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CASE REPORT

Dome-shaped macula associated with Best vitelliform macular dystrophy

Maurizio Battaglia Parodi, Ilaria Zucchiatti, Francesco Fasce, Maria Lucia Cascavilla, Maria Vittoria Cicinelli, Francesco Bandello

Department of Ophthalmology, University Vita-Salute, San Raffaele Scientific Institute, Milan - Italy

ABSTRACT

Purpose: Dome-shaped macula (DSM) has been described recently as an inward convexity of the macula typical of myopic eyes detectable on spectral-domain optical coherence tomography (SD-OCT). The authors describe a case of monolateral DSM associated with Best vitelliform macular dystrophy (VMD).
Methods: Case report.
Results: A 60-year-old man already diagnosed with VMD in vitelliruptive stage underwent SD-OCT that revealed the typical vitelliform material accumulation associated in the left eye with a convex elevation of the macula. No change was registered over a 1-year follow-up.
Conclusions: This is the first report describing a monolateral DSM associated with VMD. Dome-shaped macula could be considered as a nonspecific scleral alteration, probably due to increased scleral thickness, which can accompany many retinal disorders.

Keywords: Best vitelliform macular dystrophy, Dome-shaped macula, Optical coherence tomography

Introduction

Dome-shaped macula (DSM) was first described in association with high myopia and is characterized as an inward convexity of the macula (1, 2).

We describe a case of Best vitelliform macular dystrophy (VMD) associated with DSM.

Case report

A 60-year-old man was referred for routine examination. He had been followed for VMD for 30 years and genetic characterization confirmed mutation in the *VMD2* gene. Visual acuity was 20/31 OD and 20/40 OS. Dilated fundus examination revealed the presence of bilateral VMD with a small yellowish material centered on the fovea OD and patchy vitelliform accumulation OS. Short-wavelength fundus autofluorescence showed a focal central hypoautofluorescence OD and a patchy pattern alternating hypoautofluorescence and hyperautofluorescence OS. Electro-oculogram was extinct. Spectral-domain optical coherence tomography (Spectralis Heidelberg, Germany) showed a focal hyperreflectivity at the retinal pigment epithelium (RPE) level OD and a dome-shaped convexity OS (Fig. 1). More specifically, at the apex of the DSM, the hyperreflective vitelliform material accumulated within the subretinal space above the RPE, but below the outer segments of the photoreceptors.

The patient was followed for 1 year, revealing no change in the clinical picture.

Discussion

Recent investigations have suggested that different mechanisms may lead to DSM development, including high tangential vitreoretinal forces, increased scleral thickness, or scleral infolding secondary to insufficient pressure to maintain distension of the eye wall. More specifically, according to the most supported theory, DSM could be considered as a result of an increased scleral thickness localized under the macula.

Dome-shaped macula has been found in association not only with high myopia, but also with other conditions, including tilted disc with serous macular detachment and central serous chorioretinopathy (3, 4).

Our case report describes for the first time the monolateral association of DSM with a retinal dystrophy. The fellow eye was characterized by the presence of VMD, in the absence of any macular convexity, even though both eyes had the same refraction. It is noteworthy that the dome-shaped alteration involved the eye more affected by the disease, being classified as vitelliruptive stage. While it is difficult to hypothesize that the development of the abnormal macular convexity was

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Corresponding author:
Ilaria Zucchiatti
Department of Ophthalmology
University Vita-Salute
San Raffaele Scientific Institute
Via Olgettina 60
20132, Milan, Italy
ilaria.zucchiatti@gmail.com

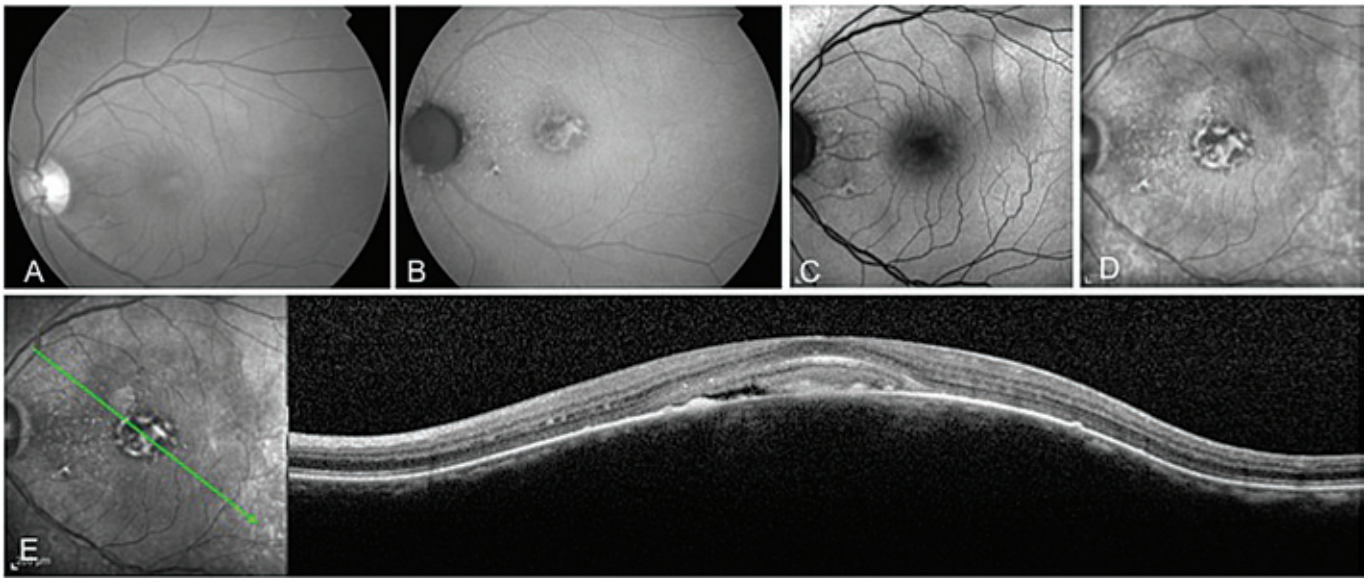

Fig. 1 - Red-free (**A**), short-wavelength fundus autofluorescence (**B**, **C**), and infrared (**D**) images show patchy vitelliform accumulation in the left eye, alternating hypoautofluorescence and hyperautofluorescence pattern. Spectral-domain optical coherence tomography (**E**) reveals a dome-shaped convexity with hyperreflective vitelliform material accumulated within the subretinal space above the retinal pigment epithelium, but below the outer segments of the photoreceptors.

promoted by the evolutive stage of VMD, a chance association may have occurred, combining different disorders in the same eye. The scleral thickness variation producing the DSM can be found in association with many conditions, including VMD.

Further investigations are warranted to evaluate the role of scleral thickness elevation in other macular dystrophies and the impact of such of such conditions on the clinical course of the disease.

Disclosure

Financial support: No financial support was received for this submission.
Conflict of interest: Francesco Bandello is a consultant for Novartis, Allergan, Farmila-Thea, Bausch and Lomb, Zeiss, and Bayer.

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