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CASE REPORT

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Dome-shaped macula with extremely extensive macular edema – a case report

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ABSTRACT

Introduction: Dome-shaped macula is characterized by a convex anterior protrusion of the macula towards the vitrous cavity, associated with high myopia and a posterior staphyloma. The etiology is not fully understood.

Case report: We report a case of a 29-year-old woman, with myopia and astigmatism presented to our department complaining of painless deterioration of vision and metamorphopsia in both eyes. The anterior segment and vitreous were unremarkable, fundoscopy showed central chorioretinal atrophy and retinal pigment epithelial changes in OU. Optical coherence tomography scans showed excessive macular edema and numerous hyporeflective sub- and intraretinal spaces in both eyes. Fundus fluorescein angiography revealed numerous punctate areas of contrast leakage at the upper-temporal vascular arch in right eye and a focus of contrast leakage at the superior-temporal vascular arch in left eye. Indocyanine green angiography presented areas with a visible intensified vascular pattern in the posterior pole of both eyes, slightly enhancing in the intermediate phase and extinguishing in the late phase.

The diagnosis of dome-shaped macula was established. After anti-inflammatory and diuretic treatment, there was no improvement in visual acuity and morphological changes. The clinical picture remains stable after 1.5 year follow-up.

Conclusions: Dome-shaped macula is still not a fully understood condition. Inconclusive clinical picture may simulate other ocular pathologies. The visual acuity and macular changes seem to be stable over the years even without treatment.

KEY WORDS: dome-shaped macula, retinoschisis, myopia.

INTRODUCTION

Dome-shaped macula (DSM) was firstly described by Gaucher and associates in 2008 as an anterior convex protrusion of the macula commonly found within a myopic staphyloma observed in optical coherence tomography (OCT) [1]. It is a rarely detected disease occurring in approximately

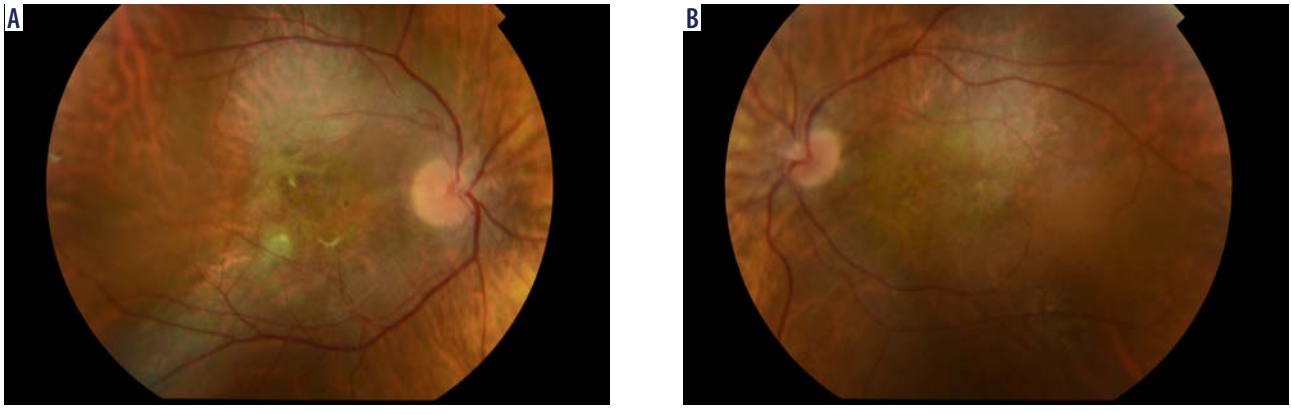

Figure 1. Colour Fundus Image: **A**) of right eye (RE); **B**) of left eye (LE)

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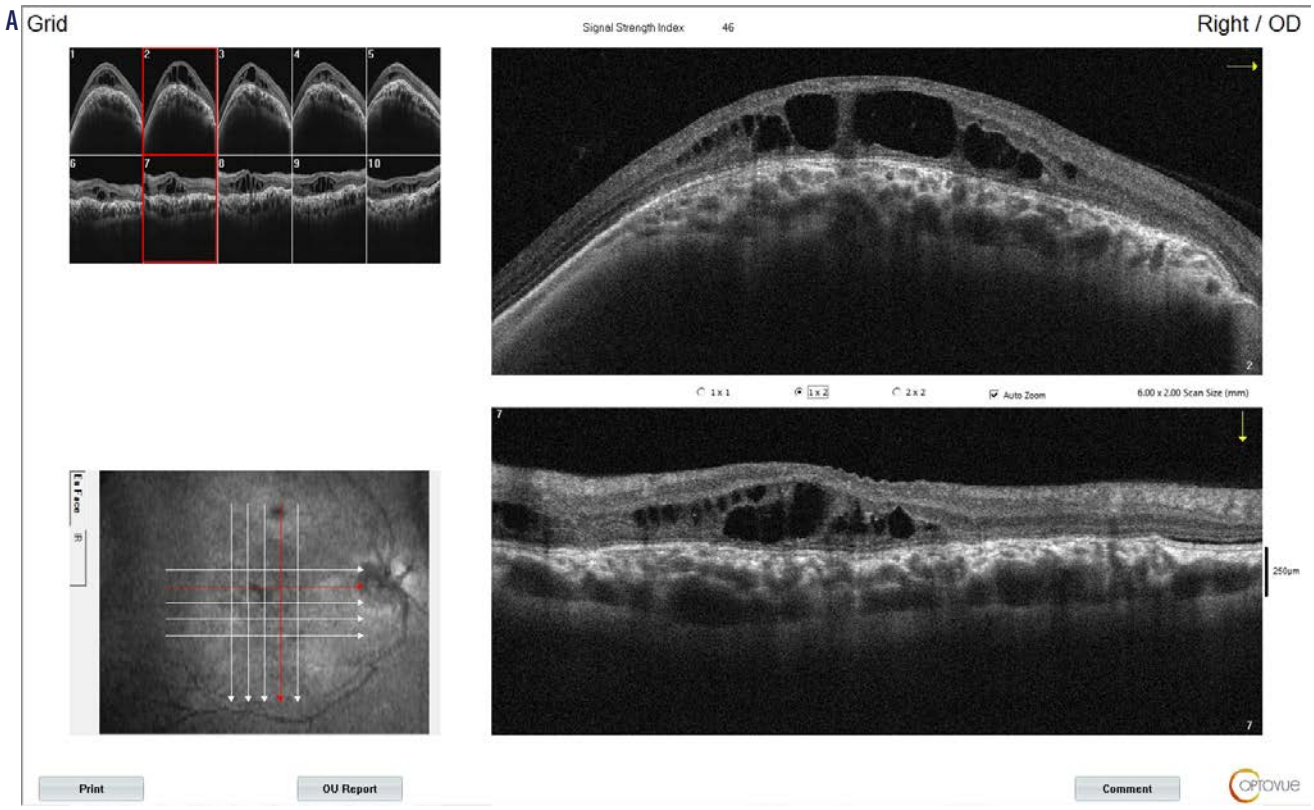

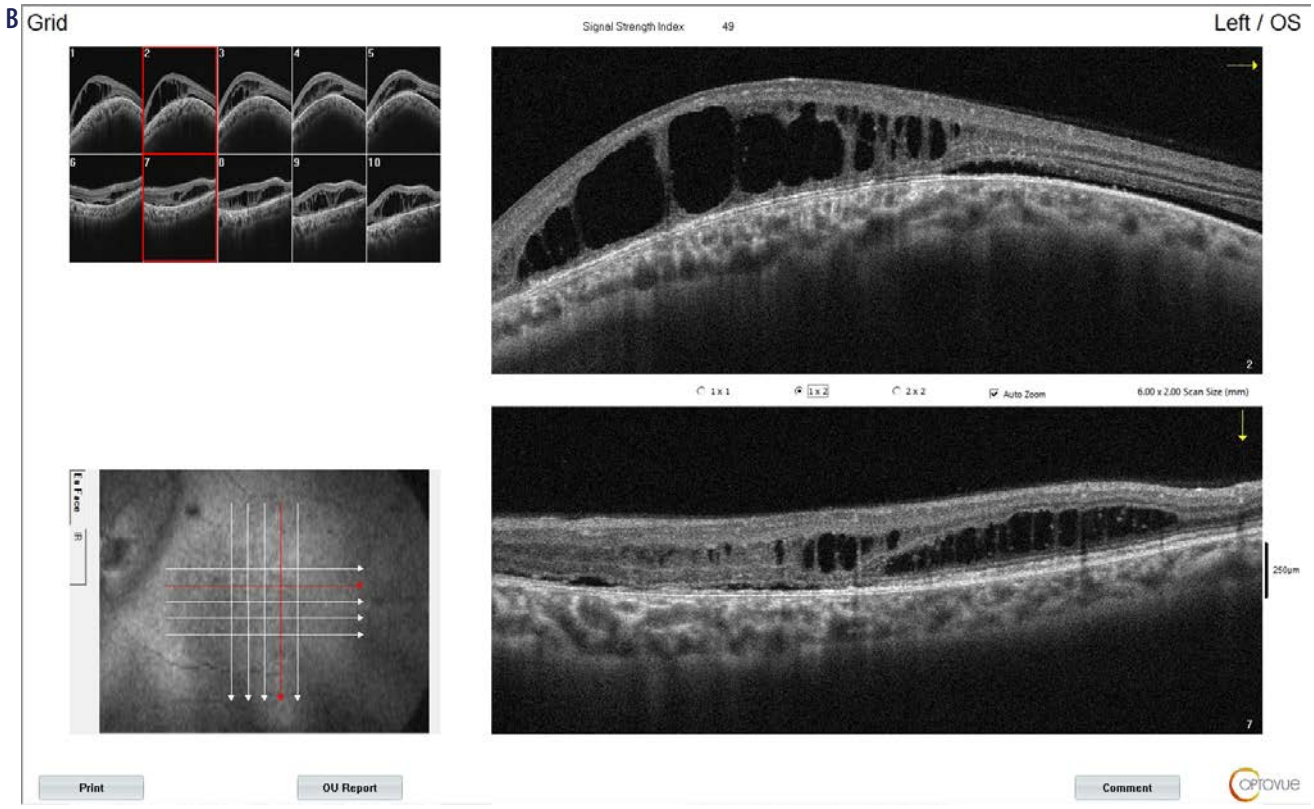

Figure 2. Optical coherence tomography (OCT): **A**) of right eye (RE); **B**) of left eye (LE)

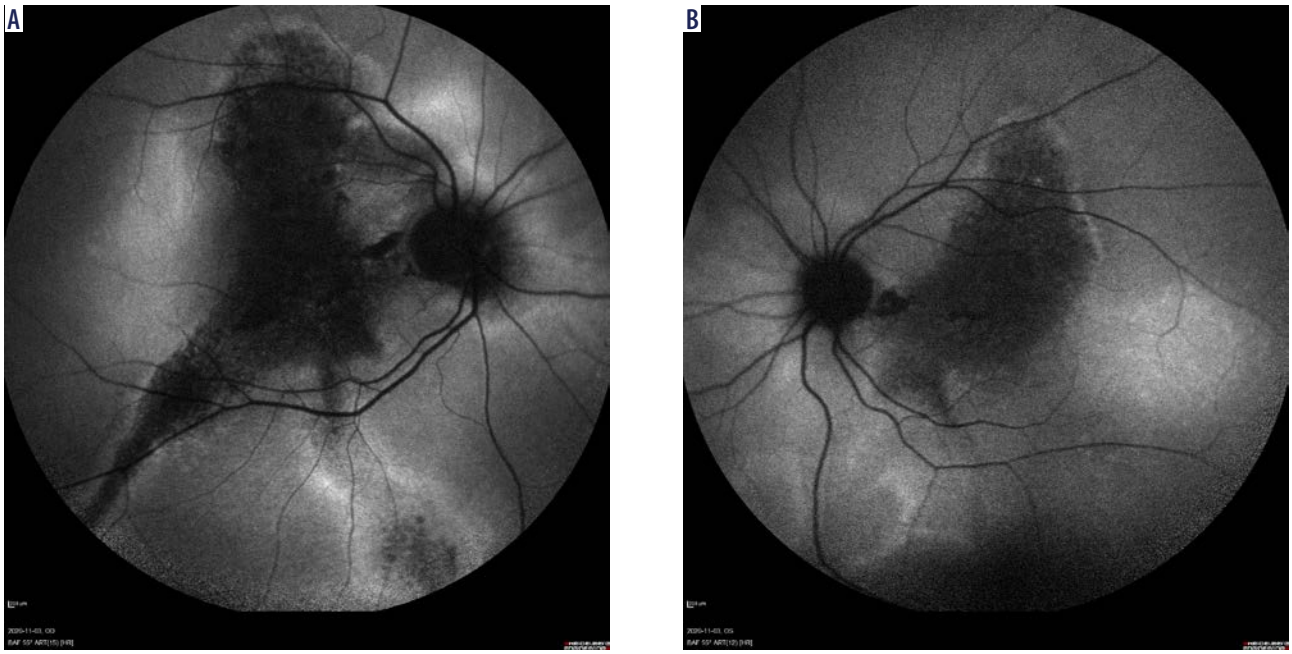

Figure 3. Autofluorescence images: **A**) of right eye (RE); **B**) of left eye (LE)

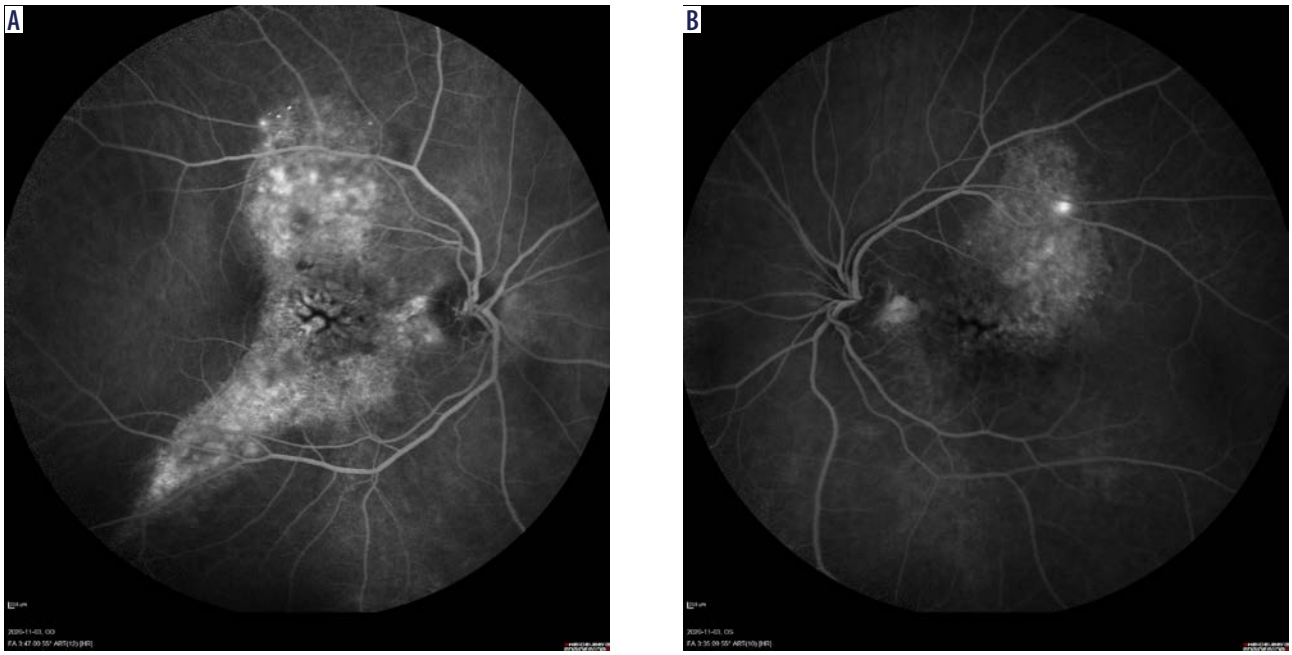

Figure 4. Fundus fluorescein angiography (FFA) reveals increasing hyperfluorescence of the hypoautofluorescent areas of the fundus: **A**) of right eye (RE); **B**) of left eye (LE)

10.7-21% of eyes with high myopia. The etiology is not fully understood. It is supposed to occur due to an increased scleral thickness which causes a disturbance of the choroidal fluid circulation [2].

The visual impairment is caused by retinal pigment epithelium (RPE) atrophic changes, sub-foveal retinal detachment, foveal and extra-foveal retinoschisis, foveal retinal detachment, sub-foveal serous detachment, macular hole and myopic choroidal neovascularization [1, 3]. In imaging examinations, dome-shaped maculopathy may simulate an intraocular tumor, including melanoma, hemangioma and choroidal metastases [4]. In differential diagnosis central serous chorioretinopathy (CSCR) and choroidal neovascular membrane (CNVM) should also be considered [5]. The article presents a case of a DSM with extremely extensive macular edema.

CASE PRESENTATION

A 29-year-old Caucasian woman with myopia and astigmatism presented to our department complaining of painless deterioration of vision and metamorphopsia in both

Figure 5. Indocyanine green angiography (ICG) – early phases: **A**) of right eye (RE); **B**) of left eye (LE)

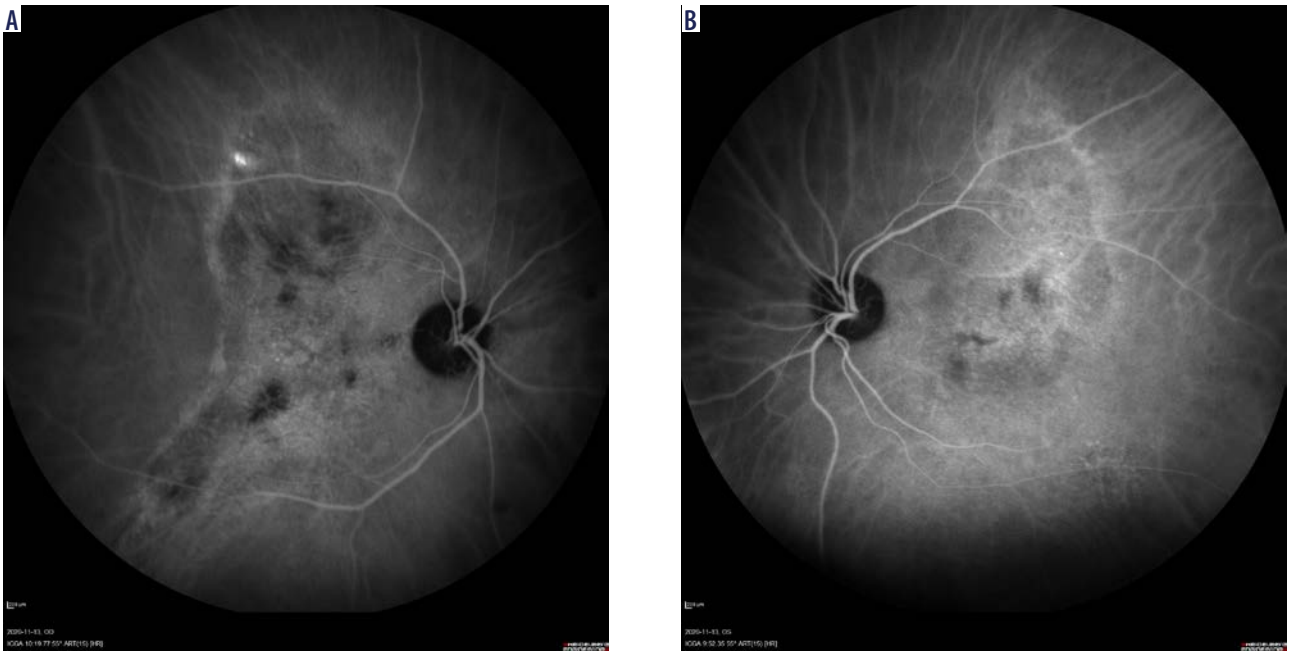

Figure 6. Indocyanine green angiography (ICG) – intermediate phases: **A**) of right eye (RE); **B**) of left eye (LE)

eyes (OU). The patient has been noticing a decrease of visual acuity from two years – since her pregnancy, most vividly within the last 3 months. There was no previous history of eye diseases, surgery, trauma or comorbidities. In the initial examination, the best-corrected visual acuity (BCVA) in the right eye (RE) was 5/50 and 3/50 in the left eye (LE) with correction –2.5[–3.5·05] and –2.0[–3.5·160], respectively. Intraocular pressure was 16 mmHg OU. The axial length of eyes measured 27 mm in RE and 26 mm in LE. The anterior segment and vitreous were unremarkable in both eyes. Pachymetry map of the cornea revealed regular astigmatism in OU. Fundoscopy showed central chorioretinal atrophy, retinal pigment epithelial changes and tilted optic discs bilaterally (Figure 1 A, B). OCT scans showed maculoschisis with large pseudocysts in outer layers of the retina and atrophic spots in fotoreceptors/RPE layers in both eyes; additionally, subretinal fluid temoprally to the macula in left eye (Figure 2 A, B).

Autofluorescence images presented hypofluorescence of the affected retina in the posterior pole with hyperfluorescence of the adjacent area in both eyes (Figure 3 A, B). Fundus fluorescein angiography (FFA) revealed numerous punctate

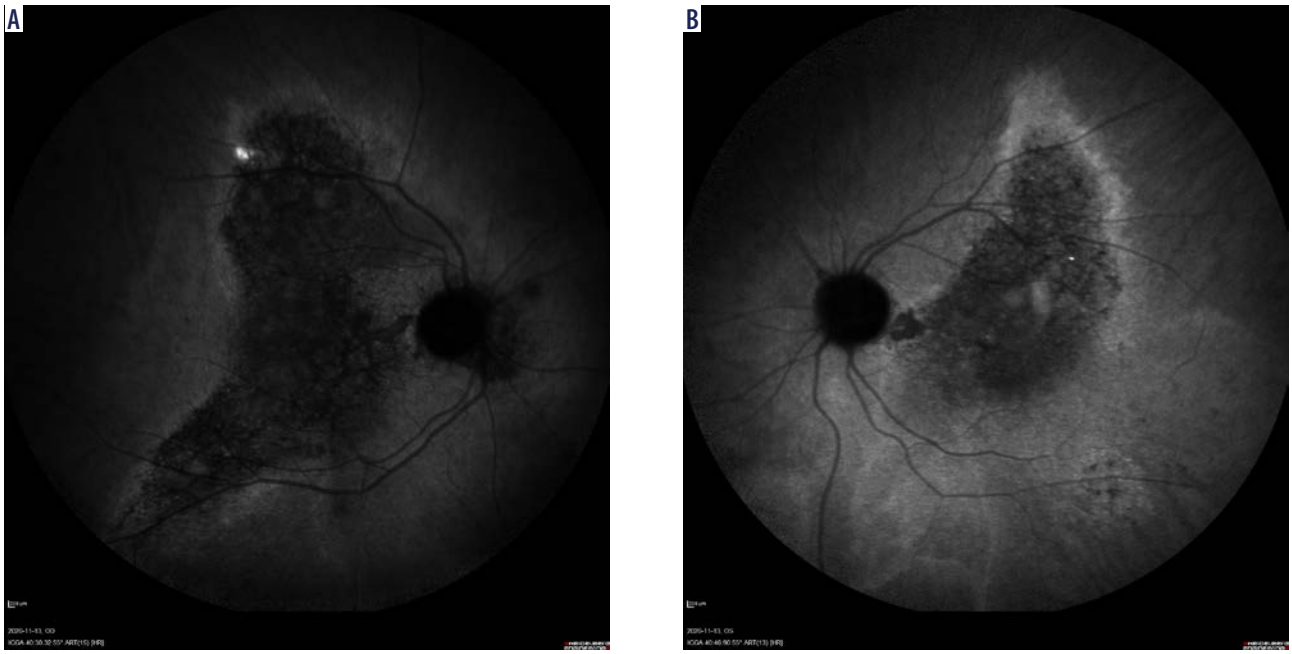

Figure 7. Indocyanine green angiography (ICG) – late phases: **A**) of right eye (RE); **B**) of left eye (LE)

areas of contrast leakage at the upper-temporal vascular arch in RE (Figure 4 A) and a focus of contrast leakage at the superior-temporal vascular arch in LE (Figure 4 B). Indocyanine green angiography (ICG) presented areas with a visible intensified vascular pattern in the posterior pole of RE, slightly enhancing in the intermediate phase and extinguishing in the late phase. Additionally in the upper-temporal vascular arch from the intermediate to the late phase, the focus of hyperfluorescence, increasing over time was noted (Figures 5 A, 6 A, 7 A). In the posterior pole of LE a limited area with a visible enhanced vascular pattern with a slight enhancement in the intermediate phase and extinction in the late phase was observed (Figures 5 B, 6 B, 7 B). Choroidal neovascularization was ruled out by OCT angiography (Figure 8 A, B).

B-mode ultrasound examination excluded fresh exudate and revealed within the macula and the vascular bundle a slightly elevated, echodense, dome-shaped mass with features of a retinoschisis in OU (Figure 9 A, B).

Laboratory results were within reference ranges, including complete blood count, sedimentation rate, C-reactive protein, random blood sugar level, urine test, renal function tests, activated partial thromboplastin time (APTT), international normalized ratio (INR), liver enzyme, homocysteine, and angiotensin converting enzyme (ACE). Results of B-hCG pregnancy testing were negative. Infectious workup revealed positive IgG antibodies against Cytomegalovirus (3796) and Herpes virus (3.64), the normal range respectively: < 0.5 and < 0.8 ratio. *Toxoplasma gondii, Bartonella henselae, Borrelia burgdorferi*, QuantiFERON-TB Gold, Venereal Diseases Research Laboratory (VDRL) and HIV test were negative.

Head and orbit magnetic resonance imaging (MRI) with contrast revealed increased thickness of the posterior pole of both eyeballs featuring contrast enhancement (Figure 10).

Initially, as retinitis or posterior scleritis was taken into considereation (due to MRI results), during hospitalization the patient received 500 mg of methylprednisolone (Solu-Medrol, Pfizer Europe) intravenously once daily for 6 days, 100 mg doxycyclinum (Doxycyclinum, Polfarmex) orally twice daily and 800 mg acyclovir (Heviran, Polpharma) orally five times daily. Dexamethason (Dexamethason 0,1% WZF, Polfa Warszawa) and diclofenac (Dicloabak, Laboratories Thea) was administered five times and twice daily respectively to OU. Improvement during hospitalization was not achieved. Further treatment consisted of 32 mg of methylprednisolone orally (Metypred, Orion) with tapering the dose, as well as antibiotic and acyclovir.

During a follow-up a control FFA and OCT scans were performed. The results were comparable with those performed on hospital admission.

Based on a constellation of clinical and angiographic features as well as the elimination of potential underlying systemic conditions, a diagnosis of dome-shaped macula syndrome was established.

On the follow up visit, the patient was administered diuretic treatment, eplerenone (Inspra, Upjohn EESV) 50 mg orally once daily. After 6 months of treatment, there was no improvement in visual acuity and morphological changes. The patient remains under regular follow-up (1.5 year). The clinical picture remains stable.

DISCUSSION

Dome-shaped maculopathy, initially described among some patients with myopic staphyloma, has been also reported in emmetropes, hypermetropes as well as eyes with other diagnoses [6].

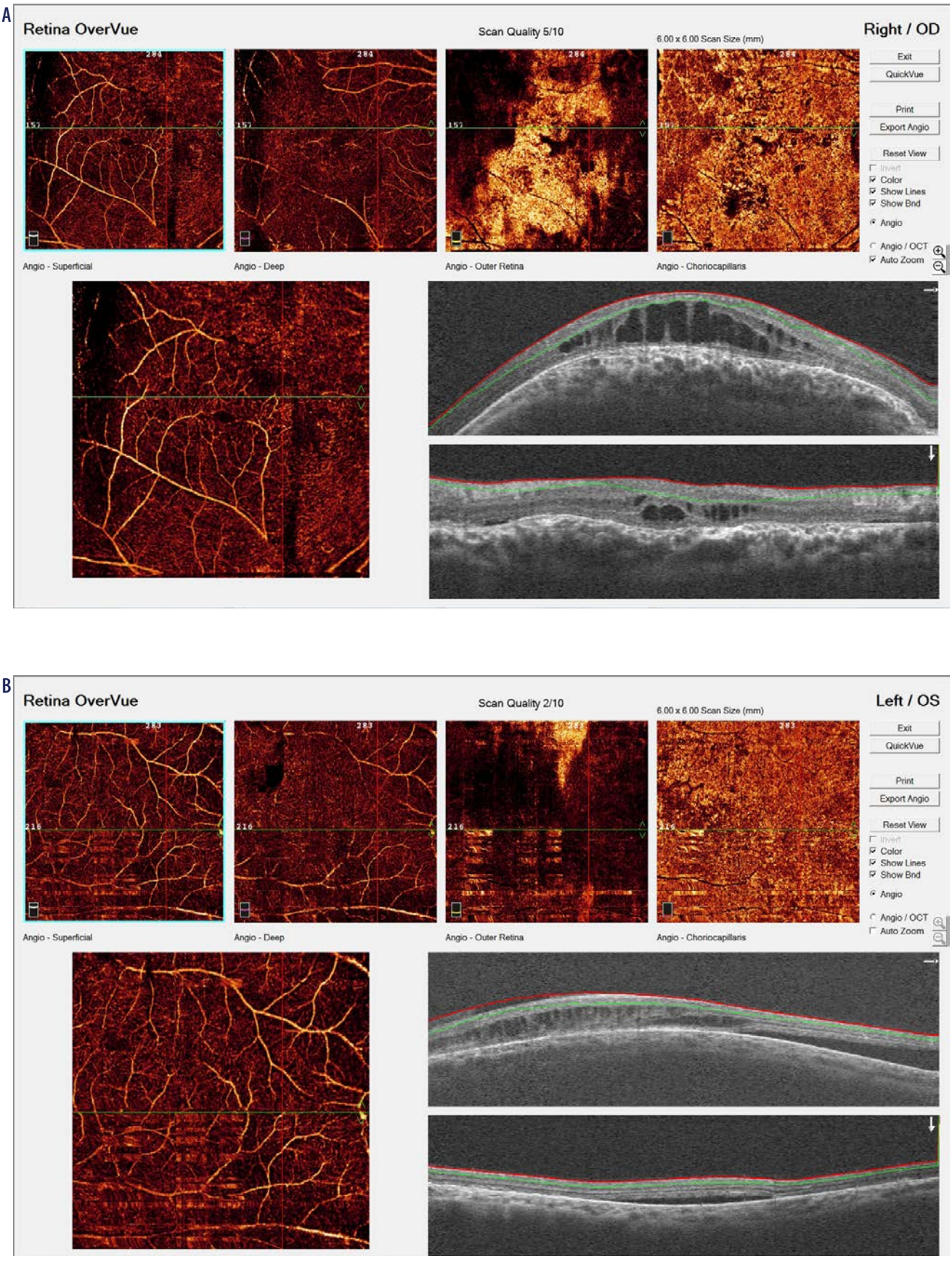

Figure 8. OCT Angiography: **A**) of right eye (RE); **B**) of left eye (LE)

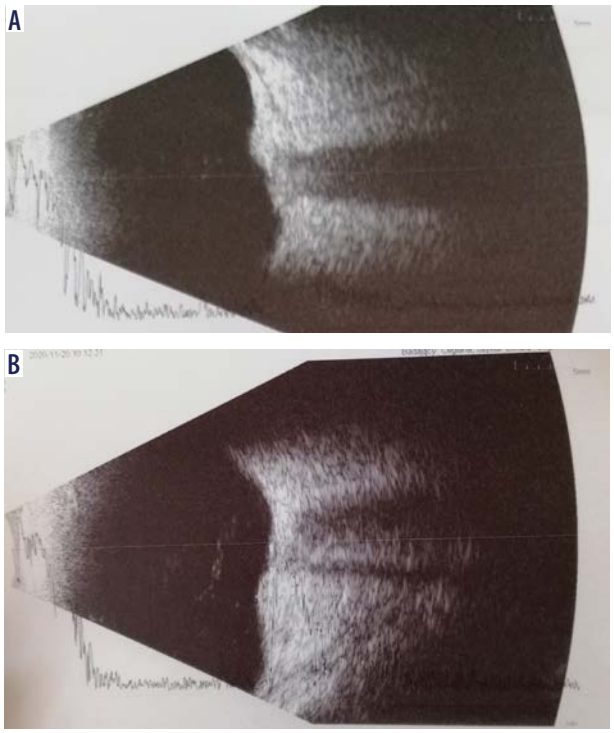

Figure 9. B-mode ultrasound examination (USG) – transverse projection: **A**) of right eye (RE); **B**) of left eye (LE)

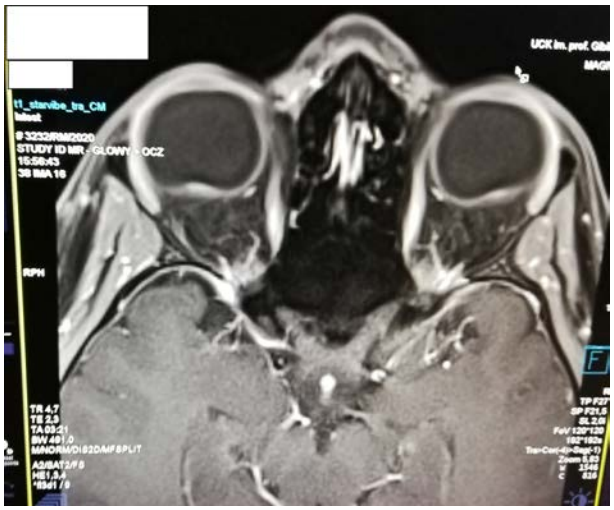

Figure 10. Orbit magnetic resonance imaging (MRI) with contrast

Dome-shaped macula is probably associated with asymmetric scleral growth. This condition seems to arise from local changes of the scleral biomechanical properties due to the emmetropization process. The clinical features are secondary to changes in the macular area and range from being asymptomatic to metamorphopsia and sometimes gradual progression to visual loss. Acute deterioration of vision in DSM is usually due to choroidal neovascular membrane (CNVM) development [2].

In the study performed by Liang *et al*., who compared retinal complications among highly myopic eyes with and without a dome-shaped macula, the incidence of CNMV has been found to be the minor factor. Moreover, DSM increases the rate of complications, especially a sub-foveal retinal detachment (SRD), which is considered as one of the major complications of DSM in Western countries [4, 7]. The formation of SRD also named as sub-foveal serous fluid (SRF) is not fully understood and is hypothesized due to choroidal vascular changes, similar to CSCR, but as in CSCR there is a generalized choroid thickening, in DSM is particularly confined to the submacular region and secondary to excessive scleral thickening [2, 8].

Dome-shaped macula is found to be a protective factor for foveal retinoschisis but increases the incidence of extra-foveal retinoschisis [7, 9]. As this is not consistent with the case we present, we suspect the profound retinal changes result from choroidal disfunction rather than mechanical factors, but further investigation is needed.

Caillaux and colleagues analyzed the morphology of the macular bulge in a dome-shaped macula. They classified DSM into 3 morphologic patterns, using spectral domain (SD) OCT, depending on the topographic features. They distinguished: round domes and horizontal (the most common) or vertical (the most rare) oval-shaped domes, which differ in the sclerochoroid elevation with hemispheric, symmetric macular convexity or horizontally and vertically elongated, respectively. The staphyloma in oval-shaped domes, referred also as ridge-shaped macula (RSM), is divided into two parts: superior and inferior in horizontally oriented and nasal and temporal in vertically oriented [10]. The RSM is also considered as an early variant of DSM seen in young myopic patients with progressive increase in height of macular bulge [5].

According to our patient, the morphologic pattern corresponds to ridge-shape macula.

In work presented by García-Zamora *et al*., in ridge-shaped macula there was a lower macular elevation than in typical round domes, thus meaningly less frequent Bruch's membrane (BM) defects. The axial length (AL) in eyes with round domes was statistically significant longer than in RSM. The differentiation of those morphologic patterns have a clinical significancer – help to identify in a daily clinical practice, eyes with a higher risk of developing severe myopic complications [11].

The red-orange hue of fundus image in DSM with central chorioretinal atrophy and retinal pigment epithelial changes as well as other features such as serous retinal detachment, cystoid macular edema may resemble a choroidal hemangioma in its circumscribed form, which is a benign vascular and typically unilateral tumor. In contrast to DSM, choroidal hemangioma on FFA, demonstrates early pre-arterial hyperfluorescence with late diffuse hyperfluorescence of the mass with leakage into the subretinal space while there is occasional staining from subretinal fluid (SRF) in cases of DSM. On ICG, DSM is iso-fluorescent, while haemangioma shows a bright hyper-fluorescence with characteristic 'washout' phenomenon [4, 12].

The potential treatment options of SRD in DSM encompass photodynamic therapy (PDT), laser photo-coagulation, anti-vascular endothelial growth factor (anti-VEGF)injec-

tion, mineralocorticoid receptor antagonists and intravitreal steroid injections. However, the efficacy of treatment has not been confirmed for any of the above-mentioned. Studies conducted so far have not shown statistically significant differences in the improvement of visual acuity among treated and untreated people [13].

The RADIANCE study, where the response for anti-VEGF injection among myopic CNVM patients with and without DSM was evaluated, revealed that the presence of DSM did not change the treatment response. However, PDT has been found to perform better in DSM patients compared to those without DSM [14].

Battaglia Parodi and coworkers performed the 12-month follow-up pilot study where the subthreshold laser treatment improved BCVA and central foveal thickness in myopic eyes with serous retinal detachment secondary to DSM. However, the study group encompasses only 12 eyes (8 patients). Further investigations in this field should be performed [15].

Fernandez and associates evaluated 24 eyes of 12 patients with SMD in the course of DSM and revealed a significant reduction in SRF and central thickness after 6-month therapy with 50 mg spironolactone daily, however, with no visual improvement [16].

We tried to cure our patient with antagonist of the mineralocorticoid receptors – 50 mg eplerenone daily, but after 6 months of treatment there was no improvement observed.

Vukkadala and coworkers presented a rare case of pediatric patient with bilateral foveal cysts, so far unprecedented in the course of DSM, who positively respond to topical drozlamid. This suggest a possible therapeutic option which should also be considered [17].

In conclusion, DSM is still not a fully understood condition in terms of its pathogenesis and complications. The visual acuity and macular changes seem to be stable over the years even without treatment [18].

Analyzing the above, the most reasonable therapeutic approach seems to be observation.

In making the diagnosis of DSM, other conditions, which clinical presentation may be similar, should be first ruled out.

DISCLOSURE

The authors declare no conflict of interest.

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