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Progressive development of dome-shaped macula and subsequent onset of punctate inner choroidopathy in a highly myopic patient: a 7-year follow-up case report

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ABSTRACT

Purpose: To describe the development of dome-shaped macula (DSM) in a highly myopic patient, followed by the subsequent onset of punctate inner choroidopathy (PIC).

Observations: A 46-year-old man with high myopia, with a systemic history of psoriatic arthritis and an ocular history of Acanthamoeba keratitis resulted in two penetrating keratoplasties (PK) in his right eye, was followed up regularly at our center for a 7-year period. During the follow-up, the left eye remained stable until the fifth year. However, at the seventh year, a routine fundus examination revealed the asymptomatic appearance of lacquer cracks, which had been absent at baseline. Multimodal imaging evaluation further identified the incidental development of a horizontal oriented DSM and active and inactive PIC foci localized near the newly detected lacquer cracks.

Conclusions and importance: This case report provides new insights into the formation of DSM, supporting the hypothesis that macular bulge may result from differential elongation of the peri-dome region. Additionally, it highlights that DSM can coexist with PIC.

1. Introduction

Axial myopia is associated with a wide range of changes in the posterior segment of the eye. These include choroidal and scleral thinning, patchy macular atrophies with defects in the retinal pigment epithelium (RPE) and Bruch's membrane (BM), temporal shifting and widening of the BM opening at the optic nerve head, and elongation and thinning of the lamina cribrosa and peripapillary structures.[1,2]

Dome-shaped macula (DSM) is another notable feature observed in some highly myopic eyes. First described by Gaucher in 2008, DSM is characterized by a convex macular profile on optical coherence tomography (OCT).[3] Its formation is thought to involve focal thickening at the subfoveal level or differential elongation of the peri-dome region, causing an inward scleral shift in the macular area. Histological studies have suggested a role for Bruch's membrane (BM) biomechanics, as DSM is often associated with central BM defects, relative scleral thickening, and choroidal changes.[4]

While DSM has been extensively characterized on multimodal imaging, its initial development and progression have not been well-documented in the literature. Complications such as subretinal fluid (SRF), choroidal neovascularization (CNV), and vitreoretinal interface abnormalities are common in DSM,[5,6] but its association with inflammatory conditions like punctate inner choroidopathy (PIC) has not been previously reported.

This case report documents the dynamic development of DSM, absent at baseline, and its subsequent association with PIC in a highly myopic patient.

2. Case report

A 46-year-old man with high myopia (axial length: 28.53 mm in the right eye and 28.14 mm in the left eye), and a history of amblyopia in the right eye was followed at our center for 7 years. His medical history included psoriatic arthritis, diagnosed in 2020 and managed with a monoclonal antibody targeting interleukin-23.

At presentation, his best-corrected visual acuity (BCVA) was 20/50

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in the right eye and 20/20 in the left eye. Anterior segment examination revealed sequelae of penetrating keratoplasties (PK) from previous infectious keratitis, and pseudophakia in the right eye, while the left eye was unremarkable. Fundus examination showed a tessellated fundus and peripapillary atrophy in both eyes.

OCT of the right eye showed a normal foveal contour. In contrast, the left eye exhibited a DSM characterized by an inward central macular bulge of the RPE exceeding 50 μm.[7] No lacquer cracks were detected in either eye.

At the seven-year follow-up examination, serial OCT scans of the right eye revealed the development of a central subfoveal scleral bulge, meeting DSM criteria (Fig. 1). According to Caillaux's classification,[8] the DSM displayed a horizontal dome configuration, with significant choroidal thinning.

Comparison of vertical OCT scans over time suggested that this structure formed due to a progressive inferior displacement of the macula, leading to the emergence of a horizontal ridge. The sclero-choroidal margin beneath the dome appeared curved, with a more pronounced profile in the vertical scan corresponding to the inferior macula. No perforating vessels were observed in or around the dome. During the same examination, additional pathological changes were detected. Linear lacquer cracks were present in the inferior half of the posterior pole—absent in all previous visits—and oriented parallel to the DSM.

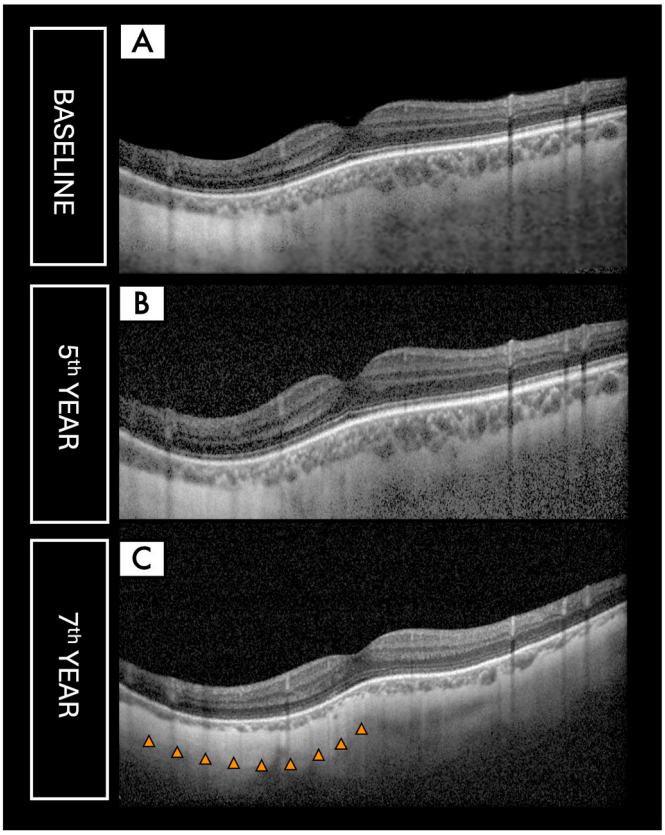

Fig. 1. Longitudinal development of DSM - (A) Baseline OCT vertical scan showing no evidence of DSM. The sclerochoroidal margin beneath the dome appears linear, with preserved choroidal thickness. **(B)** OCT scan after 5 years shows an unchanged appearance, and no evidence of DSM formation. **(C)** OCT scan at 7 years reveals the development of a central bulge characteristic of DSM. The sclerochoroidal margin beneath the dome no longer appears linear but rather curved (orange arrowheads), exhibiting increased concavity inferior to the macula. In addition, choroidal thickness shows a gradual reduction in subfoveal and peri-dome choroidal thickness. Defects in Bruch's membrane are observed at the posterior pole, in addition to the active foci of PIC. Infrared fundus images corresponding to each OCT scan are shown on the right, with orange arrows indicating the exact location of the B-scan within the macula. (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

Three months after the seven-year visit, new yellow-gray punctate lesions emerged in the posterior pole, predominantly located inferior to the fovea, extending nasally and temporally along the lacquer cracks (Fig. 1). OCT imaging of these lesions revealed focal hyperreflective disruptions of the RPE–Bruch's membrane complex, accumulation of subretinal hyperreflective material, localized choroidal thickening, disruption of the ellipsoid zone, and posterior choroidal hypertransmission (Fig. 2), consistent with active foci of PIC. Some lesions demonstrated choroidal thinning and RPE patchy atrophy, attributable to inactive PIC.

3. Discussion

This case report highlights the imaging findings associated with the onset of DSM and its subsequent coexistence with PIC, providing novel insights into DSM development and its potential link with inflammatory chorioretinal conditions.

The pathogenesis of DSM remains unclear, though it is hypothesized to involve asymmetric scleral thinning and uneven ocular expansion during myopia progression. Experimental models of myopia have demonstrated disproportionate peripheral retinal expansion relative to the fovea.[9] Dormegny[10] proposed that DSM results from differential elongation of the peri-dome region, leading to localized inward deformation of the sclera. In our case, serial imaging revealed a progressive subfoveal posterior bulge in the right eye, accompanied by changes in scleral curvature. Notably, the sclero-choroidal margin evolved from a linear to a curved configuration, particularly pronounced inferiorly on vertical scans. These observations align with Dormegny's hypothesis,[10] suggesting that DSM formation may result from lateral structural weakening and deformation rather than solely central subfoveal thickening.

BM breaks have been reported with high prevalence in patients with DSM.[11] It has been hypothesized that BM breaks may promote scleral relaxation, thereby favoring an inward bulge.[4] Several studies have indicated that the spatial distribution of these defects appears to be associated with the presence of a nearby scleral outpouching.[1,4] In our case, the de novo appearance of Bruch's membrane breaks predominantly in the inferior macular region is accompanied by localized scleral weakening that could promote the scleral bulge. Systemic factors, such as the patient's history of an inflammatory condition, such as psoriatic arthritis, may have further influenced the scleral biomechanics, complicating the pathogenesis of DSM-specific changes. Psoriatic arthritis, managed with interleukin-23 inhibitors in this case, could have indirectly contributed to scleral remodeling, underscoring the need for further research on systemic factors in DSM pathophysiology.

Longitudinal follow-up allowed evaluating the rate of DSM formation in our patient. Prior studies, such as those by Ohno-Matsui, reported an average increase in DSM height of approximately 30 μm over 10 years in pre-existing cases.[12] However, data on DSM development from baseline are limited. In this patient, DSM developed within a two-year interval, illustrating that DSM formation can occur rapidly in highly myopic eyes, potentially triggered by dynamic biomechanical changes.

This case highlights an underrecognized association between DSM and PIC. Myopia is a recognized risk factor for PIC, as axial elongation induces alterations in the sclera, choroid, and retina that predispose to inflammatory lesions.[6] In particular, it is plausible that posterior scleral thinning, choroidal thinning, increased fragility of Bruch's membrane, and defects in the RPE may enhance immune trafficking, eliciting a local immune response that subsequently results in the punch-out lesions visible in the fundus. While no causal relationship between DSM and PIC has been established, we may hypothesize that the rapid onset of DSM predisposes the eye to further pathological changes that exacerbate the structural and microvascular alterations already described in myopic

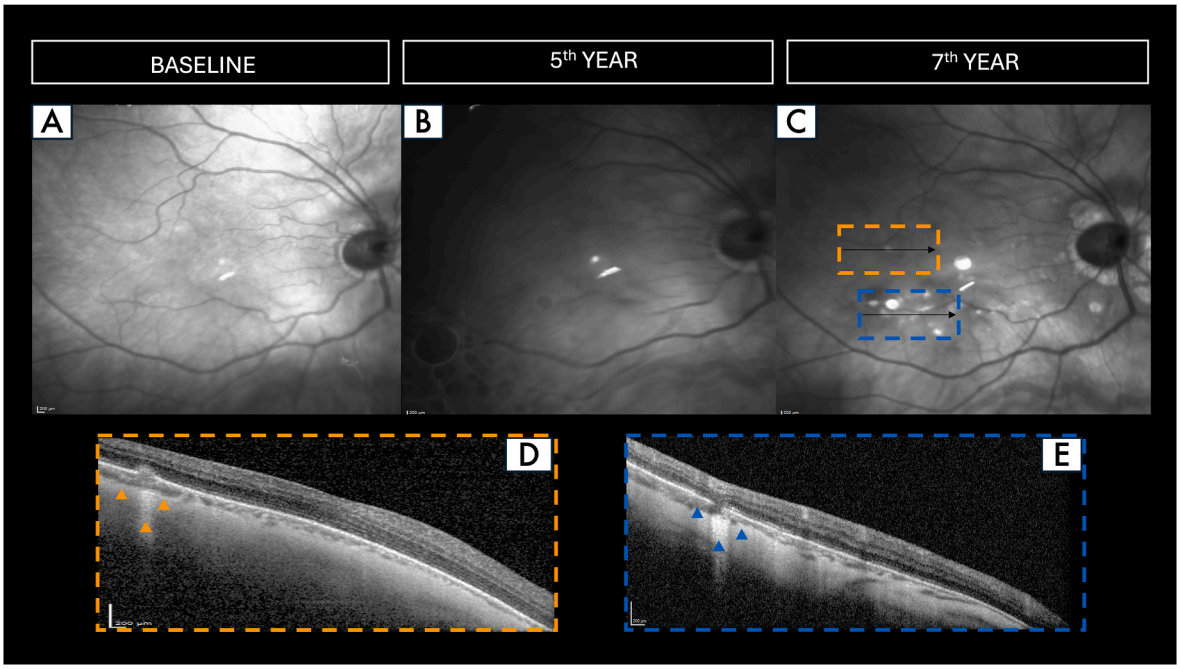

Fig. 2. Progressive onset of punched-out chorioretinal lesions on en-face infrared imaging and OCT features of PIC foci - Panels (**A**), (**B**), and (**C**), en-face infrared images illustrate the progressive appearance of new chorioretinal lesions at the posterior pole, characterized by a punched-out pattern with a linear arrangement in the inferior region of the posterior pole extending temporally. Additionally, infrared imaging shows a progressive expansion of peripapillary atrophy over time. Panels (**D**) and (**E**) show linear scans of two distinct PIC foci at different stages of evolution. Panel (**D**) displays an active lesion (orange arrowheads) with the presence of hyperreflective material, splitting of the Bruch's membrane/RPE complex, choroidal hypertransmission, and choroidal thickening. Conversely, panel (**E**) illustrates an inactive PIC foci (blue arrowheads), showing Bruch's membrane/RPE disruption and the initial collapse of the outer retina. (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

eyes, thereby favoring the development of PIC. Moreover, the spatial distribution of the PIC lesions closely mirrored the linear arrangement of the Bruch's membrane breaks, which were aligned parallel to the orientation of the dome.

This case report has limitations. First, the presentation of a single case does not allow for generalization of these findings. Second, we cannot establish a causal relationship between the development of DSM and the presence of Bruch's membrane ruptures, as both alterations appeared de novo at the final visit. Moreover, the patient was not examined between the five- and seven-year follow-up visits; consequently, the precise timing of DSM and lacquer crack onset cannot be determined. As a result, the actual interval for the development of PIC may also be broader than observed, since these lesions were first detected 3 months after the seven-year visit but could have been initiated earlier in association with the preceding structural changes. Furthermore, inflammatory conditions—including PIC—can weaken the sclera and choroid, so a causal link between DSM and PIC cannot be excluded in this instance. Finally, the patient's positive history for autoimmune disorders and the treatments administered cannot be ruled out as contributing factors in the genesis of both DSM and PIC. Additionally, it was not possible to measure scleral thickness, thereby preventing an assessment of its variations over time.

In conclusion, this case highlights the dynamic development of DSM and its novel association with inflammatory choroidopathy in a highly myopic patient. The findings suggest that DSM formation may result from differential scleral elongation and structural deformation. The coexistence of DSM and PIC underscores the need for vigilant multimodal imaging follow-up in highly myopic patients, as early detection of DSM and associated inflammatory or structural complications could guide timely management and prevent vision-threatening outcomes. Further studies are needed to explore the pathophysiological mechanisms linking DSM, PIC, and their shared risk factors.

CRediT authorship contribution statement

Sebastiano Del Fabbro: Writing – original draft, Methodology, Investigation, Formal analysis, Data curation, Conceptualization. **Alessandro Arrigo:** Validation. **Maria Vittoria Cicinelli:** Writing – review & editing, Validation, Supervision. **Francesco Bandello:** Validation, Supervision. **Maurizio Battaglia Parodi:** Visualization, Validation.

Patient consent

Each study subject provided signed informed consent.

Authorship

All authors attest that they meet the current ICMJE criteria for Authorship.

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Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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