Chapter 7

Contribution to the Optic Nerve

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Additional information is available at the end of the chapter

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Abstract

Optic nerve head drusen (ONHD) represent congenital anomaly, which is a form of calcium degeneration of optic nerve head axons. They are initially asymptomatic but may causes progressive optic neuropathy. They are presented as acellular, hyaline deposits of globular appearance in front of the lamina cribrosa (prelaminar segment). They are formed due to altered axonal transport, small diameter of scleral canal, direct compression, or ischemia. Frequent complications: progressive visual field scotoma, ischemic optic neuropathy, central retinal artery or vein occlusion, and neovascularization adjacent to the optic nerve head. Useful diagnostic tools ophthalmoscopy, angiography, standard automated perimetry, B-scan ultrasonography, CT, OCT, HRT, GDx, and electrophysiological examination. Therapeutic procedures are medical, laser, and surgical. Pilot studies confirmed benefit of topical hypotensive drugs even when drusen are not associated with glaucoma. By reducing intraocular pressure, the compression to the optic nerve axons decreases, thus improving perfusion of the optic nerve head. The paper presents young female patient with bilateral optic nerve drusen and progressive visual field defects (scotomas), which implies topical hypotensive therapy. After 6 months scotoma and loss of sensitivity of the visual field were reduced. Neuroprotective drugs are investigated to reduce potential visual morbidity.

Keywords: optic nerve head, drusen, visual field, topical hypotensive drug, glaucoma

1. Introduction

Optic nerve head drusen (ONHD) represent the congenital, developmental anomaly of the second cranial nerve. This is a form of calcium degeneration of the axons of the optic nerve head (ONH). They are initially asymptomatic, but are not so rare and can cause lower visual acuity. Thus, they are also considered to be one of the causes of the progressive type of optic neuropathy with genetic etiology [1, 2].

Their occurrence is most common in 3.4–37 per 1000 adult white people (0.34–3.7%), with a frequent bilateral presentation in 75–85% of all cases [3, 4]. They are less common in black people, equally represented among the sexes, while the average prevalence in the pediatric population is 0.4% [5–7].

For the first time, they were histologically recognized by Heinrich Muller in the nineteenth century (1858) [8, 9]. Clinically, they were described with the contribution of Liebreich in 1868, only 17 years after the construction of the direct ophthalmoscope [10]. A detailed structure was described by Tso in 1967. The author also postulates the pathophysiological mechanisms of the formation and evolution of the drusen [11].

Genetically, optic nerve head drusen are inherited in autosomal dominant (AD) type of inheritance with variable penetration. Lorentzen et al. founded that the incidence of drusen is 10 times higher among members of the family with manifest optic nerve drusen [9]. Drusen may occur primarily isolated or associated with the eye (angioid streaks, retinitis pigmentosa, open-angle glaucoma, etc.) (**Table 1**) or as systemic diseases (pseudoxanthoma elasticum (PXE) or Gronblad-Strandberg syndrome, Usher syndrome, Noonan syndrome, etc.) (**Table 2**) [7, 12]. Antclif et al. state that the inheritance of a small diameter (disk area) of the optic nerve head is a risk factor for formation of optic nerve head drusen. The small optic disk (ONH) and mesodermal dysplasia result in vascular dysplasia and therefore are considered as significant causative factors. Optic disk drusen are more commonly present in vascular and/or structural developmental anomalies of the ONH. Among the vascular anomalies, the most frequent patterns of disorders are two and three branches of blood vessels, cilioretinal artery, and optociliary shunt [9, 11].

Acquired myelinated nerve fibers

Aneurysm of the ophthalmic artery

Astrocytic hamartoma

Best's vitelliform macular dystrophy

β-thalassemia

Birdshot chorioretinopathy

Combined hamartoma of the retina and retinal pigment epithelium

Congenital night blindness

Familial macular dystrophy

Glaucoma

Gyrate atrophy

Idiopathic intracranial hypertension

Idiopathic parafoveal telangiectasia

Morning glory disk anomaly

Ocular tumoral calcinosis

Optic nerve tumors
Peripapillary central serous retinopathy
Pigmented paravenous retinochoroidal atrophy (PPRCA)
Pseudoxanthoma elasticum and angioid streaks
Retinitis pigmentosa
Severe early childhood onset retinal dystrophy (SECORD)
Tilted optic disk
Tubulointerstitial nephritis and uveitis (TINU) syndrome

Table 1. Ocular diseases associated with optic nerve head drusen.

Alport syndrome
Cystic fibrosis
Delayed language development and dyslexia
Down syndrome
Headache and seizure disorders
Psychomotor retardation
Schizophrenia
Sturge-Weber syndrome
Teeth and jaw anomalies
Trisomy 15q
Tuberous sclerosis

Table 2. Systemic diseases associated with optic nerve head drusen.

Optic nerve head drusen are acellular, hyaline deposits of globular appearance in the prelaminar segment of the optic nerve head, usually below the level of the Bruch membrane of the surrounding retina. They are formed of amino and nucleic acids, mucopolysaccharides, calcium, and iron. They represent a dynamic structure, which increases during lifetime due to calcium deposition and consequent compression of the surrounding axons [4, 11]. Among the etiological factors for optic nerve drusen formation are altered axonal transport, the small diameter of the scleral canal and the ONH, direct compression, and ischemia [4, 13].

Tso states that altered axonal metabolism causes intracellular calcification of the mitochondria, degeneration and rupture of the axons with extrusion, and deposition of mitochondrial content into the extracellular space. Seitz points out that slow antero- and/or retrograde axoplasmic transport initiates the disintegration of the axons with subsequent calcium deposition and accumulation. The small diameter of the scleral canal and the specific anatomy of the

ONH compromise the axoplasmic transport. Mechanical compression causes delay of axonal transport (stasis) [5, 11, 14]. In the younger age, optic nerve drusen are near to the lamina cribrosa, and they are overlapped by nerve and vascular structures and are called *hidden* (*buried*). Optic nerve head drusen during life, become visible, and are elevated in predilected nasal-lower sector of the ONH.

Clinical classification of ONH drusen is as follows:

Grade I: deep, hidden formations

Grade II: visible separated drusen (1–6)

Grade III: dense prominent drusen (> 6) with an unclear border ONH [4, 15]

Most patients are asymptomatic for a long time. Rarely, they can have transient visual obscurations in the form of a flicker (9%) but often are associated with progressive visual field defects (scotoma) and relative afferent pupillary defect (RAPD) in advanced cases. The incidence of visual field defects in adults with drusen is 24–87%. Visual field scotoma can be an arcuate, peripheral enlargement of the blind spot, a less frequent nasal step, and a constriction of the visual field. The central visual acuity is preserved for a long time; although after a long lasting of narrowed visual field, sudden loss of central vision is possible, thus indicating complications [11, 16, 17].

Besides visual field defects due to the compression of the retinal ganglion cells' axons, numerous vascular complications due to ischemia may occur, such as non-arteritic ischemic optic neuropathy (NAION); central retinal artery occlusion (CRAO); branch or central retinal vein occlusion (BRVO, CRVO); intravitreal, subretinal, and flamed disk hemorrhage; maculopathy; and neovascularization adjacent to the optic nerve head [4, 16, 18].

The diagnosis is made by clinical examination and additional investigation. Superficial drusen are yellowish-oval prominent structures that give an unclear appearance of the ONH border, and they are considered as the main cause of pseudoedema. Additional methods, photofundus and fluorescein angiography, fundus autofluorescence, computerized perimetry (SAP), B-scan ultrasonography, OCT (EDI-OCT, SS-OCT, OCT angiography), HRT, GDx, computerized tomography (CT), and electrophysiological testing, are applied in the confirmation of hidden or superficial drusen [7, 16, 18].

Differential diagnosis is significant if pseudoedema of ONH drusen is suspected in aim to distinct numerous causes of the true edema of ONH: inflammatory, stagnant, or ischemic etiology. Buried drusen frequently produce elevation of the optic nerve head and obscuration of the margins, making it difficult to distinguish buried drusen from true papilledema based solely on funduscopic examination. However, during examination, it could be noticed that in true papilledema, the optic nerve head swelling extends into the peripapillary retina, causing RNFL thickening and consequently obscuration of the peripapillary vasculature. In contrast, elevation from optic nerve head drusen is confined to the optic nerve disk. In the case of pseudoedema, the absence of optic nerve head hyperemia, obscuration of surface arteries, exudates, venous congestion, cotton-wool spots, and peripapillary circumferential subretinal fluid lines (Patton's lines) may help distinguish optic nerve head drusen from true papilledema [7, 19].

In differential diagnosis angiography could be helpful equally. While profuse leakage is apparent in the early angiographic phases of the edematous optic nerve head, the drusen

optic nerve should never leak. However, there is late staining of the nerve on the angiogram. It is important not to mistake this late staining for leakage in the early phase; besides an autofluorescence phenomenon and OCT ONH features can help to secure this diagnosis [20].

Therapy can be medical, laser, and surgical. There is no generally accepted and approved therapeutic protocol that is applied in the case of the ONH. Monitoring is advised in asymptomatic patients, while the application of any therapeutic modality is determined by potential complications of the drusen. A large number of studies confirm the benefit of topical hypotensive drugs even in cases where ONH drusen are not associated with glaucoma. With the reduction of intraocular pressure is achieved, which reduces mechanical pressure on the axons of the optic nerve, indirectly the reperfusion as main goal is acheived. Choroidal neovascular membranes (CNV) adjacent to the optic nerve are treated with anti-VEGF or laser therapy [12, 16, 21]. The proposed surgical therapeutic modality, radial optic neurotomia, was used to decompress the head of the optic nerve due to ischemic neuropathy. The method was described by Opremcak (2001) in the treatment of CRVO and was also applied to progressive scotoma caused by drusen. Also successful and unsuccessful excision of drusen was reported but rarely applied [21–25].

The main aim is to provide recent data of etiology, epidemiology, pathology, diagnosis, and potential treatment of optic nerve head drusen. All relevant data was collected by investigation in small samples or individual cases and reports. There is no major controlled clinical trial on therapeutic effects and protocol in drusen, but there are numerous individual reports of topical hypotensive drug benefit. The author presents the case from personal clinical praxis and the effect of hypotensive drug. The presentation is followed with the discussion of current knowledge and recommendations that are based on individual evaluation. The largest investigation found was in Grippo's study on 60 patients (13 hypertensive and 47 normotensive eyes). Therefore this is not a general therapeutic protocol, just individually based potential recommendation.

2. Case presentation

A young female (20 years old), without prior ophthalmological or systemic disease, was referred to the ophthalmologist at the eye clinic due to changes in optic nerve of her left eye. Clinical examination showed normal visual acuity (Snellen chart) and anterior segment:

VOD = cc −2.0 DSph = 1.0; VOS = cc −2.50 DSph = 0.9–1.0; and TOU = 14–18 mmHg.

CCT OU = 616 μm (−5 mmHg factor for correction).

Indirect ophthalmoscopy of the left eye showed unclear ONH borders, globular appearance, more prominent nasally (**Figure 1**).

In further observation, we performed echosonography (ultrasound A/B scanner UD-6000, Tomey) and B-scan OU: oval ONH changes of high reflectivity. FAG: the time of the arm-retina was regular. Blood vessels are completely and on time filled with contrast. Diffuse, oval, partially confluent changes with autofluorescence of the optic nerve head in the left eye (**Figures 2** and **3**).

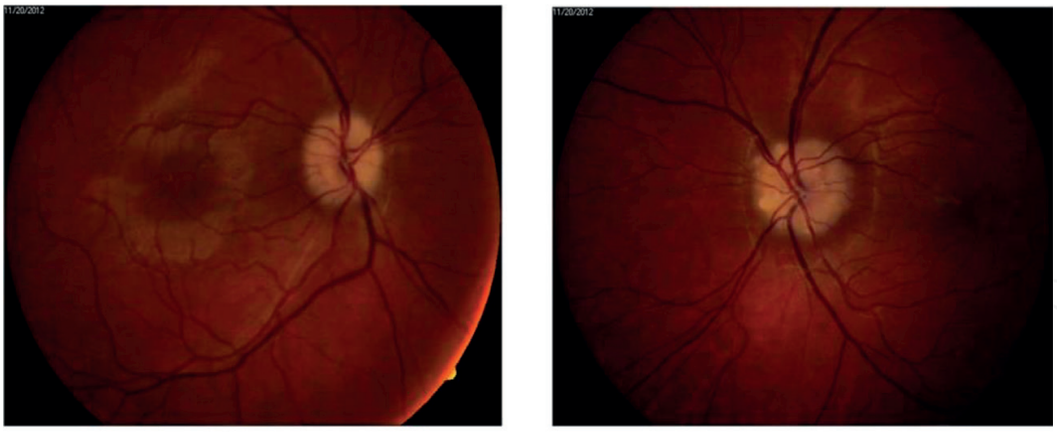

Figure 1. Photofundus OD (A) and OS (B): an unclear ONH border with superficial drusen.

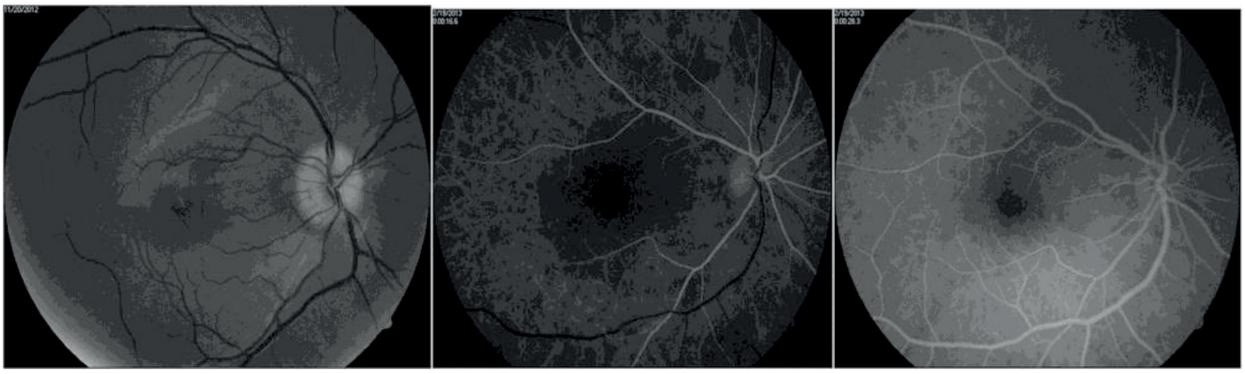

Figure 2. FAG OD: finding normal.

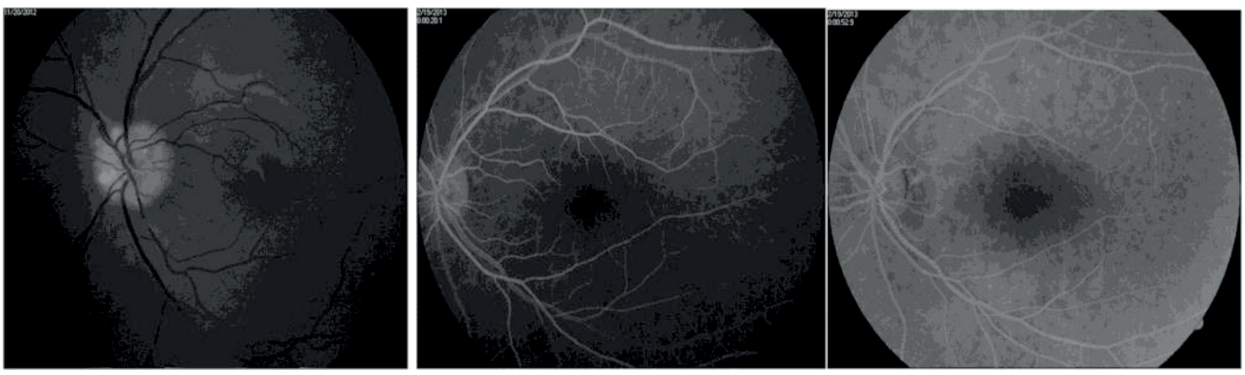

Figure 3. FAG OS: confluent autofluorescent ONH changes.

A standard automatic perimetry (Humphrey visual field analyzer, threshold test 30-2; 24-2) was performed, and scotoma progression in the visual field of both eyes was observed in four successive testing over 1 year. The most frequent defect is confluent, relative, and absolute paracentral scotoma, up to the central 15° (in the right eye; constriction of visual field) and inferior nasal quadrantanopia (left eye). After that finding, topical hypotensive therapy (brimonidine 0.2%, which was replaced by latanoprost 0.005% after 3 months due to allergic reaction) was applied (**Figures 4–7**).

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HRT detects marginal thinning in the nasal sector of the left eye (**Figure 8**).

RNFL examination OCT (Stratus optical coherence tomography); Carl Zeiss Meditec has detected sectorial thinning of both eyes (**Figure 9**).

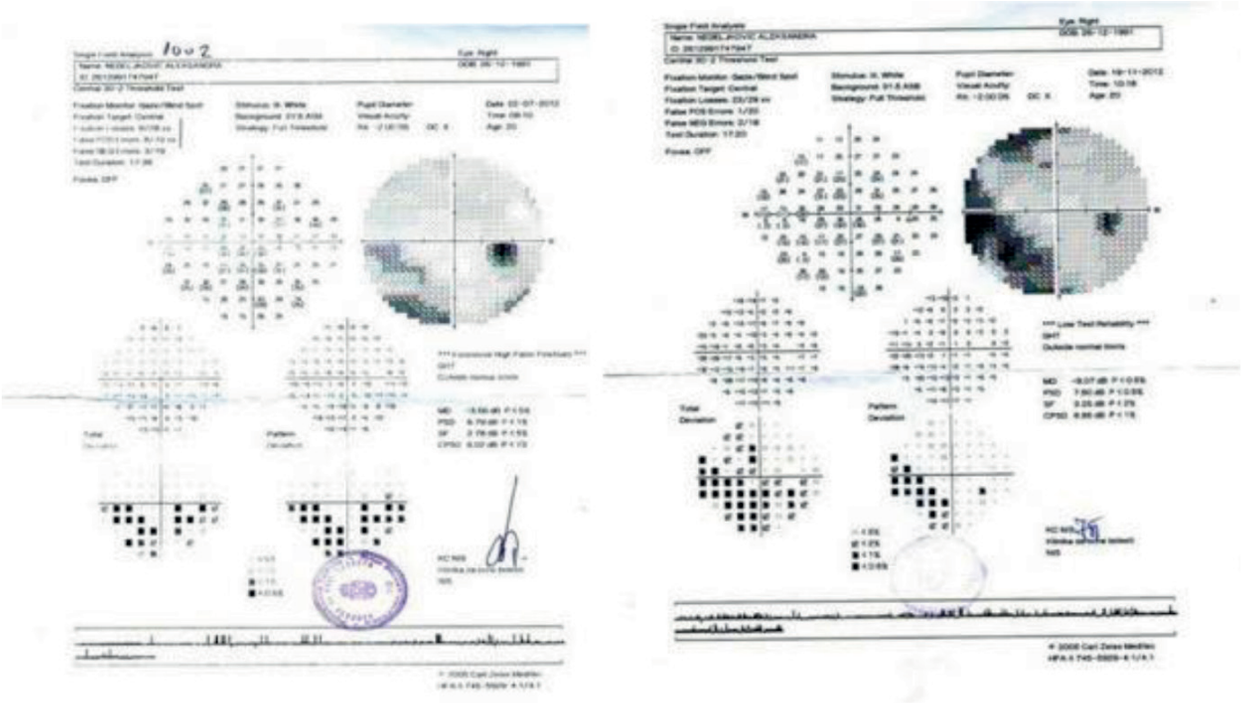

Figure 4. SAP of the right eye (VII/XI 2012) OD: relative paracentral visual field defects (MD = −9.07 dB; CPSD = 6.65 dB).

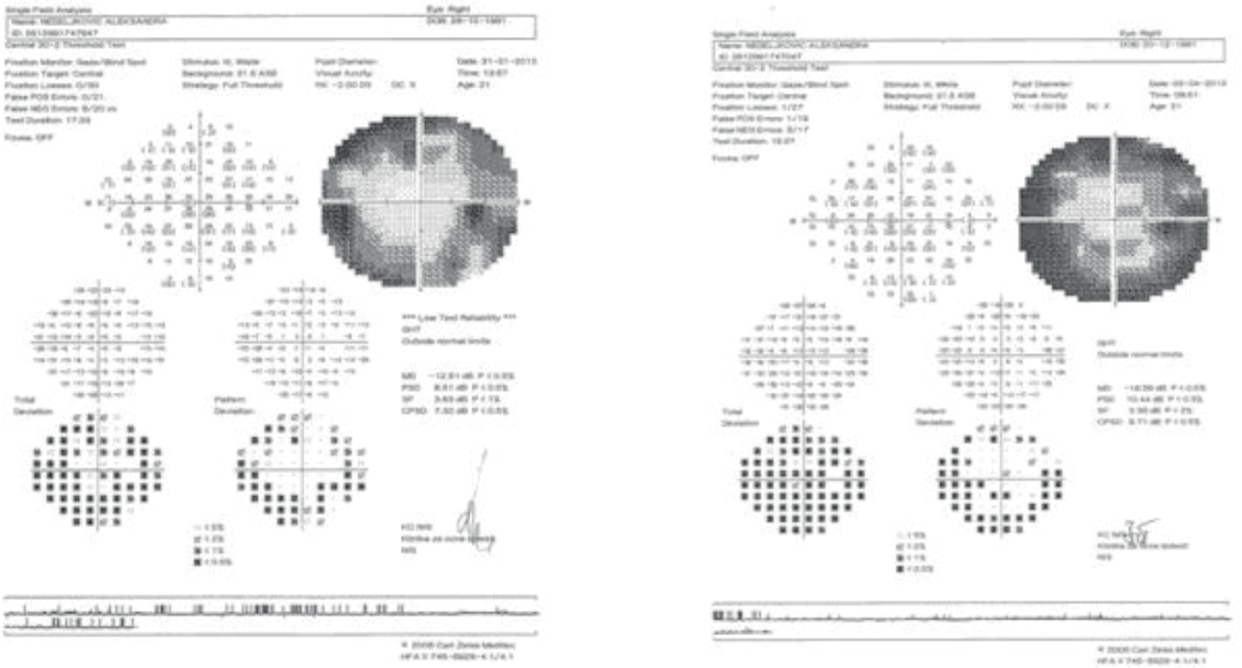

Figure 5. SAP of the right eye (I/IV 2013) OD: constriction of the visual field (MD = −18.56 dB; CPSD = 9.71 dB).

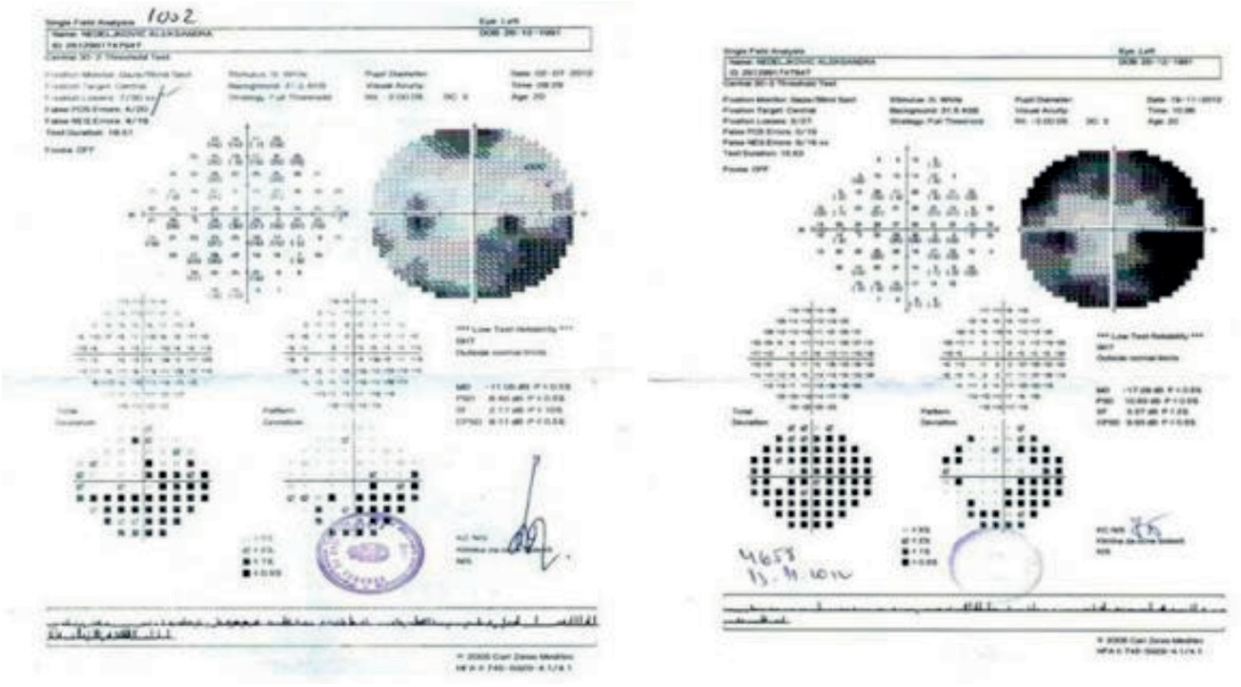

Figure 6. SAP of the left eye (VII/XI 2012) OS: blind spot enlargement, relative and absolute paracentral visual field defects (MD = −17.28 dB; CPSD = 9.93 dB).

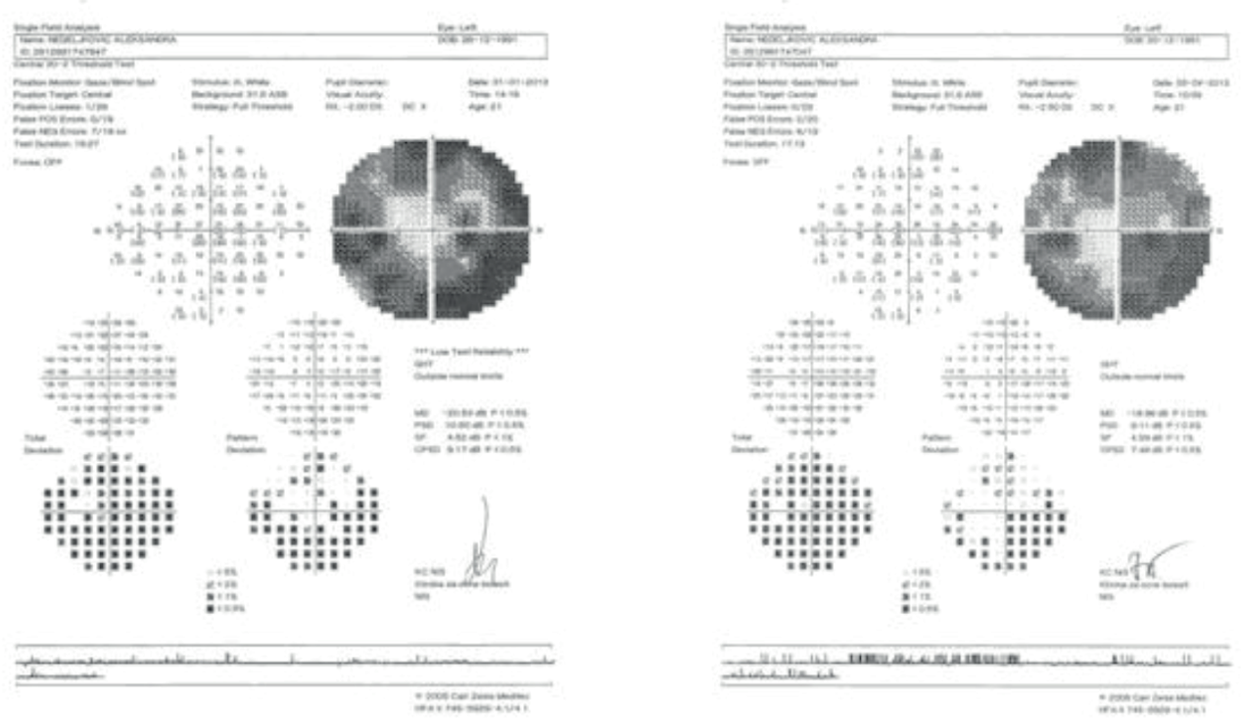

Figure 7. SAP of the left eye (I/IV 2013) OS: narrowing of the visual field up to 10°, inferior nasal quadrantanopia (MD = −18.96 dB; CPSD = 7.48 dB).

The electrophysiological examination recorded regular answers of parameters, prolonged latency of the P100 wave in the right eye (OD: P100 = 117 ms; OS: P100 = 113.7 ms).

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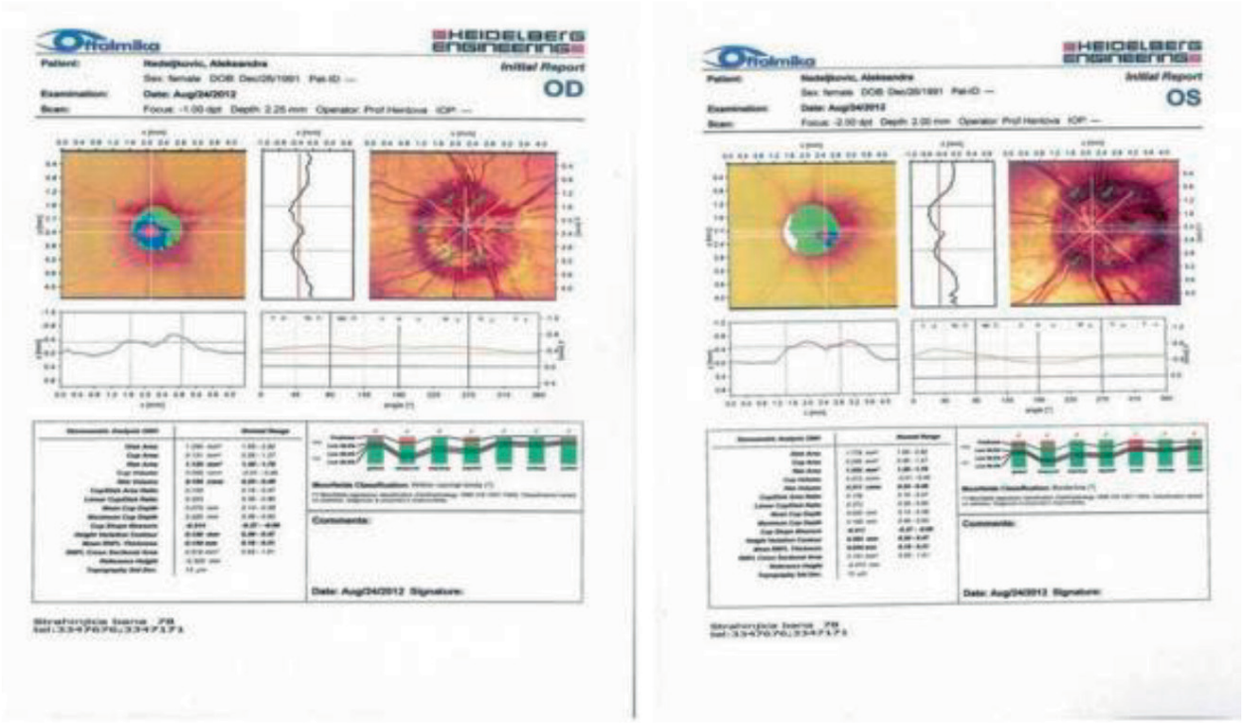

Figure 8. HRT (VIII/2012) OD: DA = 1256; mean RNFL = 0.130; OS: DA = 1778; mean RNFL = 0.034.

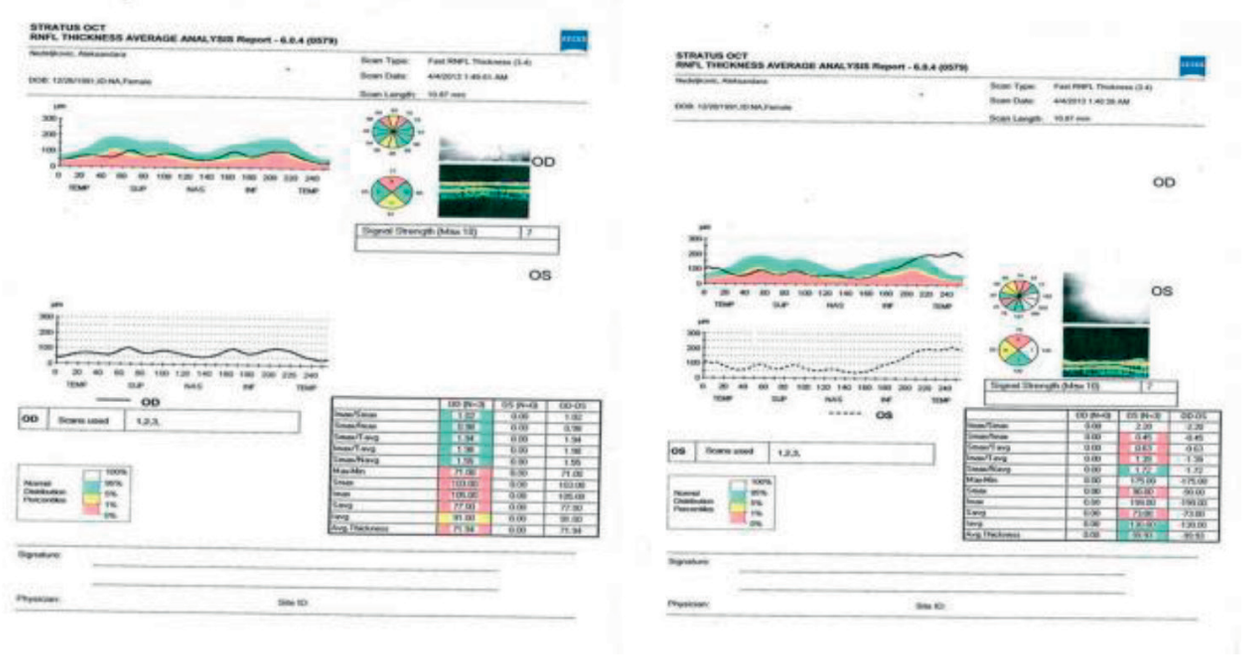

Figure 9. OCT (stratus 6.0.4. IV/2013) RNFL OD: Savg = 77.0, Iavg = 91.0, Navg = 66.0, and Avg thick = 71.94. RNFL OS: Savg = 73.0, Iavg = 130.0, Navg = 53.0, and Avg thick = 93.93.

Computerized tomography (CT) of the endocranium and the orbit on an axial scan showed bilateral calcification at the ONH (**Figure 10**).

Six months after topical hypotensive drug administration, an improvement of the visual field resulted in the improvement of reduced sensitivity. Depth and extensions of the scotomas were improved and maintained during the monitoring period of at least 3 years. The finding was also confirmed by the last visual field testing (**Figures 11** and **12**).

Relative and absolute paracentral and peripheral scotomas are still present, partly confluent, more in the nasal periphery of the left eye.

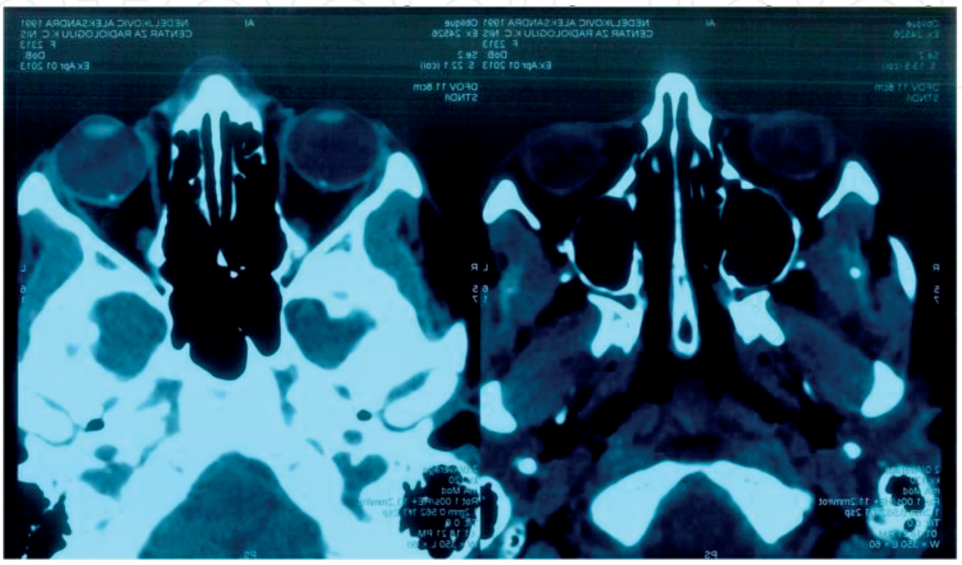

Figure 10. CT (IV/2013) No pathological changes in the endocranium. Bilateral calcification at the ONH.

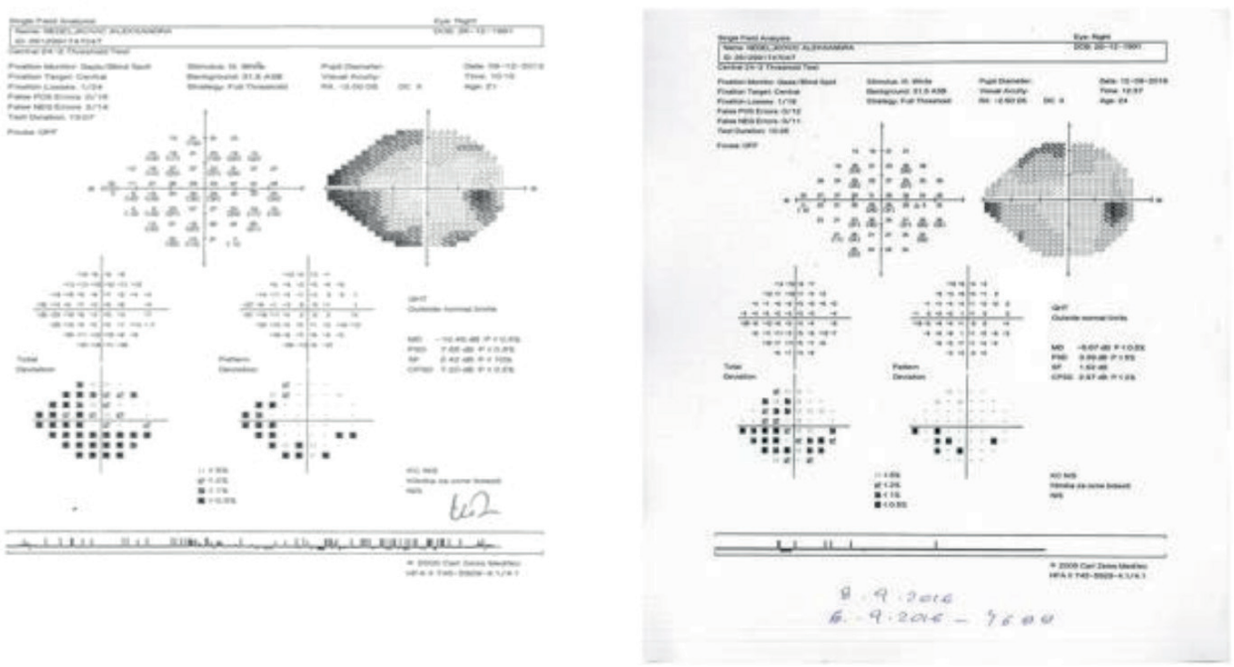

Figure 11. SAP of the right eye after therapy introduction: KP(XII/2013) OD: MD = −10.46 dB; CPSD = 7.20 dB; KP (IX/2016) OD (MD = −6.67 dB; CPSD = 2.97 dB).

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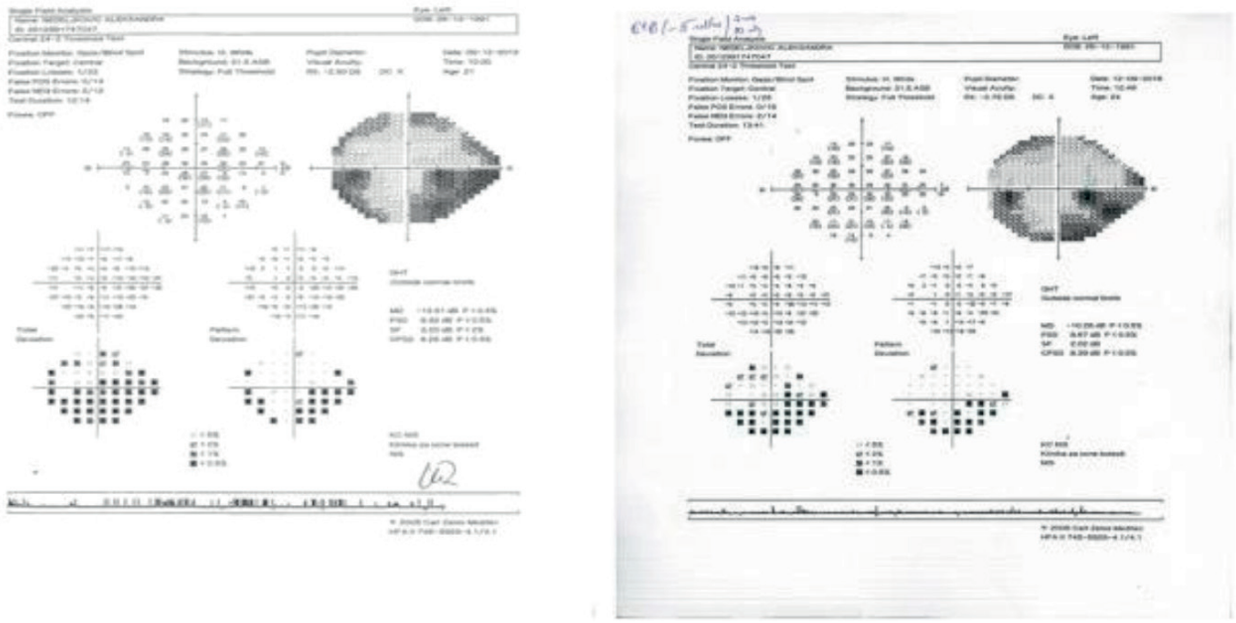

Figure 12. KP of the left eye after therapy introduction: KP(XII/2013) OS: MD = −13,51 dB; CPSD = 8,25 dB); KP (IX/2016) OS (MD = −10.28 dB; CPSD = 8.39 dB).

3. Discussion

A young female patient with advanced visual field defects because of the ONH drusen is presented. Visual field defect is one of the most common drusen complications in these cases with preserved central visual acuity. Although a patient has myopic refraction, it was concluded that the disk area is smaller (based on HRT). That was more pronounced on the right eye (OD: DA = 1.256 mm^2; OS: DA = 1.778 mm^2) when compared to the normative values of 1.69–2.82 mm^2. That indicated how a small diameter of the scleral canal can compromise axoplasmic transport. The conclusion of one of the conducted studies suggested that the incidence of drusen is greater if DA is smaller than 1.79 mm^2, with a confidence interval of $p < 0.001$. There is also confirmation that drusen are present in more than 50% of patients with a small horizontal diameter of the optic nerve head (1.68 ± 0.18 mm). Different authors agreed that the chronic obstruction of axoplasmic transport due to the small diameter of the scleral canal results in drusen formation [10, 26].

There are three basic theories in explaining pathogenesis of drusen:

1. Degenerative axonal death (Seitz)
2. Protein transudation from congenitally abnormal vessels of the disk (Sacks)
3. Alteration of axoplasmic transport (Spenser)

The slowly progressive visual field loss is characteristic of the drusen (Lauber) as a result of a direct mechanical compression followed by axonal dysfunction and degeneration. The

incidence of scotomas, as one of the most common complications of optic nerve drusen, is 24–87% in adults with a progression rate of 1.6% per year. Visual field defect may occur even in childhood [11, 27]. The rule in appearance, depth, duration, and the type of scotoma was not determined. The most common are peripheral, arcuate scotoma, enlargement of the blind spot, rarely the nasal step, and generalized constriction of the visual field. Scotomas are variable depending on the grade of drusen. In severe cases, they become confluent to inferonasal quadrantanopia to hemianopia [27, 28]. In the presented clinical case, an irregular incidence of scotomas was observed in relation to the grade of drusen (grade I in the right eye; grade II in the left eye). Savino et al. founded 71% of the visual field defects for visible drusen, compared to 21% in the case of hidden ones [14].

An attempt was made to correlate distribution of scotoma with localization and number of optic nerve drusen. In Grippo's retrospective study in 103 eyes was found that in superficial drusen (grade III), visual field defect was advanced, while in the case of hidden drusen (grade I), it was the initial, which was confirmed through this presentation. The distribution of scotoma often does not correspond completely with the localization of visible drusen, most likely due to the simultaneous presence of hidden ones and extensive axonal lesions [11]. Thus, there are scotomas with irregular distribution on the right eye of our patient with hidden drusen. Some of the reports noted 76.3% of scotomas in visible drusen compared to 46.5% in the case of hidden drusen [29, 30]. In the remaining half of patients with the first (I) grade of the drusen, in 5% of cases, an arcuate scotoma could be developed [31].

Any decrease in central visual acuity, rapidly progressive visual field loss, or arcuate scotoma in drusen requires careful consideration of the etiology in terms of glaucoma or neuro-ophthalmology background [11]. Concomitant drusen and glaucoma can cause extensive visual field damage, which can cause a diagnostic and therapeutic dilemma. It was found that the grade of drusen and IOP level directly, independently, and significantly correlate with the degree of damage of the visual field. The result is a unique attitude of introducing topical hypotensive therapy in both cases in the aim to reduce the risk of further damage [1, 16, 32].

Visual field damage occurs in 90.9% of hypertensive versus 66.7% of normotensive eyes. Higher IOP is statistically significantly associated with advanced impairments regardless of age, sex, and grade of drusen [11, 15]. In our case presentation of a young female patient, elevated IOP or association with glaucoma for the entire duration of follow-up period was not confirmed, which was confirmed by testing. Flame microhemorrhages that may occur at the RNFL in glaucoma have been observed, but not found. Hemorrhages are thought to have no effect on the occurrence of scotomas in drusen, opposed to numerous and confluent flame-shaped hemorrhages in the real existing edema of the optic nerve head. There are no major controlled clinical studies of the effects of topical hypotensive drugs in drusen, but there are numerous individual reports, as well as announcements of small sample of patients confirming the benefit of topical hypotensive drugs due to indirect enhancement of ONH perfusion (Grippo, Poda-Wilczek, Spalding, Patel, Czajor, Moris) [1, 4, 15, 18, 33–35].

Due to the extension and depth of the scotoma, as well as their progression over the monitoring period of 6 months, although the patient did not have an elevated IOP, after examination of the literature and actual reports, topical hypotensive therapy was applied. The first therapeutic choice was brimonidine 0.2% for 3 months, but after an allergic reaction, it was

replaced with latanoprost 0.005% (due to patient age and frequency of drug application). There is no specific and precise recommendation about therapeutic choice. Most commonly carboanhydrase inhibitors are used.

In these cases, the decision to use medication was made after a regular CT and the exclusion of a potential neuro-ophthalmological disease (pseudotumor cerebri) and SAP findings (right eye, MD = −18.56 dB; left eye, MD = −18.96 dB). After 6 months, the depth of visual field defects was reduced (right eye, MD = −10.46 dB; left eye, MD = −13.51 dB).

An improvement in sensitivity reduction has been achieved, from the advanced to the initial (right eye)/moderate (left eye) stage, and was confirmed by the 1-year SAP and at the last SAP finding (IX/2016) (right eye, MD = −6,67 dB; left eye, MD = 10,28 dB).

In addition to functional, it is advisable to monitor structural damage by imaging techniques. Retinal nerve fiber layer (RNFL) thinning is a pathological finding detected in drusen, by using modern technologies: OCT (EDI-OCT), HRT, and laser scanning polarimetry (GDx) [9]. Roh et al. suggested OCT as a sensitive and confident method for the early detection of RNFL thinning in drusen and/or glaucoma [36, 37].

A combination of methods that confirm anatomical lesion (OCT) and functional impairment (SAP, electrophysiological testing) is necessary in making a right decision when and how to treat by topical hypotensive drugs [4]. Recent studies confirmed that OCT is a significant method in the differentiation of ONH drusen from the real edema of the ONH by means of qualitative and quantitative criteria [10]. Both entities are presented with elevation and unclear border of the ONH, and the qualitative criterion helps in the differentiation with a sensitivity of 63%, because in real edema, an internal contour line of the optic nerve is a straight line as well as the widened subretinal hyporeflective space (SHYPS), unlike in pseudoedema with the undulating line ("lumpy-bumpy" line) and with narrow SHYPS (Savini). The thickness of subretinal hyporeflective space (SHYPS) in edema is greater than 464 μm and in a diameter of 2 mm greater than 127 μm. Using EDI-OCT, it is possible to find out hidden drusen and their posterior border with a depth of 500–800 μm, deeper than a conventional OCT [38, 39].

The quantitative parameter is the measurement of RNFL thickness (total and sectoral) with a sensitivity of 90%, with a significant thinning before occurrence of scotoma in the visual field. RNFL thinning correlates with the scotoma location, but not always with the degree of damage of the visual field. The nasal sector is the predilection area [5]. Observed according to grades, I and II are dominant superiorly, while III is more frequent inferiorly [36]. Opposite to this, in ONH edema, the thickening of RNFL is observed [38]. The OCT findings of our patient show average and sectoral thinning, at superior sector in an eye with hidden drusen and nasally with superficial drusen, that correlates with previously mentioned observations. OCT angiography, with superficial laminar segmentation, usually shows focal capillary attenuation overlying the most prominent drusen. These findings demonstrate alterations in the superficial retinal capillary network associated with ONH drusen [40].

Heidelberg retinal tomography (HRT) provides three-dimensional topographic analysis of the ONH with the measurement of the height of the peripapillary RNFL that correlates with the changes in the visual field. Thus, the area of the ONH was analyzed, and the control HRT of our patient showed total RNFL thinning in the left eye 0.107 mm (0.18–0.31) [40, 41]. HRT

is less sensitive than OCT in the RNFL estimation. It also shows anterior displacement of the cup bottom, in front of the RPE versus healthy subjects [40]. Polarimetric analysis of retinal nerve fibers (GDx) usually detects thinning, especially in the eyes with irregular and unclear ONH border and erased cupping [41–44]. In our patient it was not performed due to technical impossibility.

The retinal ganglion cell axons in the early phase of mechanical stress do not show immediate degeneration but enter the stage of functional disorder, presented with abnormal latency at the visual evoked potential (VEP) in more than 95% (Stevens). A minor clinical study of six patients with drusen but without symptoms showed prolonged P100 wave latency [11, 45]. More sensitive is the multifocal VEP (mf VEP) that allows the measurement of the latency of local VEP response from 60 sectors within the central 24° of visual field, which may be less evident on the standard VEP [46]. Grippo's analysis of patients with the drusen shows an altered finding in 28% of patients tested with conventional VEP and 70% of abnormal findings with mf VEP [15]. A careful review of the evoked potential of our patient shows a discretely prolonged P100 wave, more obvious in the right eye (right eye, 117 ms; left eye, 113.7 ms). That is a sign of initial dysfunction with normal visual acuity. In more affected eye, RAPD or reduction of amplitude N95 can be seen [9].

Vascular complications and abnormalities in patients with drusen are frequent also. They are caused by different mechanisms if they are associated with optic nerve head drusen. The most common cause of sudden visual loss may be non-arteritic anterior ischemic optic neuropathy (NAINO). Auw-Haedrich points out that people with drusen are more prone to ischemic attacks of the optic nerve head than those with a small diameter of the scleral canal and without drusen, because the arterial inflow becomes insufficient with their magnification. The younger age is more dominant among them in comparison to the years of life of patients with NAINO. CRAO, CRVO, and subretinal neovascularization adjacent to optic nerve head may also affect the younger population [2, 9]. Our patient did not have vascular complications because of the shorter duration of detected drusen and her age.

4. Conclusion

Young person with optic nerve head drusen is presented, the most common complications and performed findings of the diagnostic procedures that were carried out with the summarized current knowledge of epidemiology, pathogenesis, clinical presentation, diagnosis, and therapy of the eyes with optic nerve head drusen. Currently, there is no specific strictly indicated treatment and therapeutic protocol for the progressive damage of visual function and complications that may arise due to drusen. A preventive IOP reduction is recommended in order to reduce potential damage of the axons of the optic nerve [1]. The goal of the applied therapeutic concept was to reduce the IOP with the tendency of slowing down and stopping the progression of scotoma in the visual field, which is confirmed by the findings in this presentation and numerous pilot studies [1, 2]. Topical hypotensive drugs improve blood flow in the optic nerve head, thus reducing potential vascular damage due to the presence of drusen.

Following the correlation of years of life, the morphology of the optic nerve head, and the functional changes with drusen, it has been established that during the time, drusen can cause serious progressive optic neuropathy [47, 48]. The structural characteristics in the optic nerve head cause individual differences in the damages of retinal ganglion cell axons caused by intraocular pressure; therefore, lower IOP is recommended [49]. The importance of this, as well as other medical and surgical therapeutic modalities, especially neuroprotective drugs, is still being investigated in order to reduce potential morbidity.

Conflict of interest

The author declares no conflict of interest.

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References

[1] Morris RW, Ellerbrock JM, Hamp AM, Joy JT, Roels P, Davis CN Jr. Advanced visual field loss secondary to optic nerve head drusen: Case report and literature review. Optometry. 2009;**80**(2):83-100. DOI: 10.1016/j.optm.2008.11.004

[2] Schargus M, Gramer E. Optic disc drusen. Der Ophthalmologe. 2008;**105**(7):693-710. DOI: 10.1007/s00347-008-1762-7

[3] Auw-Haedrich C, Staubach F, Witschel H. Optic disk drusen. Survey of Ophthalmology. 2002;**47**(6):515-532. PMID: 12504737

[4] Czajor K, Misiuk-Hojło M. Heidelberg edge perimetry in optic nerve drusen—A case report. MEDtube Science. 2014;**II**(1):29-33

[5] Ubhi P, Shechtman D, Green K. Discern optic nerve head Drusen from true papilledema. Review of Optometry. 15 Dec 2015;**2015**:44-47

[6] Friedman AH, Henkind P, Gartner S. Drusen of the optic disc. A histopathological study. Transactions of the Ophthalmological Societies of the United Kingdom. 1975;**95**(1):4-9. PMID: 1064209

[7] Chang MY, Pineles SL. Optic disk drusen in children. Survey of Ophthalmology. 2016;**61**(6):745-758. DOI: 10.1016/j.survophthal.2016.03.007

[8] Gay D, Boyer S. Two differing presentations of optic nerve head drusen. Optometry. 2001;**72**(9):588-596. PMID: 11575696

[9] Davis PL, Jay WM. Optic nerve head drusen. Seminars in Ophthalmology. 2003;**18**(4):222-242. DOI: 10.1080/08820530390895244

[10] Gili P, Flores-Rodríguez P, Yangüela J, Orduña-Azcona J, Martín-Ríos M. Evaluation of optic disc size in patients with optic nerve head drusen using fundus photography. Journal of Optometry. 2013;**6**(2):75-79

[11] Grippo TM, Rogers SW, Tsai JC. Optic disc drusen. Glaucoma Today. Jan/Feb 2012;**2012**:19-24

[12] Gaillard F. Optic Disc Drusen. 2005-2017. Available from: http://radiopaedia.org

[13] Aumiller MS. Optic disc drusen: Complications and management. Optometry. 2007;**78**(1): 10-16. DOI: 10.1016/j.optm.2006.07.009

[14] Jonas JB, Gusek GC, Guggenmoos Holzmann I, Naumann GO. Pseudopapilledema associated with abnormally small optic discs. Acta Ophthalmologica. 1988;**66**(2):190-193. PMID: 3389093

[15] Grippo TM, Shihadeh WA, Schargus M, Gramer E, Tello C, et al. Optic nerve head drusen and visual field loss in normotensive and hypertensive eyes. Journal of Glaucoma. 2008;**17**:100-104. DOI: 10.1097/IJG.0b013e31814b995a

[16] Kamjoo S, Epley KD, Pihlbad MS. Optic Disc Drusen. 2015. Available from: http://eyewiki.aao.org

[17] Gossman MV. Pseudopapilledema. 2015. Available from: http://emedicine.medscape.com

[18] Patel V, Oetting TA. Optic Nerve Drusen: 19-year-old Female with Blurred Vision. EyeRounds.org. 2007. Available from: http://www.EyeRounds.org/cases

[19] Sadun AA, Wang MY. Funduscopic features. In: Handbook of Clinical Neurology. Vol. 102. New York, United States: Elsevier; 2011. pp. 2-524

[20] Lyons CJ, Wiwatwongwana A. Pediatric neurology part III. The optic nerve and visual pathways. In: Handbook of Clinical Neurology. Vol. 113. New York, United States: Elsevier; 2013. pp. 1515-1525

[21] Delyfer MN, Rougier MB, Fourmaux E, Cousin P, Korobelnik JF. Laser photocoagulation for choroidal neovascular membrane associated with optic disc drusen. Acta Ophthalmologica Scandinavica. 2004;**82**(2):236-238. DOI: 10.1111/j.1600-0420.2004.00231.x

[22] Haritoglou C, Prieglinger SG, Grueterich M, Kampik A, Kriegelstein GK. Radial optic neurotomy for the treatment of acute functional impairment associated with optic

nerve drusen. The British Journal of Ophthalmology. 2005;**89**(6):779-780. DOI: 10.1136/bjo.2004.060335.

[23] Pfriem M, Hoerauf H. Unsuccessful surgical excision of optic nerve drusen. Graefe's Archive for Clinical and Experimental Ophthalmology. 2011;**249**(10):1583-1585. DOI: 10.1007/s00417-011-1693-x

[24] Kapur R, Pulido JS, Abraham JL, Sharma M, Buerk B, Edward DP. Histologic findings after surgical excision of optic nerve head drusen. Retina. 2008;**28**(1):143-146. DOI: 10.1097/IAE.0b013e31815e98d8

[25] Pinxten I, Stalmans P. Radial optic neurotomy as a treatment for anterior ischemic optic neuropathy secondary to optic disc drusen. GMS Ophthalmology Cases. 2014. DOI: 10.3205/oc000018

[26] Jonas JB, Gusek GC, Guggenmoos-Holzmann I, Naumann GO. Optic nerve head drusen associated with abnormally small optic discs. International Ophthalmology. 1987;**11**(2):79-82. PMID: 2451648

[27] Wilkins J, Pomeranz H. Visual manifestations of visible and buried optic disc drusen. Journal of Neuro-Ophthalmology. 2004;**24**(2):125-129. PMID: 15179065

[28] McCafferty B, McClelland CM, Lee MS. The diagnostic challenge of evaluating papill-edema in the pediatric patient. Taiwan Journal of Ophthalmology. 2017;**7**(1):15-21. DOI: 10.4103/tjo.tjo_17_17

[29] Calvo-González C, Santos-Bueso E, Díaz-Valle D, Reche-Frutos J, Moriche-Carretero M, Benítez-Del-Castillo JM, et al. Optic nerve drusen and deep visual fields defects. Archivos de la Sociedad Española de Oftalmología. 2006;**81**(5):269-273. PMID: 16752318

[30] Obuchowska I, Mariak Z. Visual field defects in the optic disc drusen. Klinika Oczna. 2008;**110**(10-12):357-360. PMID: 19195165

[31] Katz BJ, Pomeranz HD. Visual field defects and retinal nerve fiber layer defects in eyes with buried optic nerve drusen. American Journal of Ophthalmology. 2006;**141**(2):248-253. DOI: 10.1016/j.ajo.2005.09.029

[32] Spalding JM. Visual-field loss with optic nerve drusen and ocular hypertension: A case report. Optometry. 2002;**73**(1):24-32. PMID: 12363235

[33] Sowka J, Kabat A. IOP, drusen and occlusion. Review of Optometry. 2016;**31**:118-119

[34] Pojda-Wilczek D, Herba E, Jedrzejewski W, Pojda SM. Optic disc drusen and glaucoma—Case report. Klinika Oczna. 2004;**106**(1-2 Suppl):263-265. PMID: 15510520

[35] Ziak P, Jarabáková K, Koyšová M. Optic disc drusen—Current diagnostic possibilities. Ceská a Slovenská Oftalmologie. 2014;**70**(1):30-35. PMID: 24862373

[36] Roh S, Noecker RJ, Schuman JS. Evaluation of coexisting optic nerve head drusen and glaucoma with optical coherence tomography. Ophthalmology. 1997;**104**:1138-1144. PMID: 9224467

[37] Roh S, Noecker RJ, Schuman JS, Hedges TR, Weiter JJ, Mattox C. Effect of optic nerve head drusen on nerve fiber layer thickness. Ophthalmology. 1998;**105**:878-885. DOI: 10.1016/S0161-6420(98)95031-X

[38] Johnson LN, Diehl ML, Hamm CW, Sommerville DN, Petroski GF. Differentiating optic disc edema from optic nerve head drusen on optical coherence tomography. Archives of Ophthalmology. 2009;**127**(1):45-49. DOI: 10.1001/archophthalmol.2008.524

[39] Silverman AL, Tatham AJ, Medeiros FA, Weinreb RN. Assessment of optic nerve head drusen using enhanced depth imaging and swept source optical coherence tomography. Journal of Neuro-Ophthalmology. 2014;**34**(2):198-205

[40] Gaier ED, Rizzo JF, Miller JB, Cestari DM. Focal capillary dropout associated with optic disc drusen using optical coherence tomographic angiography. Journal of Neuro-Ophthalmology. 2017;**37**(4):405-410. DOI: 10.1097/WNO.0000000000000502

[41] Pihlbad MS. Optic Nerve Head Drusen. 2015. Available from: http://eyewiki.aao.org

[42] Kuchenbecker J, Wecke T, Vorwerk CK, Behrens-Baumann W. Quantitative and objective topometrical analysis of drusen of the optic nerve head with the Heidelberg retina tomograph (HRT). Der Ophthalmologe. 2002;**99**(10):768-773. DOI: 10.1007/s00347-002-0639-4.

[43] Patel NN, Shulman JP, Chin KJ, Finger PT. Optical coherence tomography/scanning laser ophthalmoscopy imaging of optic nerve head drusen. Ophthalmic Surgery, Lasers & Imaging. 2010;**41**(6):614-621. DOI: 10.3928/15428877-20100929-07

[44] Tatlipinar S, Kadayifcilar S, Bozkurt B, Gedik S, Karaagaoglu E, Orhan M, et al. Polarimetric nerve fiber analysis in patients with visible optic nerve head drusen. Journal of Neuro-Ophthalmology. 2001;**21**(4):245-249. PMID: 11756852

[45] Vieregge P, Rosengart A, Mehdorn E, Wessel K, Kömpf D. Drusen papilla with vision disorder and pathologic visual evoked potentials. Der Nervenarzt. 1990;**61**(6):364-368. PMID: 2377263

[46] Grippo TM, Ezon I, Kanadani FN, Wangsupadilok B, Tello C, Liebmann JM, et al. The effects of optic disc drusen on the latency of the pattern-reversal checkerboard and multifocal visual evoked potentials. Investigative Ophthalmology & Visual Science. 2009;**50**(9):4199-4204. DOI: 10.1167/iovs.08-2887

[47] Schargus M, Grippo T, Ritch R, Gramer E. Stage of visual field loss in relation to the appearance of the optic disc in 144 eyes of patients with drusen of the optic disc. Investigative Ophthalmology & Visual Science. 2005;**46**:2483

[48] Tanaka H, Shimada Y, Nakamura A, Tanikawa A, Horiguchi M. A case of bilateral optic nerve head drusen-induced inferior altitudinal hemianopsia. Neuro-Ophthalmology. 2015;**39**(4):201-206. DOI: 10.3109/01658107.2015.1022899

[49] El-Assal K, Tatham AJ. Rapidly progressing visual loss associated with optic nerve head drusen: Is there a role for lowering intraocular pressure? Journal of Ophthalmic Science. 2016;**1**(2):23-33. DOI: 10.14302/issn.2470-0436.jos-15-763