Review paper

Dome-shaped macula: review of the literature

Zespół kopulastej plamki: przegląd piśmiennictwa

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Słowa kluczowe: zespół kopulastej plamki, optyczna koherentna tomografia, krótkowzroczność.

Abstract

Dome-shaped macula is a pathology of the posterior pole of the eye, characterized by a convex anterior protrusion of the macula. It is most commonly associated with high myopia, but can also occur in non-myopic patients. Patients may report decreased visual acuity but may also be asymptomatic. Due to the lack of specific features in the ophthalmoscopic examination, swept source optical coherence tomography (SS OCT) is the most important diagnostic tool, in which both vertical and horizontal scans should be performed. Despite the lack of effective treatment, patients should be systematically followed up. In some cases, treatment methods such as intravitreal injections of aflibercept, vitrectomy, focal laser photocoagulation and photodynamic therapy are considered.

Streszczenie

Zespół kopulastej plamki obejmuje tylny biegun gałki ocznej i charakteryzuje się przednio-wypukłym ułożeniem plamki żółtej. Najczęściej jest on związany z wysoką krótkowzrocznością, ale występuje również u osób normo- i nadwzrocznych. Pacjenci z zespołem kopulastej plamki mogą być bezobjawowi lub zgłaszać obniżenie ostrości wzroku. W związku z brakiem specyficznych cech w badaniu oftalmoskopowym swept source optyczna koherentna tomografia siatkówki jest najważniejszym narzędziem diagnostycznym, którego wykonanie powinno obejmować skany zarówno pionowe, jak i poziome. Pomimo braku efektywnego leczenia należy pacjentów systematycznie kontrolować. W niektórych przypadkach bierze się pod uwagę terapię za pomocą iniekcji afliberceptu do komory ciała szklistego, witrektomii, fotokoagulacji laserowej oraz terapii fotodynamicznej.

Patients with macular diseases complain of a deterioration in distance and near visual acuity, the presence of metamorphopsia and scotomas in the visual field. Availability of diagnostic devices, such as optical coherence tomography (OCT), fluorescein angiography (FA), B-scan ultrasonography allows to make an appropriate diagnosis and propose adequate treatment. The dome-shaped macula (DSM) is one of the pathologies of the posterior segment of the eye, which should be taken into account in the differentiation of macular diseases, especially in younger patients with myopia [1].

DSM is the pathology of the posterior pole of the eye, which was first described by Gaucher [2]. The diagnosis of this pathology is made on the basis of a fundus examination as well as additional examinations. It is characterized by the presence of a convex elevation of the RPE, retina and choroid in the posterior segment of the eye identified by OCT. The swept source OCT (SS OCT) examination enables a reliable diagnosis of DSM due to the deep penetration of light waves into the choroid and more accurate imaging of the structures of the posterior pole of the eye. The height of the dome is greater than 50 µm above the line connecting the RPE on both sides of the DSM in the vertical or horizontal measurement in the OCT [3] (Figures 1, 2).

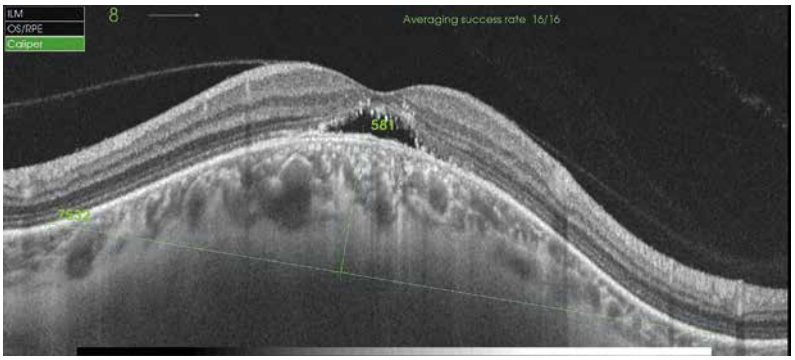

Figure 1. Swept source optical coherence tomography (SS-OCT) of RE with DSM – horizontal scan

Figure 2. SS-OCT of RE with DSM – horizontal scan – the height of the inward bulge of RPE

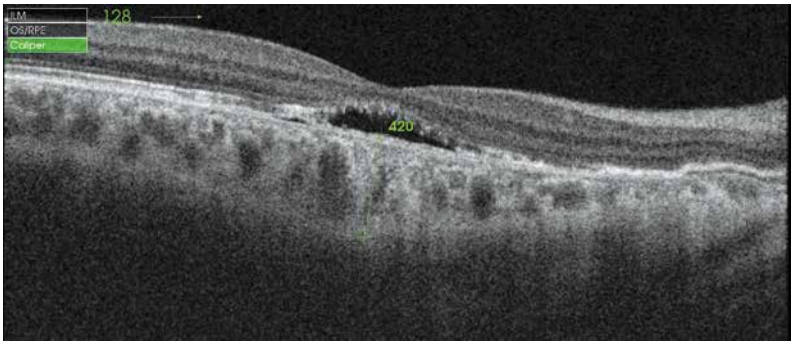

Figure 3. SS-OCT of RE with DSM – vertical scan – increased choroidal thickness

DMS occurs mainly in myopia, but also in emmetropia and hyperopia. The syndrome is usually diagnosed in the 4th and 5th decade of life. The occurrence of the above pathology does not depend on sex. Bilateral DSM is found in 30–80% of cases. This syndrome is diagnosed in adults as well as children and adolescents. DSM is a pathology that remains stable over time [4].

The pathogenesis of DSM is unclear. Genetic predisposition is taken into account, as well as mechanical dysfunction of RPE resulting from anatomical disorders at the site of the lesion. In the eye with DSM, the Bruch's membrane defects may occur [5]. The choroid thickening within the lesion with the presence of thin choroid around the DSM is also taken into account [6] (Figure 3). The pathogenesis of the above pathology may also involve a localized thickening of the sclera within the lesion with subsequent disturbance of choroidal outflow [7].

The diagnosis of DSM may be random as it could be asymptomatic. Sometimes symptoms such as decreased visual acuity, the presence of metamorphopsia and central scotoma in the visual field occur during the course of the disease. It may result from complications of CNV accompanying DSM and serous macular detachment, the incidence of which is estimated, depending on the researcher, at a level of 2% to 67% [8].

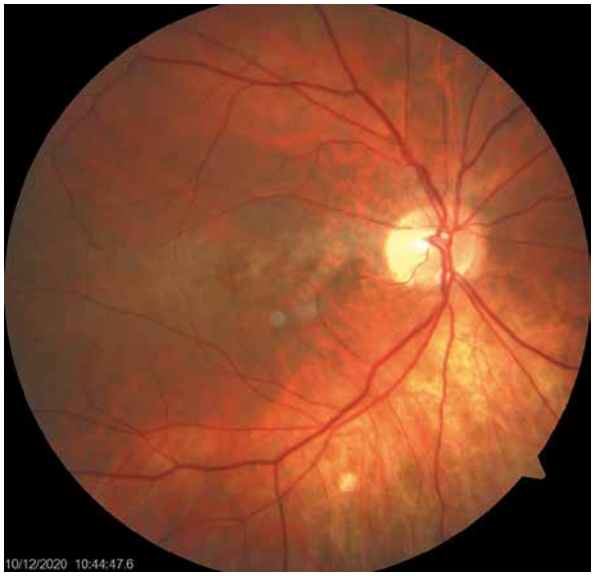

Figure 4. The fundus of the right eye (RE) with dome-shaped macula (DSM): atrophic changes in the macular retinal pigment epithelium (RPE) and dull macular reflex

The fundus examination in patients with DSM reveal the presence of a dull reflex, changes of the retinal pigment epithelium (Figure 4). Sometimes intraretinal or preretinal hemorrhages occur, if CNV is present. Angiography OCT is a useful tool to detect CNV (Figure 5). For myopia patients, the central and peripheral retina may be thin and the optic disc paler [9].

The early phase of the fluorescein angiography shows mild hyperfluorescence in the macula (Figure 6) gradually increasing in the late phase (Figure 7). The autofluorescence examination of the fundus shows hypofluorescence in the macula (Figure 8). For myopia patients, the central and peripheral retina may be thin and the optic disc paler [9].

DSM should be differentiated from other pathologies, including myopia with a posterior hump, where the entire choroid is thinned [10]. Another pathology that should be taken into account in the differentiation of DSM is the Tilted Disc Syndrome, characterized by a typical oblique appearance of the optic nerve head and thinning of the entire choroid [11]. Eyeball tumors, which can cause the typical 'modeling symptom' in OCT, must be excluded. These might be malignant melanoma, metastasis, granuloma or choroidal hemangioma.

The B-scan ultrasonography is useful to verify the diagnosis [12]. A hyperechogenic lesion may be demonstrated in the posterior pole of the eye (Figure 9). If the CNV occurs in the OCTA, it should lead us to exclude other macular pathologies connected with CNV including neovascular age-related macular degeneration (nAMD), CNV associated with myopia and central serous chorioretinopathy [13, 14]. Anti-VEGF therapy is an effective method of treatment for macular neovascularization in the course of nAMD

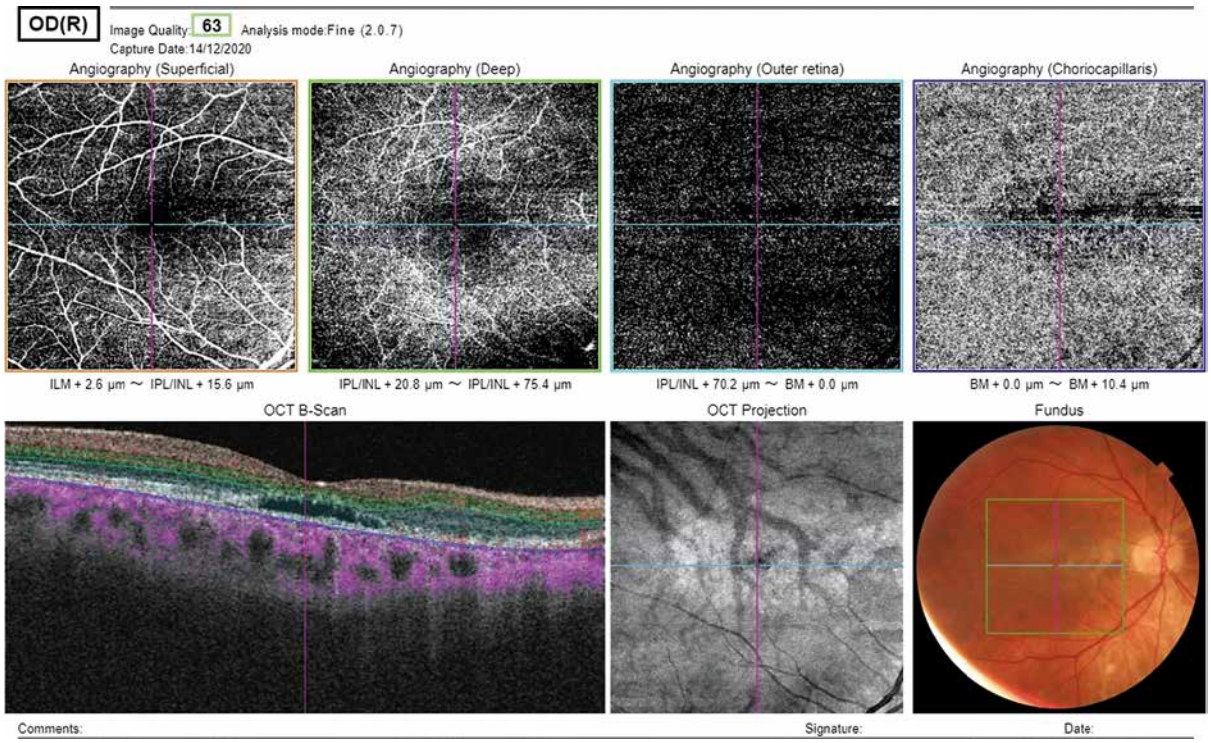

Figure 5. OCTA with DSM – no choroidal neovascularization detected

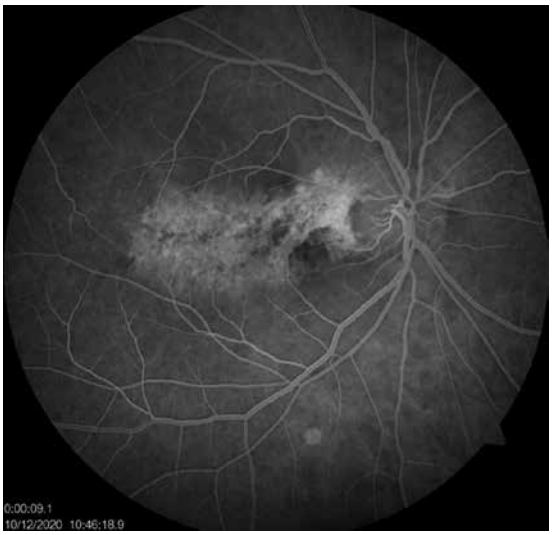

Figure 6. The early phase of the fluorescein angiogram (FA) of RE – mild hyperfluorescence in the macula

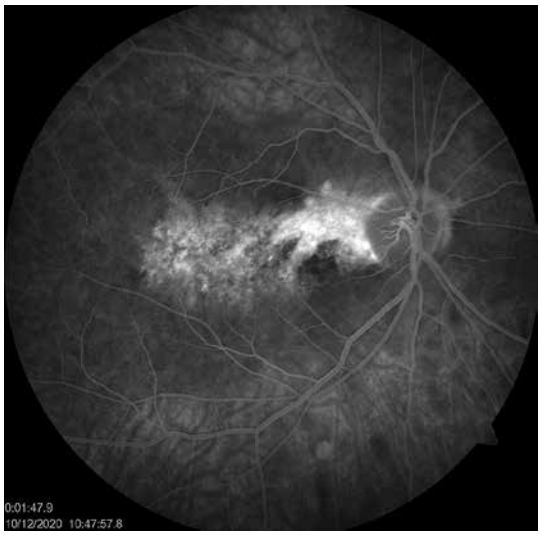

Figure 7. The late phase of FA of the RE – gradually increasing hyperfluorescence in the macula

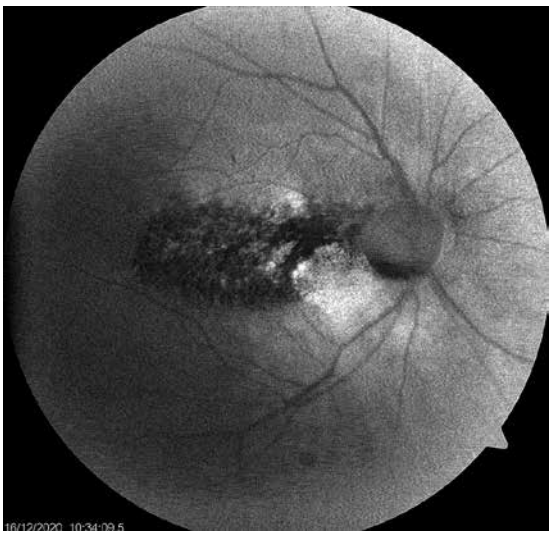

Figure 8. Autofluorescence of the fundus of RE with DSM – hypofluorescence in the macula

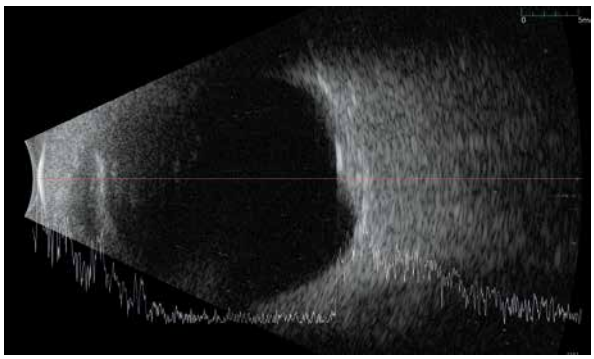

Figure 9. Ultrasonography, scan B of RE with DSM – a hyperechogenic lesion in the posterior pole

however some patients do not respond positively to therapy [15]. There is no clearly defined treatment for patients with DSM but patients should be monitored regularly. Available publications mention the possibility of using intravitreal injections of anti-VEGF medicines if CNV coexists, in addition, focal laser photocoagulation (micropulse laser), photodynamic therapy (PDT), local carbonic anhydrase inhibitors and orally administered mineralocorticosteroid receptor antagonists are used. The effectiveness of the above methods in improving visual acuity is unsatisfactory [16–19].

In conclusion, choroidal disorders are more and more often considered in the development of many diseases of the posterior segment of the eye. The SS-OCT technology enables detailed imaging of the choroid, improves the accurate diagnosis of the DSM and the differential diagnosis of the posterior segment diseases.

Conflict of interest

The authors declare no conflict of interest.

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