Swept-source optical coherence tomography angiography of choroidal neovascularization in vertically oriented oval dome-shaped maculopathy

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A 48-year-old female presented with complaints of recent onset diminution of vision of the left eye (OS) for the past 2 months. She was highly myopic and was using glasses for the past 30 years. Ocular examination revealed presence of a myopic fundus with high axial lengths in both the eyes. Fundus examination of the OS revealed a myopic tessellated fundus with prominent choroidal vessels and a blunted foveal reflex. There was a small pale whitish lesion just superior to the foveal center. Optical coherence tomography (OCT) scans (both horizontal and vertical) confirmed presence of dome-shaped maculopathy. There was subretinal fluid in the OS. A vertical OCT scan also revealed a subretinal hyperreflective material, which was confirmed to be due to a small mixed type 1 and type 2 choroidal neovascularization (CNV) on swept-source (SS) OCT angiography in the OS. The patient was given intravitreal injection of ranibizumab (0.5 mg/0.05 mL) in the OS. At 1-month follow-up, the subretinal fluid completely resolved. The CNV lesion regressed significantly on SS-OCT angiography. The best-corrected visual acuity improved from 20/80 to 20/20 in the OS, which was maintained at 3 months of follow-up.

Key words: Choroidal neovascularization, dome-shaped maculopathy, OCT, optical coherence tomography angiography, ranibizumab, swept-source

Dome-shaped maculopathy (DSM) is a rare condition characterized by a forward convex bulge of the macula that is observed in highly myopic eyes with staphylomas using imaging tools such as optical coherence tomography (OCT).[1-3] Sclerochoroidal thickness abnormalities are observed in the macular areas in cases with DSM.[1,4,5] These changes may be easily missed on cursory examination unless a detailed evaluation of the macula is performed using variably oriented OCT scans.[6] Eyes with DSM may develop various macular complications such as serous retinal detachments (SRDs) greatly impacting central vision.[7,8] SRD is observed in eyes with DSM in the presence of higher macular bulge, or associated with pigment epithelial detachments (PEDs).[7] Choroidal neovascularization (CNV) is a rare complication of DSM, which may also be associated with SRD.

Imaging tools such as OCT angiography (OCTA) have greatly revolutionized the management of retinal diseases. Noninvasively, these imaging modalities are able to detect abnormal microvascular networks observed on conventional techniques such as fluorescein angiography (FA) and indocyanine green angiography (ICGA).[9-11] In this index case report, we describe serial imaging of a rare case of a 48-year-old lady who presented with diminution of vision due to development of a mixed (type 1 and type 2) CNV and SRD at the edge of a vertically oriented dome.

Case Report

A 48-year-old lady presented to the Department of Ophthalmology with chief complaints of blurring of vision in her left eye (OS) for the past 2 months. The visual symptoms were insidious in onset and continued to persist during this period. She was a high myope and using spectacle correction of −8.25 diopters in the right eye (OD) and −7.75 diopters in the OS. At another center, she was diagnosed with myopic maculopathy and referred for further opinion and management. There was no systemic history of any medical illnesses, and family history was noncontributory. She was a nonhypertensive and nondiabetic. There was no history of intake of any medicines or eye drops in the past 6 months.

On examination, her best-corrected visual acuity (BCVA) was 20/25 in the OD and 20/80 in the OS (measured by Snellen's visual acuity chart for distance). The intraocular pressure by Goldmann applanation tonometry was 16 and 18 mmHg in OD and OS, respectively. Slit-lamp examination revealed a normal anterior chamber with no signs of inflammation. Posterior segment examination revealed a clear media. Retinal examination revealed tessellated fundus with prominent choroidal vasculature in both the eyes (OU) with a small peripapillary myopic crescent. Foveal reflex appeared sharp in OD and blunted in OS. Fundus autofluorescence (FAF) imaging revealed a normal pattern of autofluorescence in OD and a small, subtle area of hyperautofluorescence superonasal to the foveal center. Combined FA and ICGA were performed. The angiographic images of OD did not reveal any abnormality. In OS, there was an area of hyperfluorescence corresponding to the hyperautofluorescent lesion on FAF in the early and late phases [Fig. 1; only OS shown]. The FA image revealed mild,

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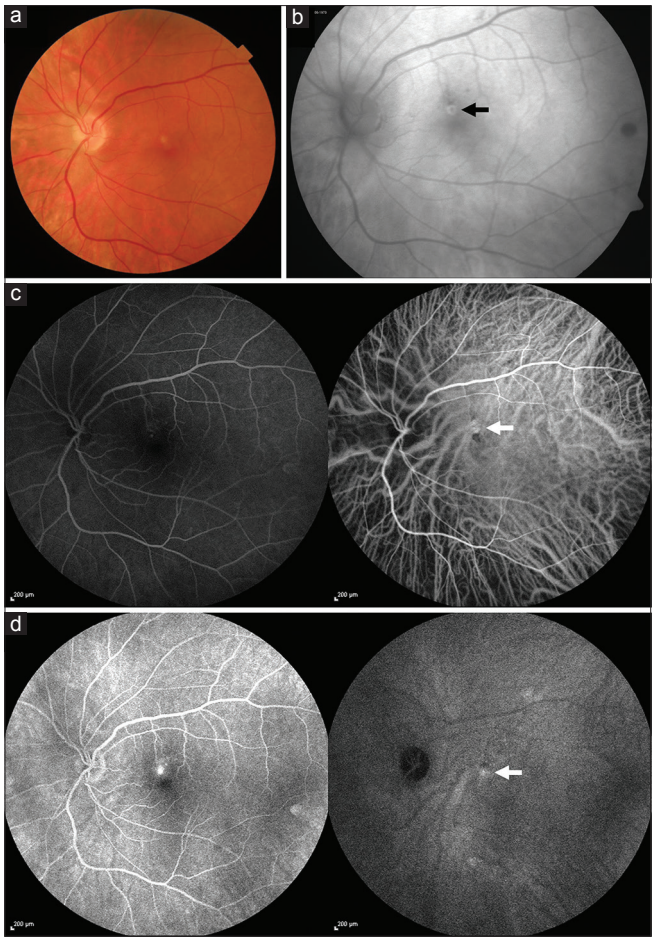

Figure 1: (a) A colored fundus photograph of the patient with dome-shaped maculopathy (DSM) shows a tessellated fundus with prominent choroidal vasculature and a blunted foveal reflex. (b) Autofluorescence imaging (FAF) shows a small area of hyperautofluroescence (black arrow). (c) Combined fluorescein angiography (FA) and indocyanine green angiography (ICGA) of the same patient shows a subtle increased hyperfluorescence in the same area of hyperautofluorescence on FAF. In the late phase (a), FA shows a hyperfluroescent lesion which shows mildly increased hypercyanesence on ICGA. Prominent large choroidal vessels are marked by a white arrow in (c and d)

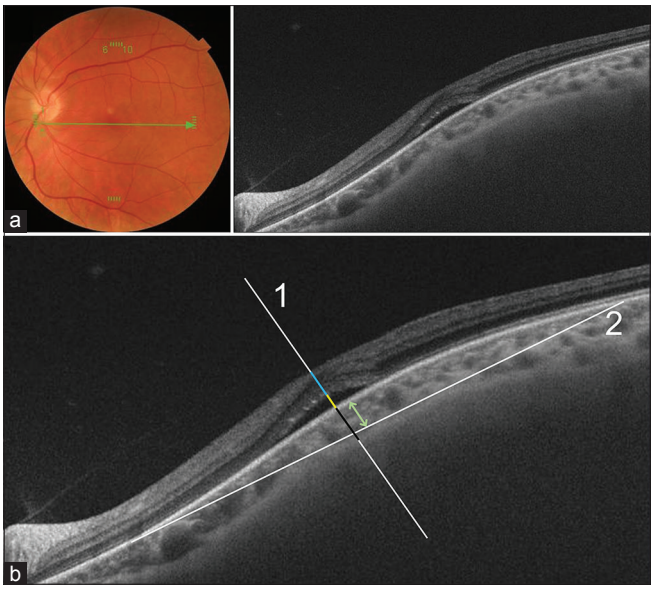

Figure 2: Horizontal swept-source optical coherence tomography (SS-OCT) B-scan line passing through the fovea reveals a dome-shaped maculopathy (DSM) with serous retinal detachment (SRD) (a). (b) Illustrative description of measurements is shown. Line 1 (passing through the foveal center) is a perpendicular line to Line 2, which is a tangent drawn to the outer border of the retinal pigment epithelium (RPE) at the edges of the bulge. Blue portion on line 1 represents retinal thickness, yellow portion represents the height of the SRD, and the black portion represents the choroidal thickness. Green line with double arrowheads is the height of the macular bulge (distance of line 2 from the RPE)

focal, ill-defined early hyperfluorescence, which increased in the late phase. The ICGA showed mildly increased focal hyperfluorescence in the early and late phases.

A horizontal swept-source (SS)-OCT B-scan passing through the center of the fovea revealed a convex, anteriorly protruding macula suggestive of DSM. There was presence of SRD which could be visualized in the same horizontal B-scan of the OCT [Fig. 2]. The measurements of the macular bulge height were obtained as described by Caillaux *et al*.[6] Briefly, a line was drawn through the center of the fovea (Line 1) perpendicular to another tangent line (Line 2) along the outer border of the RPE at the bottom of the staphyloma. The distance between the intersection of Line A with RPE and intersection of Line A and Line B was measured as the bulge height. The retinal thickness at the fovea was 196 μm, and the choroidal thickness was 263 μm. The macular bulge height was 206 μm, and the height of SRD was 123 μm [Fig. 2]. A vertical OCT B-scan was also obtained passing through the center of the macula. The area corresponding to the subtle area of hyperautofluorescence on FAF imaging appeared as a small outer retinal hyperreflective lesion (at the edge of the SRD) with pigment epithelial elevation on vertical OCT. SS-OCTA passing through this lesion revealed a small CNV with loopy vessels and intricate branching. The corresponding OCT B-scan showed high flow through the neovascular lesion [Fig. 3].

The patient was counselled regarding antivascular endothelial growth factor (anti-VEGF) therapy. After explaining the risks and potential benefits, the patient elected for therapy and was administered intravitreal ranibizumab (0.5 mg) injection. At a follow-up visit 4 weeks later, the BCVA in OS improved from 20/80 to 20/25. The SRD regressed completely in OS on both horizontal and vertical B-scan OCT. The area of outer retinal hyperreflectivity also resolved, and only an area of RPE irregularity and mild elevation persisted. OCTA showed reduction in the neovascular flow signal and significant regression of the loopy vessels [Fig. 3].

At 3-month-follow-up, the BCVA was maintained at 20/25, and there was no recurrence of SRD on OCT [Fig. 4]. There was no change in the neovascular flow signal on OCTA, and the patient was asymptomatic.

Discussion

In highly myopic eyes, DSM is a rare entity that has been recently evaluated using OCT imaging. In 2013, Caillaux *et al.* proposed a classification of DSM based on the orientation

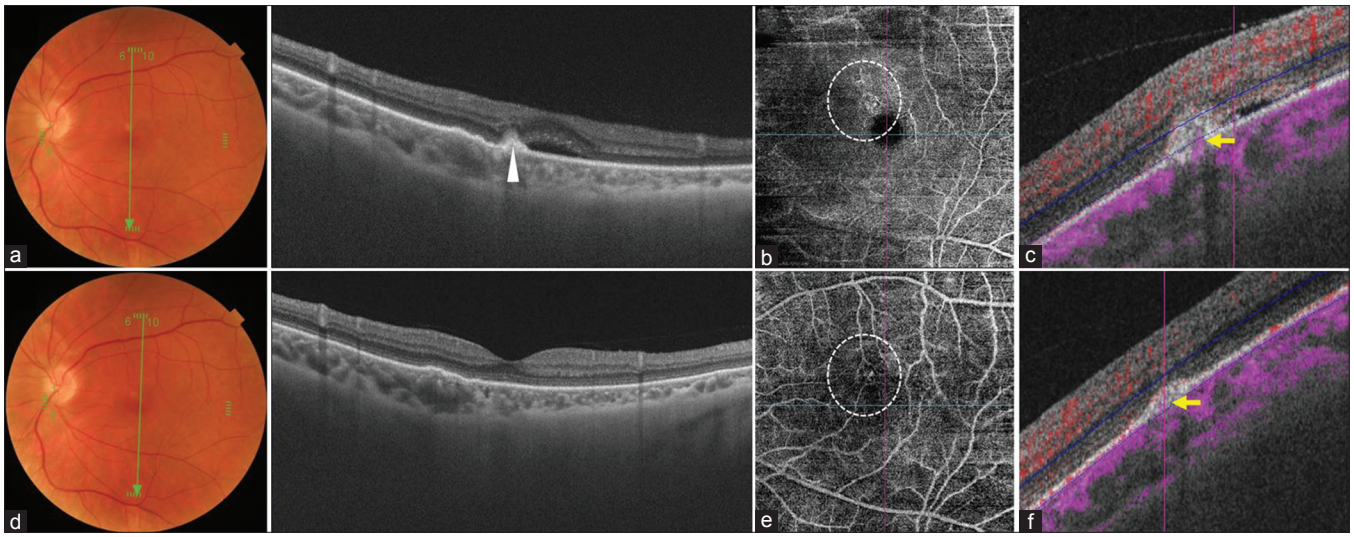

Figure 3: (a) A vertical swept-source optical coherence tomography (SS-OCT) B-scan passing through the fovea shows presence of an outer retinal lesion (white arrowhead) at the edge of the serous retinal detachment (SRD). On swept-source optical coherence tomography angiography (SS-OCTA), there is a small vascular network of loopy vessels (b) (white dashed circle), which shows neovascular flow signals on corresponding OCT B-scan (c) (yellow arrow). Follow-up vertical SS-OCT line scan passing through the fovea shows resolution of the outer retinal lesion and SRD (d). A residual irregularity and elevation of the retinal pigment epithelium is seen. Corresponding follow-up SS-OCTA (e) shows regression of the choroidal neovascular network (white dashed circle) and disappearance of the neovascular flow signal on the OCT B-scan (f) (yellow arrow)

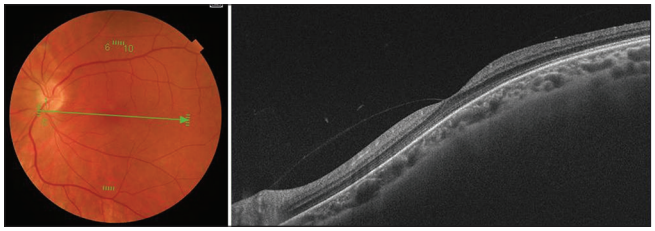

Figure 4: Follow-up horizontal swept-source optical coherence tomography (SS-OCT) B-scan line passing through the fovea at 3-month follow-up shows resolution of the serous retinal detachment (SRD)

of the dome: round dome (21% – dome present on both horizontal and vertical OCT scans), horizontally oriented oval dome (63% – dome visible on vertical OCT B-scan), and vertically oriented oval dome (16% – dome visible on horizontal OCT B-scan).[6] Our patient presented with vertically oriented dome, which is the rarest of the three types but easily detectable on horizontal OCT B-scan.

Macular complications such as SRD are a major cause of visual morbidity in eyes with DSM.[5,7,12,13] Till date, the pathogenesis of SRD in eyes with DSM is unclear but has been attributed to higher macular bulge height. It has been postulated that SRD in DSM may occur due to slow leakage of fluid from PEDs, potentiated by the thick underlying choroid beneath the bulge. Mechanisms similar to those responsible for SRD in central serous chorioretinopathy may also lead to accumulation of subretinal fluid in DSM.[5] Imamura *et al.* postulated that localized thickening of the sclera in eyes with DSM may result in abnormal choroidal blood flow and RPE function leading to SRD.[5] Ohsugi *et al*.[12] in their series of 49 myopic eyes observed SRD in five eyes (10.2%); Ellabban *et al*.[14] observed SRD in 5.9% eyes, and Imamura *et al.* observed SRD in 9% eyes.[5] On the other hand, Gaucher *et al.* observed SRD in 5 of 15 eyes (66.7%).[1] Caillaux *et al.* observed SRD in 25 of 48 eyes (52.1%) with DSM.[6] However, while SRD may be frequently observed in eyes with DSM, there is no satisfactory therapeutic modality that can effectively reduce the amount of subretinal fluid.[7,13,15,16]

CNV is a rare cause of SRD in eyes with DSM. While CNV has been reported as a complication in eyes with DSM by Ohsugi *et al*., Caillaux *et al*., and Imamura *et al*., among others, the detection was entirely based on OCT imaging alone.[5,6,12] There have been no studies using OCTA to detect CNV and determine the cause of SRD in these eyes. In our case, SS OCTA helped in detecting neovascular flow lesion responsible for accumulation of subretinal fluid. While FA images showed presence of a hyperfluorescent lesion, it could not definitively confirm the presence of CNV meriting treatment. Following anti-VEGF therapy, the SRD resolved completely, and mixed CNV also regressed leaving behind an area of irregular RPE elevation (regressed type 1 lesion).[17] Our patient had significant visual improvement which persisted till her last follow-up at 3 months.

In summary, SRD is a major cause of visual disturbance in eyes with DSM. Using novel technologies such as SS OCTA, it may be possible to identify the presence of macular complications such as CNVs which represent a treatable cause of SRD (unlike the chronic form of SRD due to high foveal bulge and abnormal choroidal flow). Further studies that evaluate eyes with SRD and DSM using SS OCT and OCTA are needed to determine the true incidence of CNV in these patients.

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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Nil.

Conflicts of interest

There are no conflicts of interest.

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