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REVIEW ARTICLE

Dome-shaped maculopathy: a review

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Abstract

First described by Gaucher and associates in 2008 in eyes with high myopia, dome-shaped maculopathy (DSM) is an anterior convex protrusion of the macula towards the vitreous cavity observable on OCT. This seems to be related to a localized scleral thickness, which might be the result of regional variation in the scleral bio-mechanical properties and the process of emmetropization causing asymmetric scleral growth. The presence of DSM can be associated with an increased risk of complications. The clinical spectrum ranges from being asymptomatic to metamorphopsia and mild-to-moderate gradual visual loss over years. Visual impairment in DSM results from retinal pigment epithelial changes, sub-foveal serous detachment, retinoschisis and myopic choroidal neovascularization. In this review, we compile and review the available information on the pathophysiology, nomenclature, classification, clinical features including imaging, differential diagnosis, complications associated with DSM and the gaps in our understanding of this entity thus far.

Introduction

The prevalence of myopia and high myopia is increasing steadily with considerable variations world-wide. High myopia is now one of the leading causes of legal blindness, especially in south-east Asian countries [1, 2]. Recently published data from 145 countries show that nearly 1460 million (22.9% of the world population) and 163 million (2.7% of the world population) people globally have myopia and high myopia, respectively [3]. It is predicted that by the year 2050, these figures will grow to nearly 4758 million (49.8% of the world population) and 938 (9.8% of the world population) million people, respectively. This is alarming as young people are affected bilaterally in the productive years of life [3].

Progressive globe elongation in high myopia leads to significant visual morbidity resulting from early cataract, glaucoma, retinal detachment and macular complications like myopic maculopathy, myopic retinoschisis, choroidal neovascularization and many more [4]. The direct and indirect consequences on visual performance, quality of life and economic burden cannot be over-emphasized.

Optical coherence tomography (OCT) allows in vivo optical biopsy of the retina. OCT has revolutionized our practice today and helps us to diagnose, follow-up and prognosticate diseases. With further advances and refinement of technology, swept-source OCT (SS-OCT) today allows us to image choroidal structure over a larger area with greater depth resolution including the sclera and orbital fat in pathological myopia [5].

Dome-shaped maculopathy is a relatively new condition associated with high myopia, which was first described by Gaucher and associates in 2008 using OCT [6]. Visual impairment in DSM results from retinal pigment epithelium (RPE) changes, sub-foveal serous detachment, retinoschisis and myopic choroidal neovascularization. Considerable progress has been made in the present understanding of this condition. In this article, we present a review of the available information on the pathophysiology, nomenclature, classification, clinical features including imaging, differential diagnosis, complications associated with DSM and our gaps in understanding of this entity so far.

Methods

A literature review was conducted in May 2020 of all English language articles in PubMed resulting from searches of the following terms: ‘dome-shaped macula’ OR

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'dome-shaped maculopathy' AND 'high myopia' OR 'degenerative myopia' OR 'pathological myopia' OR 'pathologic myopia'. Search results were combined and duplicates were removed. Clinical studies were included; however, where necessary, experimental studies were used in supporting hypothesis. We excluded, studies without abstracts, and those of other conditions (e.g. tilted disc syndrome), genetic studies, and studies with dome-shaped maculopathy secondary to conditions other than myopia. We screened a total of 65 abstracts and deemed 36 articles relevant for full publication review. We identified additional eight articles after a search of reference lists within the publications selected. This resulted in 44 articles being used to develop this review.

Historical perspective

While evaluating several myopic patients with unexplained visual loss, Gaucher and associates found an unusual feature —an anterior convex protrusion of the macula towards the vitreous cavity on OCT, which they termed as 'dome-shaped maculopathy' [6]. They retrospectively reviewed OCT images of 140 myopic eyes and found that 15 of these had similar findings. All the eyes had a Curtin type 1 or 2 staphyloma with a mean refractive error of −8.25 D [7]. Of these, ten eyes had sub-retinal fluid (SRF) at the fovea.

The initial definition of DSM was qualitative in nature, thus subjected to observational bias and heterogenicity among studies. Ohsugi et al. and Ellabban et al. defined DSM as the presence of an inward bulge of the macular RPE of >50 µm in the vertical or horizontal section of the OCT image (Fig. 1) [8, 9].

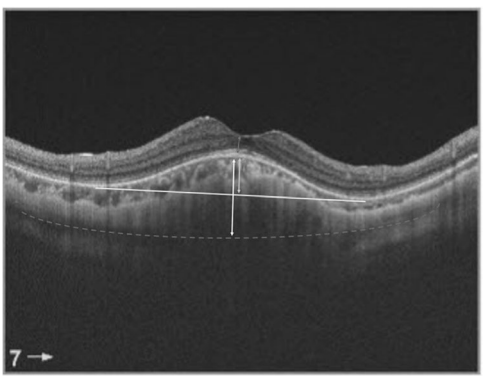

Fig. 1 A multi-averaged vertical section OCT through the fovea obtained with swept-source OCT machine in a high myope with dome-shaped macular configuration with the parameters associated. At the fovea, three parameters are measured: retinal thickness (red arrow), choroidal thickness (green arrow) and sclero-choroidal thickness (white arrow). The height of the forward convexity of the retinal pigment epithelium (orange arrow) was measured above the tangent plane (yellow line) at the bottom of the posterior pole. Blue dashed line indicates approximated outer scleral border (colour figure online).

Hypotheses on formation of DSM

In the initial description, Gaucher et al. could not ascertain the cause largely since imaging beyond the RPE was not possible with the OCT machines available then [6]. They proposed that either a localized resistance of the sclera to the staphylomatous deformation or a localized thickening of the choroid might be the possible underlying cause.

Mehdizadeh and Nowroozzadeh in their letter to the editor in response to the published article by Gaucher et al. hypothesized that in high myopia there is progressive thinning of the sclera with reduced rigidity [10]. They further hypothesised that within the staphyloma because of its smaller radius of curvature, there is a localized hypotony (Laplace's law) at the macula, which causes the macula and choroid to bulge, on the lines of hypotony maculopathy. However, this seems unlikely as none of the eyes had other associated signs of hypotony like choroidal/retinal folds and disc oedema. They also hypothesized that inside the staphyloma the vitreous tractional forces become more perpendicular resulting in this inward convex deformation. However, on OCT there is no evidence of vitreous traction to support this hypothesis.

Imanura et al. in 2011, studied the choroid and sclera using enhanced depth imaging OCT in highly myopic eyes [11]. They found that the sub-foveal scleral thickness in eyes with posterior staphyloma and DSM was significantly greater than eyes with posterior staphyloma but no DSM, 570 ± 221 microns versus 281 ± 85 micron [11]. Ellabban et al. found that increased scleral thickness was localized to the sub-foveal region [9]. However, significant staphylomatous scleral thinning was noted in all four para-foveal quadrants similar to high myopic eyes without DSM (sub-foveal scleral thickness versus para-foveal scleral thickness: 518.6 ± 97.6 versus 277.2–360.3 microns; $p < 0.001$). More interestingly, asymmetry was even present among the quadrants, with nasal being significantly thicker than the superior and inferior.

Longitudinal studies by Ellabban et al. measuring the changes in the scleral thickness over time in DSM eyes further confirmed the concept of asymmetric scleral thinning [12]. Over a follow-up period of 24.8 months, the mean change in the scleral thickness at fovea, superior, inferior, temporal and nasal para-foveal region was 5.6, 11.1, 12.1, 10.4 and 5.8 microns, respectively.

Although Imanura et al. and Chebil et al. did observe that the choroid in the DSM group was thicker than the no DSM group, the difference (~10–22 microns) was small and could not account for this anatomical change, therefore refuting choroidal thickness as the cause [9, 13]. Moreover, there are conflicting reports on comparison of choroidal thickness in eyes with DSM versus those without DSM [14]. SS-OCT confirmed that there was no inversion or collapse of the

scleral wall, and both the inner and outer sclera maintained a concavity throughout with a localized thickness at the macula [9].

The possible hypothesis to the regional variation of the scleral thickness is the asymmetric growth of the eyeball [15]. Experiments from various animal models lend support to this hypothesis that the growth of fovea and periphery is locally modulated in the process of adaptive emmetropization [16]. The peculiar shape of the eyeball in high myopia makes the periphery relatively hypermetropic. It is possible that during near work, although the image at the central macula is maintained in a relatively high degree of sharpness, the periphery areas remain largely defocussed. It results in asymmetric growth pattern where the periphery expands disproportionately as compared to the fovea. A more complex interplay between genetics and environmental factors, however, could only explain why DSM is found only a small portion of high myopic eyes. Another contributing factor could also be a regional difference in the structure and organization of the scleral lamellae, which makes the central part of the sclera mechanically stronger than the surrounding region.

An interesting observation by Fang et al. was the presence of macular Bruch's membrane defect being significantly associated with DSM in high myopia ($p < 0.001$; odds ratio: 1.96 after adjusting for longer axial length) [17]. The focal relaxation of the posterior sclera by the deficient Bruch's membrane could allow partial bulge of the macula inwards even when the eyeball is expanding out.

Incidence

To date, there are no data on the incidence of DSM from population-based studies. Hospital-based data estimate the prevalence to be 10.7–21% in eyes with high myopia [6, 14, 18, 19]. The large variation in the reported rate results from the small sample size, definition of high myopia, inclusion criteria (only symptomatic case versus all high myopia), diagnostic modality (spectral-domain versus enhance depth/swept source), scan length and scan protocols (6 versus 9 mm/12 mm) and others.

Natural history

The prevalence of DSM reported across the various categories of myopic maculopathy according to the international photographic classification is: C0 (no changes) 1.7%; C1 (tessellated fundus): 3.34%; C2 (diffuse chorio-retinal atrophy): 21.37%; C3 (focal chorio-retinal atrophy): 6.53% and C4 (macular atrophy): 6.59% [14].

Over time, asymmetric progressive elongation of the globe and scleral thinning results in the increase in the bulge height. Ellabban et al. found that over a mean follow up of 24.8 ± 2.5 months, the bulge height increases from 136.5 ± 60.9 to 157.6 ± 67 microns with a generalized choroidal thinning from 28.3 ± 17.2 to 22.9 ± 17.2 microns [12]. There was no hypermetropic shift noted suggesting that the increase in DSM height results from the deepening of the surrounding staphyloma.

Similarly, Soudier et al. over a follow-up period of 37.89 ± 33.04 months found that the dome height increased from 338 to 364 microns [20]. Interestingly, they also observed that a case of horizontal oval dome at baseline changed to a round dome during follow-up, suggesting the progressive asymmetric expansion of the staphyloma.

Although DSM is an adaptation of the macula to preserve vision despite progressive axial elongation, an increase in bulge height and choroidal thinning with age results in photo-receptors and RPE-Buch's membrane-choriocapillary complex damage contributing to visual loss.

Symptomatology

People with DSM have a history of high myopia. The symptomatology is not so much due to the presence of DSM but due to secondary changes in the overlying fovea and perifoveal retina. Thus, the clinical spectrum of presentation varies from being asymptomatic to metamorphopsia and gradual mild-to-moderate visual loss over a few years. Rarely, DSM can cause acute symptoms due to growth of a choroidal neo-vascular membrane.

Fundus features

Clinical clues for the diagnosis of DSM

Although DSM can only be confirmed on OCT, Liang et al. described some clinical clues on fundus evaluation in high myopic eyes with posterior staphyloma to enable suspecting the presence of DSM [14]. They reported the highest sensitivity with the presence of a horizontal ridge connecting the disc and fovea in 91.4% eyes. Hyperpigmentation at the macula and horizontally oval disc were seen only in 4.9% and 3.5% eyes, respectively. On stereoscopic view, a horizontal ridge consists of two parallel minimally elevated linear protrusions towards the vitreous cavity. On auto-fluorescent and infra-red imaging, the ridge is hyper-autofluorescent and hyper-reflectant, suggesting an increased reflex from the protruded sclera. On OCT cross-sectional imaging, the ridge appears as two circular hypo-reflective structures within the scleral stroma with a notch in between. Fajardo et al. in their small case series found an oblique insertion of the optic disc as the most common clinical clue and the horizontal ridge was noted only in eyes with dome height more than 400 microns [21].

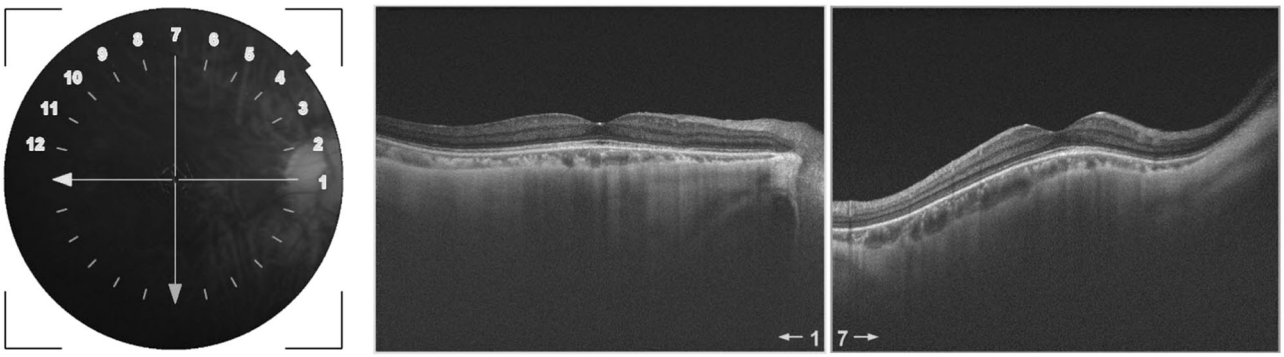

Fig. 2 Optical coherence tomography scan of horizontal and vertical sections through the fovea in a high myope using swept-source technology. The left images show the posterior pole fundus photographs with tessellated background. The middle image representing the horizontal section shows a relatively flat macular contour. The right image representing the vertical section shows a convex forward bowing of the foveal contour with normal retinal anatomy overlying it.

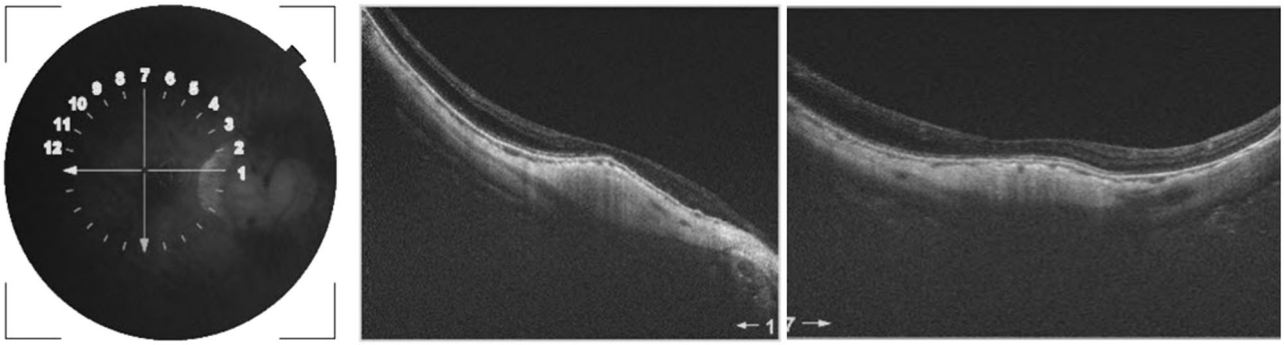

Fig. 3 Optical coherence tomography scan of horizontal and vertical sections through the fovea in a high myope using swept-source technology. The left images show the posterior pole fundus photographs with tessellated background with peri-papillary atrophy and retinal pigment epithelial changes encroaching fovea. The transfoveal horizontal line scan in the middle image shows a convex forward bowing of the foveal contour. The corresponding vertical line scan on the right hand side shows a relatively flat macular contour. The overlying retina looks grossly normal with mild irregularity of the photo-receptors.

Morphology of DSM

According to the orientation of the protrusion, three anatomical types of DSM have been described.

(1) Horizontally oriented dome: This is the most common type. The vertical OCT shows the anterior protrusion of the macula in the background of outward bowing in the peri-macular region. The horizontal OCT shows a flat contour of RPE and choroid (Fig. 2).

(2) Round-shaped dome: Second most common. Both the vertical and horizontal OCT shows an anterior protrusion of the macula in the background of outward bowing in the peri-macular region (Fig. 3).

(3) Vertically oriented dome: Least common type. The horizontal OCT shows the anterior protrusion of macula in the background of outward bowing in the peri-macular region. The vertical OCT shows a flat contour of RPE and choroid. (Fig. 4).

Liang et al. in their large series found horizontal variety to be the commonest (77%) followed by round-shaped (20%) and vertical variety (2%) [14]. Similar results were found by Hocaoglu et al.—horizontal (71%), round (27%) and vertical dome (2%) [22]. Ellaban et al. reported that the central scleral thickness in the round-shaped dome (mean thickness 598 microns) was greater than the horizontally oriented dome (mean thickness 503 microns) [9]. This was later validated by Ohsugi et al.; however, no difference in the rate of complications was noted across the three groups was noted [8].

A longer scan of 9-mm or preferably 12-mm length with both vertical and horizontal sections at the fovea is essential in the diagnosis of this condition. Although a radial scan protocol may yield more information and has been advocated by Christenbury et al., an obvious advantage over is doubtful [23].

Another less popular way of classification of DSM is based on dome height; low (50–350 μ), medium (351–650 μ) and high (>650 μ) [21]. Eyes in the medium and high group had more complications as compared to those in the low group.

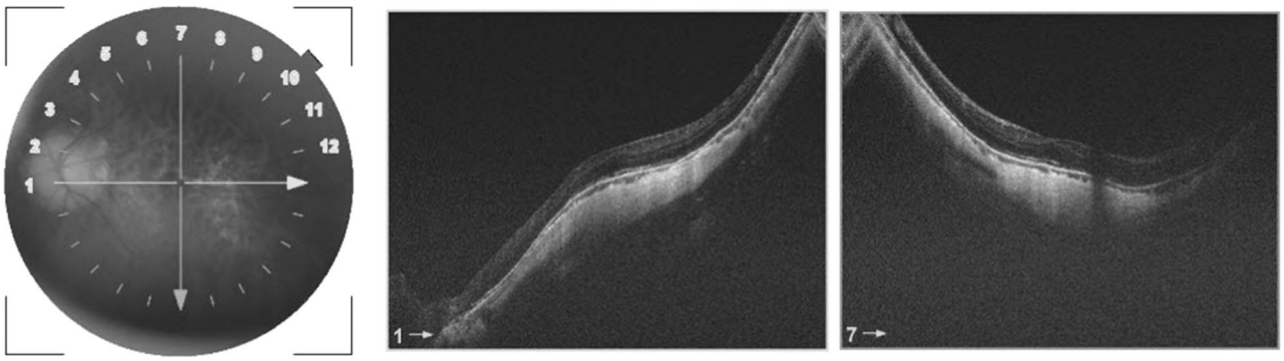

Fig. 4 Optical coherence tomography scan of horizontal and vertical sections through the fovea in a high myope using swept-source technology. The left images show the posterior pole fundus photographs with tessellated background with peri-papillary atrophy encroaching fovea. The middle image representing the horizontal section shows a convex forward bowing of the foveal contour. The right image representing the vertical section shows a shallow forward convexity of the macular. The overlying retina looks thinned out.

Macular changes

The presence of DSM is associated with an increased risk of complications. Eyes with complications have thinner choroid, more scleral thickness and higher dome height [8].

Similar prevalence of complications has been noted in high myopic eyes with inferior staphyloma and a 'macular bent', implying that the observed complications are the result of a similar anatomical configuration [24]. DSM increases the risk for shallow sub-foveal serous fluid and extra-foveal retinoschisis [18]. The incidence of CNVM has been found to be independent of the presence of DSM. An odds ratio for any complications in DSM group was 3.4 as compared to the group without DSM [14]. The OR for sub-foveal retinal detachment, foveal retinoschisis, extra-foveal retinoschisis, macular hole and CNVM calculated was 35.5, 0.06, 1.6, 0.8 and 0.8, respectively [14].

The presence of DSM is associated with a lower incidence of foveal retinoschisis, that can be explained by the protective 'macular buckle' effect at the fovea. However, this also translates to an increase in the incidence of extra-foveal retinoschisis, which is commonly noted on the slopes of the bulge with a preferential affection of the superior quadrant [19]. A higher bulge height versus width ratio predisposes to retinoschisis [19]. No association has been found between the macula hole and DSM.

Serous retinal detachment without associated CNVM

The presence of SRF in a sub-set of patients with DSM was described even in the initial report by Gaucher et al. [6]. The incidence of SRF in DSM is highly variable and ranged from 66.7 to 1.8% [6, 8, 14, 25, 26].

Multiple hypotheses for SRF generation in DSM have been postulated; however, the exact pathogenesis is presently unknown. A thickened choroid at the dome has been attributed to SRF formation on the same line as in central serous chorioretinopathy (CSCR). However, the relatively localized choroid thickening in eyes with DSM and SRF is nearly nine times smaller as compared to generalized thickening observed in CSCR [11, 25, 27]. No hyper-permeability has been noted on ICG in DSM eyes [26]. However, an abrupt change in the scleral thickness at macula could be contributory [28]. The thickened sclera has been proposed to impair the flow of choroidal fluid, similar to cases of nanophthalmos. However, it seems unlikely that that a localized scleral thickness at fovea would impair the flow of choroidal fluid at the equatorial region thorough vortex veins [8].

The localized bulging at macula would result in significant mechanical stress to the RPE, which results in anatomical and functional damage [8]. The macular pigment atrophy correlated with the bulge height and enlarged over time [20]. There may also be some compressive effect of the thickened sclera on overlying choroid and RPE with resultant damage [29].

Mateo and Bures described three patients with a macular buckle for myopic foveoschisis who on follow up developed a serous retinal detachment [30]. They postulated that an iatrogenic 'dome-shaped configuration' caused compression at the border to reduce the choroidal flow at the foveal area increasing the choroidal thickness and SRF accumulation [30]. This is supported by the observation of greater scleral thickness and bulge height in eyes with DSM and SRF as compared to eyes with DSM but no SRF [22, 27, 31].

Most commonly, patients complain of gradual onset decrease in vision/metamorphopsia, although few may be asymptomatic. It is at times difficult to suspect SRF in these eyes with high myopia. OCT helps detect more than what is seen clinically and can reveal forward convex protrusion of the macula, shallow SRF, photo-receptor layer disruption or

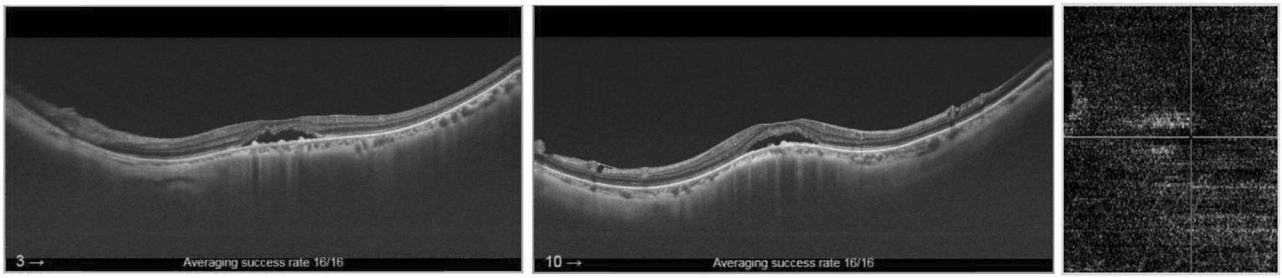

Fig. 5 (Left) Optical coherence tomography scan of vertical transfoveal line scan showing a convex forward protrusion of the central macula, a shallow sub-foveal neurosensory detachment at the apex of the dome. (Middle) Magnified image of another line scan showing retinal pigment epithelial irregularity, choroidal thinning and elongation of the photo-receptor outer segments overlying it. (Right) OCT-angiogram of the outer retinal layer slab confirms the absence of Choroidal neo-vascular membrane.

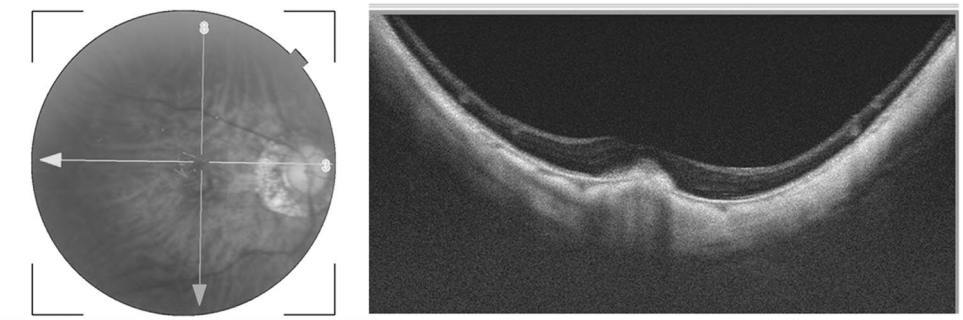

Fig. 6 (Left) Fundus photograph of an eye with high myopia showing peri-papillary atrophy, tessellations and a small dirty grey membrane with a speck of sub-retinal haemorrhage noted at fovea. (Right) Optical coherence tomogram, vertical transfoveal line scan showing grossly thinned choroid and forward protrusion of the sclera, choroid and retinal pigment epithelium at the fovea. An ill-defined hyper-reflective material was noted in the sub-retinal space at the fovea, suggestive of choroidal neo-vascular membrane.

a flat irregular pigment epithelial detachment overlying the dome (Fig. 5).

The presence of SRF is associated with window defect and pin-point leakage on FFA and hyper-cyanescence spots on ICG [26]. Ruling out associated CNVM is necessary in cases with associated PED as the treatment and prognosis would differ significantly.

The natural course of SRF is fluctuating. However the vision remains relatively preserved [20, 26, 32, 33]. Over a follow-up of 20 months with no intervention, few eyes showed a decrease/resolution of SRF while others showed an increase or appearance of new SRF [26]. The mean SRF increased by a mean of 24 ± 12 microns with no change in mean BCVA [26]. Shallow SRF may not be detrimental to the diffusion of nutrients and metabolic products to the overlying RPE and photo-receptors [34].

Various treatment modalities have been tried in the same lines of CSCR. It includes photodynamic therapy, laser photo-coagulation, anti-VEGFs, mineralocorticoid receptor antagonists and intravitreal steroid injections [23, 35–42]. Although these case reports and small case series did find a modest reduction in SRF in some of the patients, this did not transform to visual improvement in most [23, 35–42]. Poor outcome is secondary to the presence of RPE atrophy. At present, there is little evidence to support the effectiveness of these treatment modalities given the relatively benign course of the disease.

CNVM

The development of CNVM is independent of the presence of DSM and is associated with increasing age, higher axial length and thinning of choroid [14]. Liang et al. reported that 21.3% of high myopic eyes with DSM eyes had CNVM, which was similar to myopic eyes without DSM (27.4%) [14]. Similarly, of the total 277 eyes included in RADIANCE study, 50 eyes (18%) had DSM [43].

Patients would generally present with sudden onset decrease in vision, metamorphopsia, recent onset floaters and/or scotoma. Clinically, a greyish membrane may be noted with a shallow SRF with speck of haemorrhage. These findings may be very subtle to be missed even after a meticulous clinical examination.

An OCT through the lesion would show a sub-retinal hyper-reflective material (Type 1 CNVM) with minimal SRF. Rarely, intra-retinal fluid or significant SRF are noted (Fig. 6). On FFA, the lesion is hyper-fluorescent in

Table 1 List and frequencies of various complications associated with dome-shaped maculopathy in the published literature.

Study (no. of eyes)	RPE atrophy	SRF without CNVM	CNVM	Foveal retinoschisis	Extra-foveal retinoschisis	FTMH	Lamellar MH	No complications	Others
Cocco et al. [24] (48 eyes)	5/48	4/48	10/48	9/48[a]		2[b]		18/48	
Ohsugi et al. [8] (49 eyes)	–	8/49	6/49	–	–	–	–	35/49	
Gallego-Pinazo et al. [27] (52 eyes)	–	17/52	13/52	2/52	5/52	–	1/52	14/52	
Liang et al. [14] (225 eyes)		4/225	48/225	2/225	62/225	7/225	8/225	59/225	
Soudier et al. [20] (29 eyes)	20/29	15/29	15/29[c]					5/29	
Zhao et al. [18] (149 eyes)	–	3/149	3/149	10/149	23/149	2/149	2/149		14 eyes ERM;1 eye haemorrhage
Hocaoglu et al. [22] (167 eyes)	–	13 /167 (7.8%)	62/167 (37%)	15/167 (9%)[d]		4/167 (2.4%)	5/167 (3%)	62/167 (37%)	7 eye ERM

CNVM choroidal neo-vascular membrane, *ERM* epiretinal membrane, *FTMH* full thickness macular hole, *MH* macular hole, *No.* number, *RPE* retinal pigment epithelium.

[a]Value represents Internal limiting membrane detachment leading to retinoschisis.

[b]Separate information of lamellar and full thickness macular hole.

[c]Value represents other complications except sub-retinal fluid and macular atrophy. Separate values not available.

[d]Denotes combined foveal and extra-foveal retinoschisis.

the early phase, which increase with time with minimal leakage.

Intravitreal anti-VEGF drugs are the standard of care for myopic CNVM even in eyes with DSM. A post hoc analysis of RADIANCE trial was done to evaluate if presence of DSM affected the response to ranibizumab injection in myopic CNVM [43]. The authors found that eyes with DSM and myopic CNVM ($n = 38$; 5012 letters) had a worse baseline visual acuity as compared to eyes with no DSM and myopic CNVM ($n = 183$; 5713 letters) [43]. However, no substantial difference in the visual gain between the two groups was noted at 3 months (with DSM 11.19 letters versus no DSM 12.09 letters) and 12 months (with DSM 12.11 letters versus no DSM 14.01 letters) [43]. The mean number of injections was similar in both groups (with DSM 3.9 versus no DSM 3.7 injections) [43] Similar results were replicated in other studies, thus validating the fact that DSM does not alter anti-VEGF response in myopic CNVM [44, 45]. Interestingly, it was noted that eyes with DSM fared better after PDT than eyes without DSM, which could be related to the thicker choroid in eyes with DSM [43].

In a rare case report by Naysan et al, polypoidal choroidal vasculopathy was reported in an eye with DSM. OCT and ICG showed the presence of thumb like polyps and focal 'hotspots', respectively, which corresponded to polys on OCT-angiography and En-Face OCT [46].

Differential diagnosis

(1) Choroidal haemangioma: A localized bulge with SRF can mimic choroidal haemangioma [47]. OCT-EDI or SS-OCT would be sufficient in most cases, to aid the diagnosis —revealing the choroidal thickening in choroidal haemangioma [47]. Fluorescein angiography showed early diffuse hyper-fluorescence with leakage in choroidal haemangioma while there is occasional staining from SRF in cases of DSM [47]. On ICG, DSM is iso-fluorescent, while haemangioma shows a bright hyper-fluorescence with characteristic 'washout' phenomenon [47].

(2) Inferior staphyloma: Coco et al. reported that in myopic eyes with inferior staphyloma, the superior edge of the staphyloma at fovea may cause a sudden sharp slope resulting in a similar macular 'bend' on OCT [24]. They found similar complication rates between the two groups, thus suggesting that these changes could be secondary to anatomical distortion [24].

Ridge in young myopic patients

A similar convex elevation of the macula termed 'ridge-shaped macula' has been noted in young myopic individuals even as young as 4 years [48]. In contrast, DSM has been typically described in patients above 20 years with a mean age ranging between 50 and 60 years [6, 8–10].

None of eyes with 'ridge-shaped macula' have an associated posterior staphyloma and macular Bruch membrane defect [48]. Moreover, these eyes are less myopic with smaller axial length, better visual acuity and lower grade of myopic maculopathy [48]. Interestingly, the dome height was significantly lower (124 ± 123 versus 206 ± 114 mm, $P = 0.036$) with a broader base (4413 ± 1085 versus 2956 ± 709 mm, $P = 0.001$) [48].

It is not known at present whether these patients will develop staphyloma in future and convert to DSMs. Ridge-shaped maculas may result from the folding of Bruch membrane at the posterior pole resulting from an asymmetrical enlargement of Bruch membrane in the equatorial region [48].

The gaps in our understanding and future directions

Further research is needed to understand why only certain proportion of eyes with myopia develop DSM. The natural course of SRF and efficacy of various treatment modalities in DSM needs to be validated in large longitudinal studies.

The terms 'dome-shaped macula' and 'dome-shaped maculopathy' at present are used inter-changeably. There is a sub-set of eyes that show this configuration without reduction in vision and changes in overlying retina as shown in a sub-set of eyes mentioned in Table 1. Probably they could just be termed as dome-shaped macula while those with overlying secondary changes (retinal pigment epithelial changes, irregular PEDs, sub-foveal detachment, photo-receptor layer irregularities and retinoschisis, resulting in visual loss) could be described as dome-shaped maculopathy. Serial long-term longitudinal follow up of macula of eyes with high myopia could probably through light on the temporal relationship of maculopathy developing over a dome-shaped macula and would further strengthen the above nomenclature. Long-term follow-up and documentation of macular changes of children of parents with high myopia will give us an opportunity to build up the story on how the changes evolve over time. This could probably add to our existing knowledge on the pathogenesis of this entity so far.

Conclusion

DSM is an anterior convex protrusion of the macula in a small sub-set of eyes with high myopia. This seems to be related to a localized scleral thickness, which might be the result of regional variation in the scleral bio-mechanical properties and process of emteropization causing asymmetric scleral growth.

Longer OCT scan, both vertical and horizontal, is essential for the diagnosis of DSM. The bulge height increases minimally with time as a result of progressive staphyloma elongation. SRF and extra-foveal retinoschisis are commonly associated complications. CNMV in DSM responds favourably with anti-VEGF injections.

Summary

What is known about this topic

- Dome Shaped Macula is a forward convex protrusion of the macula detected on OCT.
- It results from asymmetric scleral growth in a small subset of eyes with high myopia.
- SRF at the dome and extra-foveal macular schisis are well known complications assocaited with DSM.

What this study add

- Comprehensive review of present literature on the pathophysiology, classification, clinical features and complications associted with DSM.
- Lacunae in our present understanding are described.
- The need for future large longitudinal studies to understand the natural history and treatment of DSM and its associated complications.

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Author contributions MJ, TRP and LG conceived and designed the review. MJ was involved in literature search and manuscript writing. TRP and LG were involved in critical review. LG was fundamental in overall supervision and feedback. All authors were critical in manuscript writing and improvement of present work

Compliance with ethical standards

Conflict of interest The authors declare no competing interests.

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