DOME SHAPED MACULOPATHY

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DOME SHAPED MACULOPATHY-DEFINITIONS

• The entity Dome Shaped Macula ( DSM ) was first described by Gaucher and associates in 2008 by the use of time-domain optical coherence tomography (OCT) as an inward convexity or bulge of the macula within the concavity of the posterior staphyloma

  “We describe in this study an entity characterized by a convex elevation of the macula within a myopic staphyloma.”

• Only in myopic eyes and always with Posterior Staphyloma, Type I or II

• It is seen in approximately 10.7% of eyes with high myopia

SYMPTOMS

- The dome-shaped macula seems to be a new cause of visual impairment in myopic eyes

➤ **Symptoms**

- Visual loss and
- Metamorphopsia
EXAMPLES

(Top Left) Fundus photograph showing pigmentary changes present in the fovea. Arrowheads delimit the area of the staphyloma;

(Top right) Late-phase fluorescein angiography (FA) image showing a leakage point (arrow) Serous retinal detachment (arrowheads).

(Middle left) ICGA showing no sign of choroidal new vessels.

(Middle right) Late-phase ICGA image showing hypofluorescence in the area of RPE disturbance and focal hyperfluorescence (arrow) corresponding to the leakage point observed on FA image.

(Bottom left) Vertical OCT scan. This bulge affects the retina, the RPE, and the inner choroid. The foveal retina is slightly detached at the top of the macular bulge (asterisk) and above the fovea (arrow), corresponding to the serous detachment seen on FA image.

(Bottom right) Ultrasonography image confirming the abnormal anterior bulge at the posterior pole of the globe,
(B, C), OCT images across the central fovea. The sclera is thickened in the macula and shows an inward bulge in the horizontal section (B) and vertical section (C). The fovea (arrowhead in B) is located temporal to the top of the dome in the horizontal section.

(E, F), OCT image across the central fovea. The sclera is thickened in the macula and shows an inward bulge in both the horizontal (E) and vertical (F) sections. The fovea (arrowhead in E) is located slightly temporal to the top of the dome.
An inward bulge due to a DSM is observed only in the horizontal OCT section (B) and not in the vertical section (C). The central fovea (arrowhead in B) is located temporal to the top of the dome.
A staphyloma is defined as an abnormal local protrusion of thin sclera and uveal tissue and was categorized into 10 types by Curtin.

Drawings showing the classification of the different types of posterior staphyloma according to Curtin.

Because the DSM feature could not be categorized to any subtypes of staphyloma, it should be considered a novel, posterior anatomic characteristic of some highly myopic eyes and not as a staphyloma subtype.

It is believed that the dome-shaped macula is not related to the type of staphyloma, but rather is related to an anatomic or structural change within the sclera and can occur in eyes with any type of staphyloma.

Three-Dimensional Tomographic Features of Dome-Shaped Macula by Swept-Source Optical Coherence Tomography
Clues that a dome-shaped macula (DSM) might be present

A, B, Macular pigmentation (arrow) is seen funduscopically as increased pigmentation (A) and as hypoautofluorescence (B).
C-E, Horizontal ridge-like lesion connecting the optic disc and the fovea (between arrows).

1 - Macular pigmentation
2 - Ridge-like connection
3 - Oval optic disc

The horizontal ridge is observed funduscopically as a yellowish-white linear lesion (C), as hyperautofluorescent (D), and as hyperfluorescent (E).
In stereoscopic observations, the horizontal ridge seems to slightly protrude toward the vitreous.
F, G, Horizontally oval optic disc (arrowheads).
Horizontal Ridge

A, Right fundus showing 2 yellowish linear lesions connecting the optic disc and the macula horizontally (arrows).

B, Fundus autofluorescence are observed as hyperautofluorescence (arrows).

C, Infrared image shows a horizontal ridge as hyperreflectance (arrows).

D, E, OCT images. An inward bulge due to a DSM is observed only in the vertical OCT section (E) and not in the horizontal section (D).

F, G, Vertical OCT images across the horizontal ridge nasal to the central fovea. Two cross-sections of hyporeflective structures (arrowheads) are observed in the sclera (F). There is a notch-like dent (arrow) between the 2 horizontal protrusions (G).
Causes

- Several authors have suggested possible causes for this abnormal morphology, including
  - Scleral infolding similar to hypotony maculopathy,
  - Vitreous traction, and
  - Choroidal thickening.

- However, more recent evidence provided by enhanced depth imaging OCT (EDI-OCT), has shown that the usual cause is localized thickening of the sclera underneath the macula in highly myopic eyes.


The external surface of the sclera is seen clearly (white arrowheads), and the sclera shows uneven thinning in the posterior pole, being relatively thick beneath the fovea.
Complications in the Eyes With a Dome-Shaped Macula

- Choroidal neovascularization
- Serous retinal detachment
- Diffuse chorioretinal atrophy
- Patchy chorioretinal atrophy
- Lamellar macular hole
- Full-thickness macular hole
- Foveal schisis
- Extrafoveal retinal schisis
Macular abnormalities in DSM

- Foveal serous RD was the most common finding,
- Observed in almost one third of the eyes with dome-shaped macula and
- In 44% of the eyes with dome-shaped macula without myopic CNV

- The origin of subretinal fluid in dome-shaped macula eyes without choroidal neovascularization is not known, as is the therapeutic mechanism of action of verteporfin PDT in this condition
- Hypothesis - Subretinal fluid can occur in eyes because of the obstruction of outflow of choroidal fluid by a thick sclera
- The effect is similar to central serous chorioretinopathy, in which PDT is presumed to close hyperpermeable vascular channels in the choroid

Serous RD is generally described in highly myopic eyes with
- CNV
- Tractional phenomena
- Dome-Shaped Macula or
- Tilted disc syndrome
Treatment

- The only two studies reporting treatment of subretinal fluid in dome-shaped macula describe the unsuccessful use of argon laser photocoagulation.


- Anti-VEGF injections are without benefit.

  - Although very limited, it is reasonable to consider **half-fluence verteporfin PDT** as treatment for patients with nonresolving subretinal fluid associated with dome-shaped macula.
  - Application of **low-intensity laser photocoagulation**, similar to that employed in central serous retinopathy, seems a reasonable alternative for eyes with focal areas of leakage that are located safely outside the fovea, particularly if PDT has failed to resolve the fluid.
  - Although additional experience is needed to confirm the efficacy of these treatments in dome-shaped macula,
  - The risk-benefit ratio appears acceptable for patients with significant visual symptoms or disability.
RADIANCE Study

- Patients with visual impairment due to myopic CNV
- A total of 277 baseline scans have been analyzed for the presence of DSM.
- Identified a total of 50 patients (18%) with the DSM feature.

- Of the 50 patients with DSM, 38 were treated with intravitreal ranibizumab and 12 with verteporfin photodynamic therapy (vPDT) at baseline.
- Comparing visual outcome in patients treated with ranibizumab showed no differences in visual gain at month 3, in patients with DSM compared with patients without DSM.
- The number of injections at month 12 was similar in both groups.
- At month 12 follow-up, no relevant differences in visual gain between the groups were detected.
- Comparison of patients treated with vPDT having DSM or no DSM showed numerically better outcomes at month 3 for patients with DSM at baseline.
- This is an important finding, because the DSM feature is a frequent finding in patients with myopic CNV.
Case report-Treatment with PDT

Early (A) and late (B) fluorescein angiography images of the right eye show mild late leakage nasal to the fovea without definable choroidal neovascularization.

(C) OCT demonstrates DSM with subretinal fluid in the right eye and
(D) without fluid in the left eye.

(E) OCT of right eye 6 months after PDT shows resolution of subretinal fluid.
(F) Subretinal fluid has reaccumulated in the right eye 2 years after PDT.

(G) Six months after a second PDT treatment, subretinal fluid has decreased but not completely resolved.

(H) OCT imaging 3 months after grid laser shows complete resolution of subretinal fluid.
Case 2. Fundus photographs of the right (A) and left (B) eye demonstrate absence of drusen and pigment mottling.

Early (C) and late (D) fluorescein angiography images of the left eye show mild diffuse late leakage in the central macula without definable choroidal neovascularization.

OCT of the right (E) and left (F) eyes shows bilateral dome-shaped macula with subretinal fluid in the left eye.

Four months after PDT (G), the subretinal fluid has resolved completely in the left eye.
Differential Diagnosis

- Idiopathic central serous chorioretinopathy,
- Neovascular Age-related Macular Degeneration, or
- Myopic Choroidal Neovascularization
FIBROVASCULAR PED

The conventional OCT scans before (E) and 1 month after (F) PDT show complete resolution of central serous retinal detachment.

The EDI-OCT scans before (G) and after (H) PDT show that the choroid was thickened initially and decreased 1 month after PDT (H, double arrowhead).

- Choroidal vascular changes seen only inside the inward bulge within the posterior staphyloma and not widely spread in the entire choroid like CSR

Central Serous Retinopathy

Serous detachments of the neurosensory retina in the macular region.
Differential diagnosis for myopic choroidal neovascularisation (CNV):

(A and B) haemorrhage due to lacquer cracks;

(C) dome-shaped macula with serous retinal detachment; and

(D and E) macular fluid due to staphyloma.
Conclusion

• New Entity
• Symptomatic patients
• Exclude CNV
• Anti – VEGF ineffective
• PDT and Argon laser
Ευχαριστώ πολύ