Dome shaped maculopathy with tilted disc misdiagnosed as central serous chorioretinopathy

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DESCRIPTION

A 63-year-old woman presented with blurring of vision in the left eye (LE) of unknown duration, and had been treated as central serous chorioretinopathy (CSCR). The visual loss was non-progressive and painless. There was no history of previous trauma or surgical interventions. The best corrected visual acuity was 6/6 (axial length = 25.11 mm, refractive error = −0.25 DS/−2.5 DC @ 14°) and 6/18 (axial length = 24.55 mm, refractive error = −0.25 DS/−2.0 DC @ 1°) in the right eye (RE) and left eyes, respectively, and both eyes had brisk pupillary reactions. Apart from early nuclear sclerosis of the lens, bilateral anterior segments and intraocular pressures were within normal limits. Fundus examination revealed peripapillary scleral crescent, increased fundus tessellations in inferior fundus and a dull foveal reflex in both eyes (LE>RE) (figure 1). Swept-source optical coherence tomography (SS-OCT) of RE showed a dome shaped elevation of the macular region. The choroid had irregular thickness. The LE SS-OCT images showed central sub retinal fluid (SRF) with an even more remarkable elevation of the macular region as seen on the three-dimensional (3D) map, with irregular choroidal thickness (figures 1 and 2). The diagnosis was hence changed to dome shaped maculopathy (DSM) with tilted disc (TD). TD was inferred due to the 3D asymmetry of optic nerve head as it entered the sclera, peripapillary crescent that involved area inferior to disc, disc torsion and asymmetric presence of tessellations in the inferior fundus. Both fluorescein and indocyanine green angiography did not reveal leakage.

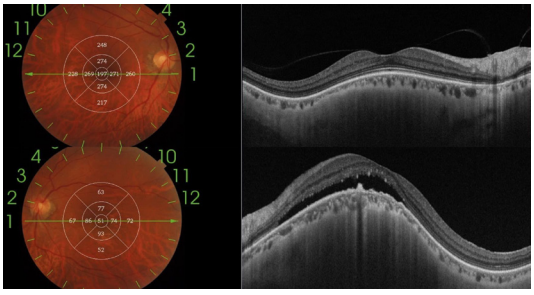

Figure 1 Fundus photographs and OCT pictures of the right eye (upper panel) and the left eye (lower panel). Dome shaped maculopathy is easily appreciable (LE>RE), and the choroidal thickness can be seen to be irregular. LE additionally shows areas of thickened RPE, with sub retinal fluid and deposits on the outer surface of the neurosensory retina. The scleral thickness appears to be increased in both eyes, though outermost sections of the sclera are not clearly imaged due to poor OCT penetration. LE, left eye; OCT, optical coherence tomography; RE, right eye; RPE, retinal pigment epithelium.

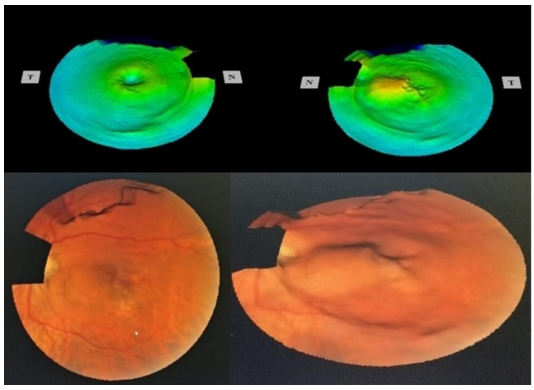

Figure 2 3D map images as reconstructed from the swept-source optical coherence topography images. Upper panel shows the dome to be smoother in the right eye and uneven and exaggerated towards the optic disc in the left eye. Lower panel shows the 3D images as superimposed on the fundus of the images of the LE. These images give a realistic and readily understandable picture of the dome. 3D, three dimensional; LE, left eye.

Presence of SRF is typical of CSCR. However CSCR is known to have a large number of macular masqueraders. Hence, presence of any atypical sign should arouse the suspicion of an alternative pathology, in our case the TD. In cases of TD, there is partly normal and partly abnormal curvature of posterior sclera.[1 2] DSM is considered by most to be a scleral anomaly and as seen in our case, has been rarely reported with TD in absence of a staphyloma.[1] As opposed to thin sclera of myopic DSM, other types of DSM show a thickened sclera that impairs fluid outflow and vascular supply, subsequently impairing the RPE and choroidal function and leading to findings like pigment epithelium detachment and SRF.[2] It has been demonstrated that DSM may be missed when only single B scan images are used, prompting use of radial OCT scans.[3] In our case, the 3D map offered a very realistic view of the anomaly due to readily understandable images of the dome (figure 2).

The treatment options of photodynamic therapy, laser photocoagulation and anti-vascular endothelial growth factor therapy have only a limited success and should be considered only in cases of persistent or quickly recurrent SRF and deterioration of vision.[1] As spontaneous resolution of SRF in DSM has also been seen and stability of vision is known in cases with persistent SRF, we observed the patient. The clinical condition of this patient remained stable until a follow-up of 3 months.

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Learning points

- Presence of subretinal fluid in cases of dome shaped maculopathy can lead to misdiagnosis of central serous chorioretinopathy.
- Findings like tilted disc should alert the surgeon to possibility of associated posterior scleral contour anomalies. Three-dimensional optical coherence tomography maps offer an excellent way of studying such anomalies.

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