progression of the fundus fluorescein angiography

lesion has been described as paramacular coloboma.^[2] The pathogenesis of such lesions is still highly debatable. Pian *et al.*^[2] considered incomplete differentiation of the arcuate bundles during development along the horizontal raphe of the macular architecture to be the cause of the lesion. Teitelbaum *et al.*^[3] hypothesized that disturbance of the posterior ciliary arteries and veins before birth produce a retinal pigment epithelium defect resulting in a torpedo-like lesion. Two essential features of TM are the torpedo-shaped lesion and location in the temporal macula. Both these features are present in our case. The nasal margin of the lesion is slightly pointed and directed toward the foveal center.

Torpedo lesions usually do not tend to involve the foveal center and the vision is usually normal.^[1,2] Lesions vary between 1 and 2 disc diameters in their horizontal extent and 0.5–1 disc diameter in their vertical extent.^[4,5] Published reports find that the nasal edge of the lesion thins out and disappears within one disc diameter of the foveal center.^[1,2] In our case too, the nasal margin of the lesion thins out within 1 DD of the foveal center. Lesions are usually unilateral, but bilateral cases have been reported.^[6] TM usually does not lead to any loss of vision. If present, it can be attributed to CSCR.

Optical coherence tomography findings of the lesion include an optically clear space between the RPE and neurosensory retina which has been described as NSD.^[7,8] This clear space might be due to photoreceptor degeneration or loss of RPE at the borders of the lesions. Other OCT findings include mild disorganization of outer and inner retina.^[5,9] Our case has similar OCT findings of a NSD at the site of lesion and inner retina changes overlying the lesion.

Conclusion

TM is a rare lesion which might be accidentally seen during routine fundus examination. The lesion is usually nonprogressive and not associated with vision loss. We have presented a rare case of TM associated with multifocal CSCR. Both these conditions might not be related to each other. There is a possibility of the association being coincidental rather than causal. Such association has not been reported previously. This is the first reported case of TM from India to the best of our knowledge. Our report adds to the body of literature available on TM.

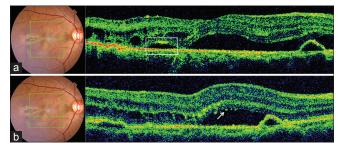


Figure 3: (a) Optical coherence tomography scan passing through the torpedo lesion shows neurosensory detachment (white box). Cystic changes can be seen in the inner retina overlying the lesion. Scan passing through the foveal center shows neurosensory detachment because of the underlying subretinal fluid and a pigment epithelial detachments nasal to the foveal center. (b) Optical coherence tomography scan passing below the foveal center shows the presence of neurosensory detachment and a small pigment epithelial detachments. Scan passing through the yellowish subretinal lesions show hyperreflective dots (white arrow) on the under surface of the neurosensory detachment suggestive of fibrin

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.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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