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Case report

Swelling of the optic nerve head due to the coexistence of idiopathic intracranial hypertension and optic nerve drusen

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Summary

Introduction. Optic nerve head drusen (ONHD) is one of the most common congenital disorders of the optic nerve with prevalence from 0.5 to 2% in general population. Idiopathic intracranial hypertension is a condition occurring in children, unlike adults, regardless of sex or the presence of obesity. Both conditions in children present on the eye as swelling of the optic disc, which is difficult to distinguish by clinical examination. They are extremely rarely associated.

Case report. A 15-year-old girl was urgently admitted due to decreased vision in both eyes for the past seven days. Computerized visual field (CVP) testing revealed annular scotoma in both eyes, with a residual of 10 degrees and decreased sensitivity. Fundus examination showed an edematous of both ONH. Echo B scan showed highly reflective changes corresponding to drusen in ONH. Acetazolamide 250 mg orally three times a day was introduced into the therapy. Neurological examination and MRI of endocranium were normal. After three weeks, VA in both eyes was 0.5 corrected with -0.50 Dsph to 1.0. ONH had significant regression with unclear boundaries temporally, superiorly and inferiorly. The follow-up period lasted over the following year, visual acuity was stable, and CVP was normal, only slightly unclear boundaries of the ONH were present.

Conclusion. The diagnosis of optic nerve drusen is possible in children with benign intracranial hypertension. For this reason, it is necessary to thoroughly diagnose the patient in order to start treatment in a timely manner and prevent the development of visual impairment.

Key words: optic disk drusen, idiopathic intracranial hypertension, benign intracranial hypertension, optic disc oedema, children

Introduction

Optic nerve head drusen (ONHD) is one of the most common congenital conditions of the optic nerve. The prevalence of optic nerve drusen in the general population is 0.5–2% [1].

These largely acellular calcifications, usually detected incidentally on an exam, may progress and become more visible during earlier decades of life but rarely change significantly over a patient's lifetime and are generally asymptomatic. Although most are asymptomatic, optic

nerve drusen have been associated with visual field defects, anterior ischemic optic neuropathy, transient visual obscurations, and more rare complications [2].

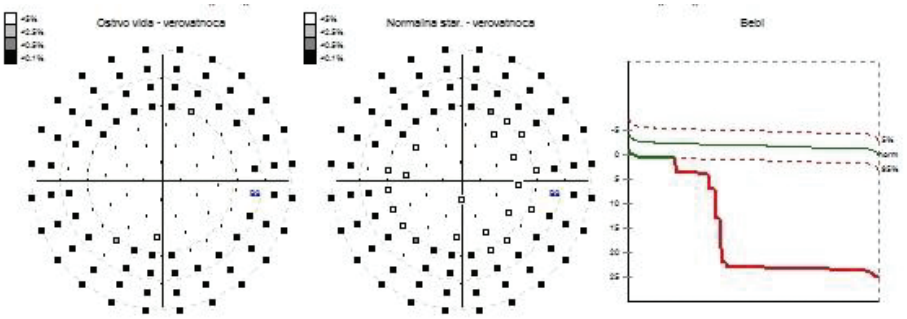
A variety of imaging modalities may be employed to evaluate the presence of ONHD, including ultrasound, optical coherence tomography (OCT), enhanced depth imaging-OCT, fluorescein angiography, fundus autofluorescence, and optical coherence tomography angiography [3].

Idiopathic intracranial hypertension (IIH) occurs in children, but unlike adults, it is not specific for sex and the presence of obesity. IIH is manifested on the fundus as swelling of the ONH.

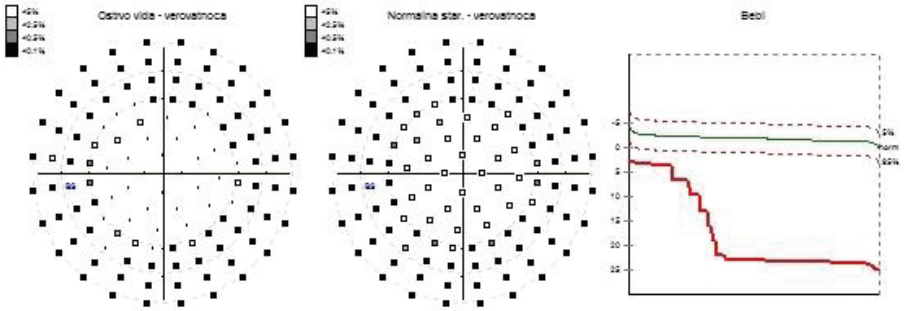
Acetazolamide is the mainstay of medical treatment, but some patients with significant visual loss may require surgical intervention [4].

Case report

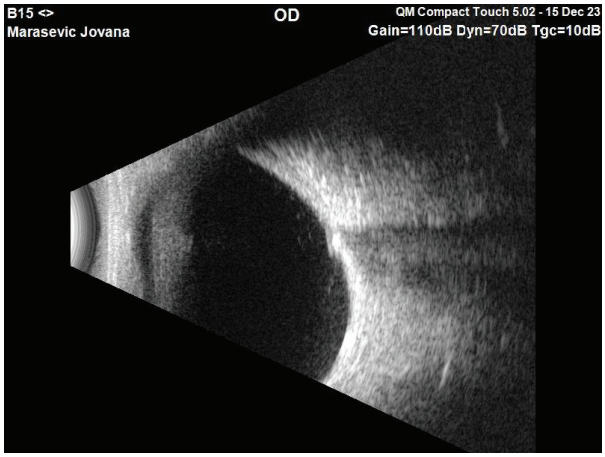
A 15-year-old girl was urgently admitted for an ophthalmological examination due to decreased vision in both eyes for the past seven days. Her previous history denied any systemic diseases. She had worn glasses for vision correction for a short time. The ophthalmological examination revealed the following: Visual acuity in the right eye was 0.3 according to Snellen, and in the left eye 0.2, without the possibility of correction. Intraocular pressure measured by Goldman applanation tonometry was 15 mmHg in both eyes. In the strabismus examination, there was orthoposition at near and far, regular motility and convergence, without double images and nystagmus. Color vision tested with Ishihara charts was 8/11 in both eyes. Examination under a biomicroscope revealed a completely normal anterior segment. The pupils were isochoric, circular, reacting promptly and abundantly to light, RAPD was negative. Computerized visual field testing revealed annular scotoma in both eyes with a residual of 10 degrees and decreased sensitivity. After mydriasis on two occasions with 1% tropicamide solution, an edematous optic nerve papilla was seen with normal findings in the macula, blood vessels, and periphery of the fundus. Refraction in cycloplegia for RE was 0.00 and LE +0.50 Dsph. Optical coherence tomography of the optic nerve head and macula was performed. The findings indicated the presence of ONH swelling, a decrease in the number of ganglion cells in the macula of the left eye, and preserved foveolar depression and macular stratification. Echo B scan was performed: in the PNO area there were highly reflective changes corresponding to drusen. In the CV area, there were low reflective opacities. The retina was lying. Acetazolamide 250 mg orally three times a day was introduced into the therapy. Neurological examination did not reveal any pathological symptoms or signs. MRI of endocranium did not reveal any pathological changes, except pronounced inflammatory changes in the right maxillary sinus, ethmoidal, sphenoidal, and right frontal sinuses. After four days of starting acetazolamide therapy, visual acuity in both eyes recovered and the symptoms decreased. Visual RE was 0.3 with -1.00 Dsph was 1.0 according to Snellen. LE was 0.4 with -1.00 Dsph 1.0. ONH edema was in regression. After three days, a computerized visual field was performed – and there was a completely normal finding. Examination of the fundus of the eye in mydriasis registered a significant improvement: ONH was with clear temporal boundaries, upper, lower and nasal edema was less than before. After three weeks, VA in both eyes was 0.5 corrected with -0.50 Dsph to 1.0. ONH had significant regression with unclear boundaries temporally, superiorly and inferiorly. Acetazolamide was recommended twice daily for seven days and once daily for seven days, after which it was excluded from therapy. During the follow-up period over the next year, visual acuity was stable, and findings in the visual field were normal, as were those in the anterior and posterior segments of the eye except slightly unclear boundaries of the ONH.



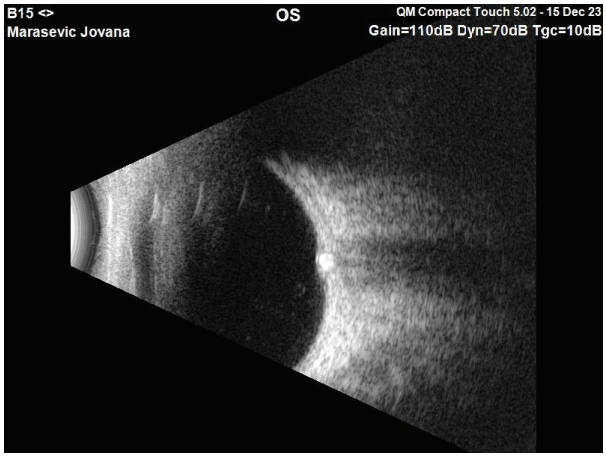
Picture 1. Computerized visual field of right eye before treatment



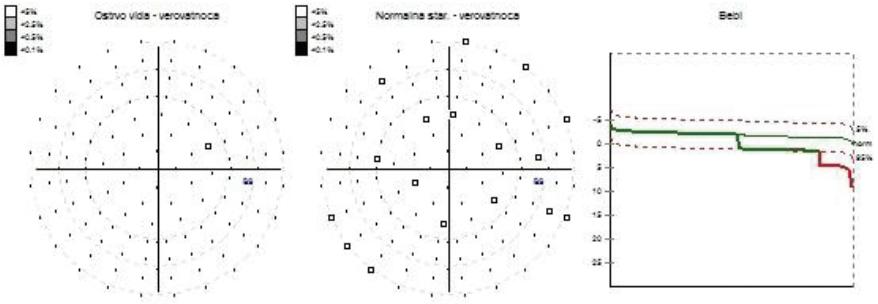
Picture 2. Computerized visual field of left eye before treatment



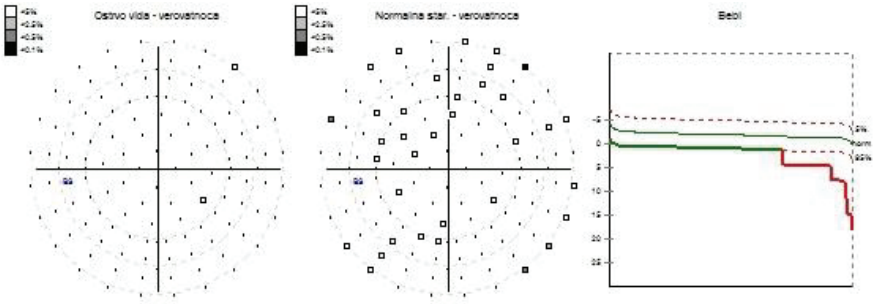
Picture 3. Ultrasonography - B scan RE



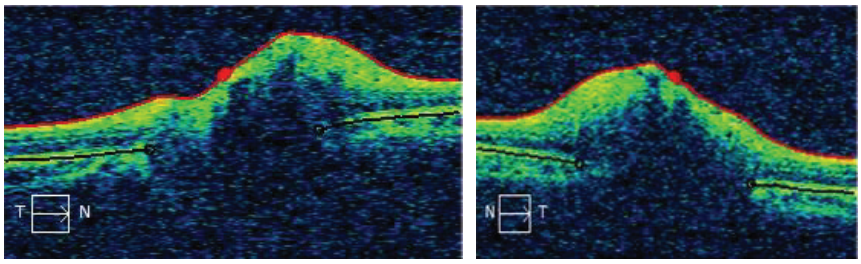
Picture 4. Ultrasonography - B scan LE



Picture 5. Computerized visual field of right eye after treatment



Picture 6. Computerized visual field of left eye after treatment



Picture 7. OCT ONH right and left eye

Discussion

Although both drusen and IIH cause the clinical picture of ONH edema, care should be taken when making the diagnosis. In the aforementioned patient, measurement of intracranial pressure was not possible due to the technical unavailability of the method. Ultrasound B-scan remains the most reliable method for demonstrating drusen, as OCT in many cases cannot distinguish drusen from edema of other etiology. Certainly, the findings of clinical and ultrasound measurements of edema at the beginning of treatment and at the end of follow-up indicate a difference in its size. The clinical course (sudden decrease in vision without refractive correction at the beginning, then complete recovery with acetazolamide) suggests a dynamic component – typical for IIH, and atypical for isolated drusen, which usually do not affect central visual acuity. In ONH drusen, visual acuity is stable and not subject to change, i.e. in almost all cases, drusen do not affect visual acuity. The rapid and complete regression of symptoms after acetazolamide in this case strongly supports IIH, with the drusen probably contributing to the clinical picture and prolonging the presence of mildly blurred papillary borders.

The diagnosis of optic nerve drusen is possible in children with benign intracranial hypertension. For this reason, it is necessary to thoroughly diagnose the patient in order to start treatment in a timely manner and prevent the development of visual impairment.

Conclusion

The specificity of this presentation lies in the extremely rare combination of idiopathic intracranial hypertension and optic disc drusen in a child without the usual risk factors, the diagnostic complexity due to the clinical picture mimicking papilledema, and the rapid and complete therapeutic response to acetazolamide. These elements emphasize the need for a multimodal diagnostic approach and early treatment.

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Ethical approval. The Ethics Committee of the Clinical Center of Montenegro, Podgorica, Montenegro, approved the study and informed consent was obtained from all individual respondents. The research was conducted according to the Declaration of Helsinki.

Conflicts of interest. The authors declare no conflict of in terest

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Edem glave optičkog nerva izazvan istovremenim prisustvom idiopatske intrakranijalne hipertenzije i drūza optičkog nerva

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Uvod. Drūze optičkog nerva (ONHD) predstavljaju jedan od najčešćih urođenih poremećaja optičkog nerva, sa prevalencijom 0,5–2% u opštoj populaciji. Idiopatska intrakranijalna hipertenzija je stanje koje se kod djece javlja, za razliku od odraslih, nezavisno od pola i prisustva gojaznosti. Oba stanja se kod djece na očima manifestuju kao otok optičkog diska, koji je kliničkim pregledom teško razlikovati. Njihova međusobna povezanost je izuzetno rijetka.

Prikaz bolesnika. Djevojčica stara 15 godina primljena je hitno zbog smanjenog vida na oba oka u trajanju od sedam dana. Kompjuterizovano ispitivanje vidnog polja (CVP) pokazalo je prstenasti skotom na oba oka, sa preostalim vidnim poljem od 10 stepeni i smanjenom osjetljivošću. Pregled očnog dna pokazao je edem oba optička diska. B-sken ultrazvuk pokazao je visoko reflektivne promjene koje odgovaraju drūzama u ONH. U terapiju je uveden acetazolamid 250 mg peroralno, tri puta dnevno. Neurološki pregled i magnetna rezonancija endokranijuma bili su uredni. Nakon tri nedjelje, vidna oštrina (VA) na oba oka bila je 0,5 korigovana sa -0,50 Dsph na 1,0. Zapažena je značajna regresija edema optičkog diska, sa nejasnim granicama temporalno, superiorno i inferiorno. Tokom jednogodišnjeg praćenja, vidna oštrina je ostala stabilna, CVP je bio uredan, a granice optičkog diska samo blago nejasne.

Zaključak. Dijagnoza drūza optičkog nerva moguća je i kod djece sa benignom intrakranijalnom hipertenzijom. Iz tog razloga je neophodno temeljno dijagnostikovati pacijenta kako bi se pravovremeno započelo liječenje i spriječio razvoj oštećenja vida.

Ključne riječi: drūze optičkog diska, idiopatska intrakranijalna hipertenzija, benigna intrakranijalna hipertenzija, edem optičkog diska, djeca