Extramacular dome-shaped elevation: a novel finding in a case of high myopia

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DESCRIPTION

Dome-shaped macula (DSM) is an elevation at the macula seen in about 5%–10% cases of high myopia, usually within a posterior staphyloma, caused by localised inward scleral and choroidal convexity. It may lead to visual deterioration when associated with serous foveal detachment (44% cases). Herein, we describe a similar but ‘Extramacular dome-shaped elevation (EDSE)’ associated with a large retinal hole.

A 23-year-old myopic female patient was referred to our clinic for pre refractive surgery fundus screening. She had a history of diminution of vision in the left eye more than the right eye since childhood and was using myopic glasses for the same. Best-corrected visual acuity on Snellen chart was 20/20 in the right eye and 20/200 in the left eye. Lower acuity in the left eye was attributed to anisometropic amblyopia as her refractive error was –6.75 D sphere OD and −17.25 D sphere OS with an axial length of 26.62 mm OD and 32.85 mm OS on IOL Master (Carl Zeiss, Jena, Germany). Fundus examination on indirect ophthalmoscopy revealed a myopic fundus with tessellations and tilted disc, Curtin type I posterior staphyloma, myelinated nerve fibres, a large hole superiorly above the retinal vascular arcades of about three disc diameter size along with a Weiss ring suggestive of a complete posterior vitreous detachment in the left eye (figure 1A). Fundus examination was per se normal without any myopic degenerative changes in the right eye.

Swept source optical coherence tomography (SSOCT) (Triton - Topcon, Tokyo, Japan) of the left eye: radial scans (12 mm) passing via the fovea showed a dome-shaped macula with increased foveal thickness (539 µ) and cystoid changes at the fovea with altered foveal contour. Radial scans (12 mm) passing via the retinal hole showed a minimum hole diameter of 3341 µ and an underlying dome-shaped elevation with a choroidal thickness of 114 µ and an inward convex bulge of sclera (white arrow). A 12 mm scan via the fovea in the left eye (B) shows a dome-shaped macula (black arrow), altered foveal contour and an increased retinal thickness of 539 µ with cystoid changes (white asterisk). The 12 mm scan via the fovea in the right eye (C) showing normal foveal contour with a thickness of 211 µ (all measurements were done manually).

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Various theories proposed for DSM are sublesional scleral thickening, inward convex contour of the choroid and sclera and localised choroidal thickening. Inward convex contour of the sclera and choroid along with sublesional scleral thickening was present in our case at the site of DSM and EDSE, and thus it appears to contribute towards the formation of both DSM and EDSE, suggesting that they originate from a similar but unknown pathophysiology. To the best of our knowledge, an EDSE alone or along with a large retinal hole, as in our case has not been reported before.

Figure 1 Optos widefield fundus photograph (A) of the patient’s left eye showing tessellated fundus, tilted disc, superotemporal myelinated nerve fibres (red asterisk) and a large retinal hole above the superior arcade (black arrow). Fluorescein angiography (B) and indocyanine green angiography figures (C) are suggestive of window defect through the retinal hole (white arrow).
Learning points

- Dome-shaped elevation can be found in extramacular location also, in high myopic eyes with posterior staphyloma.
- Both dome-shaped macula and extramacular dome-shaped elevation (EDSE) occur as a result of inward scleral bulge and can be diagnosed on optical coherence tomography.
- EDSE can be associated with a retinal hole.

Contributors

VM: Corresponding and first author; AK: Case selection and imaging; RC: Case description and image analysis; RDR: Text editing and literature search.

Competing interests
None declared.

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