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Optic Disc Drusen Presenting with Binasal Hemianopia

Binazal Hemianopsi Prezentasyonlu Optik Disk Druzeni

Hasan Ali Tufan, Baran Gencer, Barış Kömür, Selçuk Kara, Betül Kızıldağ*, Fatma Uysal*

Çanakkale Onsekiz Mart University Medical Faculty, Department of Ophthalmology, Çanakkale, Turkey

*Çanakkale Onsekiz Mart University Medical Faculty, Department of Radiology, Çanakkale, Turkey

Summary

Optic disc drusen (ODD) are defined as a form of calcific degeneration of the optic nerve. Visual field defects are seen in nearly 90% of the cases. We report a 32-year-old woman who was referred to our clinic with the complaint of longstanding blurred vision and nasal visual field defect in her both eyes. The diagnosis of ODD was made on the basis of clinical appearance of the optic nerves and confirmed by ancillary tests. Binasal visual field defect was observed in the automated visual field test. In conclusion, ophthalmologists should consider ODD in the differential diagnosis of advanced visual field loss and binasal hemianopia. *(Turk J Ophthalmol 2013; 43: 371-3)*

Key Words: Optic disc drusen, visual field defect, binasal hemianopia

Özet

Optik disk druzeni (ODD), optic sinirin bir çeşit kalsifik dejenerasyonu şeklinde tanımlanmaktadır. Olguların %90'ında görme alan defekleri görülmektedir. Her iki gözde uzun zamandır görme bulanıklığı ve nasal görme alan defekti mevcudiyeti nedeniyle kliniğimize yönlendirilen 32 yaşındaki bayan hasta sunulmuştur. Optik sinirin klinik görünümü ve yardımcı testler ile ODD tanısı konulmuştur. Otomatik görme alan testinde binazal görme alan defekti saptanmıştır. Sonuç olarak, göz hekimleri ileri görme alan kaybı ve binazal hemianopi ayırıcı tanısında optic disk druzenini dikkate almalıdır. *(Turk J Ophthalmol 2013; 43: 371-3)*

Anahtar Kelimeler: Optik disk druzeni, görme alan defekti, binazal hemianopi

Introduction

Optic disc drusen (ODD) are congenital and developmental anomalies characterized by calcific degeneration of the optic nerve head. Even though the pathogenesis remains obscure, it is thought that alteration in the axoplasmic flow of ganglion cells is the possible underlying reason. ODD occur in 0.4% to 3.7% of the population. ODD are usually bilateral and asymmetric and seem to favor the female gender.[1] Most patients with ODD are asymptomatic and they are noticed incidentally in a routine ophthalmologic examination. Clinically, ODD may infrequently cause transient visual obscurations. However, visual field defects appear in nearly 90% of the cases, even before the drusen appear on the disc. The most commonly found visual field alterations are concentric constriction, enlargement of the blind spot, and arcuate scotomas.[2]

We report herein a case with bilateral advanced visual field loss secondary to ODD which may be confused with binasal hemianopia.

Case Report

A 32-year-old white woman was referred to the eye clinic with the complaint of long-standing blurred vision and nasal visual field defect in her both eyes. The patient's medical history was unremarkable. Her best-corrected visual acuity was 18/20 in the right eye and 20/20 in the left. She had normal colour vision and no relative afferent pupillary defect was present.

Address for Correspondence/Yazışma Adresi: Hasan Ali Tufan MD, Çanakkale Onsekiz Mart University, Department of Ophthalmology, Çanakkale, Turkey
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Slit lamp examination was unremarkable in both eyes (OU) with grade IV open angles. Intraocular pressure measured by applanation tonometry was 14 mmHg bilaterally at 09:40 AM. Dilated optic disc examination revealed some small and bright yellow deposits in the both nerves. Disc margins were irregular and slightly elevated bilaterally. Peripapillary atrophy was seen in the temporal region of both nerves. Macula, vessels and the peripheral retina were normal in both eyes. Fundus and optic nerve photography with autofluorescence was obtained with the prediagnosis of ODD (Figure 1). Both optic nerves showed autofluorescence, giving strong evidence of ODD. At this point, B-scan ultrasound was ordered to finalize the diagnosis of ODD. The presence of calcification was confirmed by ultrasonography (Figure 2).

An automated visual field test was performed to investigate the visual field defects that the patient complained of. A binasal visual field defect was observed in the visual field testing. An inferiotemporal shallow scotoma was also noted in the left eye (Figure 3). Further optic nerve evaluation was performed with optical coherence tomography, and the retinal nerve fiber layer thickness measurement, revealing advanced nerve fiber thinning, supported the visual field loss (Figure 4). An MRI of the brain and orbits was ordered to rule out any mass or neurologic pathology. The MRI scans showed no pathologic appearances.

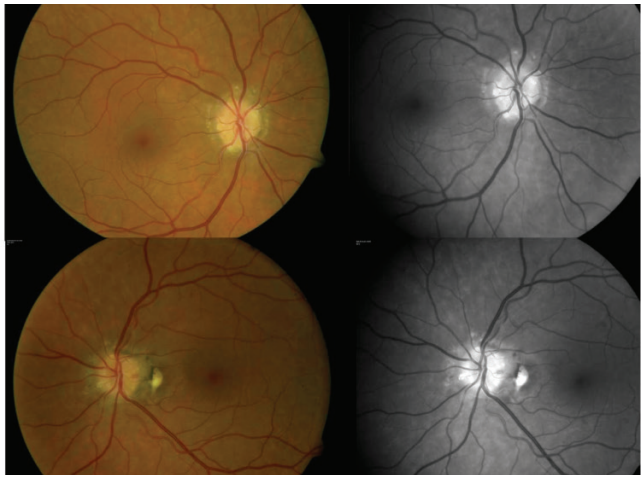

Figure 1. Fundus and red-free photographs of both eyes. Note the blurry margins and lumpy bumpy surfaces of the discs. Red-free photos reveal autofluorescence

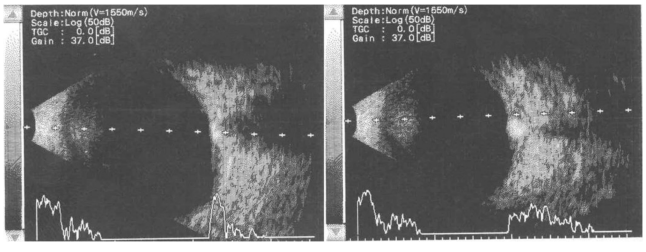

Figure 2. B-scan ultrasounds of both eyes. The optic nerve head have an area of hyperechoic reflections consistent with drusen

The diagnosis of ODD was made on the basis of clinical appearance of the optic nerves, which showed autofluorescence, the advanced visual field defects, and the normal MRI. The patient received prophylactic topical antiglaucomatous treatment and was suggested to be followed-up every 6 months with dilated fundus examinations, automated visual fields, and retinal nerve fiber analysis to monitor for progression.

Discussion

Visual field defects may arise due to ocular or neurologic pathologies. Different neurological difficulties cause characteristic forms of visual disturbances, including hemianopia and quadrantanopsias. Isolated loss of the nasal fields of vision is uncommon. Binasal hemianopsia, unlike types of visual field loss such as bitemporal heteronymous hemianopia, unilateral homonymous hemianopia, is not common and cannot be explained by a single visual tract lesion. It develops from bilateral involvement of the uncrossed fibers at the chiasma such as bilateral internal carotid artery atherosclerosis or aneurysm, olfactory groove meningioma, empty sella syndrome, chronic raised intracranial pressure, and neurosyphilis affecting both optic nerves.[3,4] Binasal hemianopia commonly occurs due to

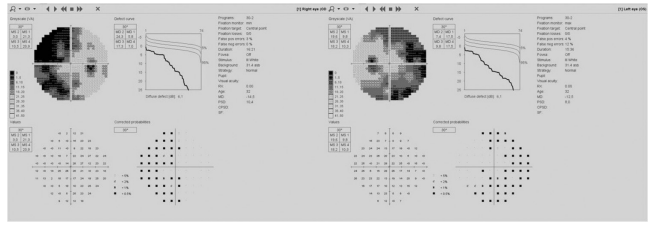

Figure 3. Automated perimetry shows binasal hemianopia and a shallow inferiotemporal scotoma in the left eye

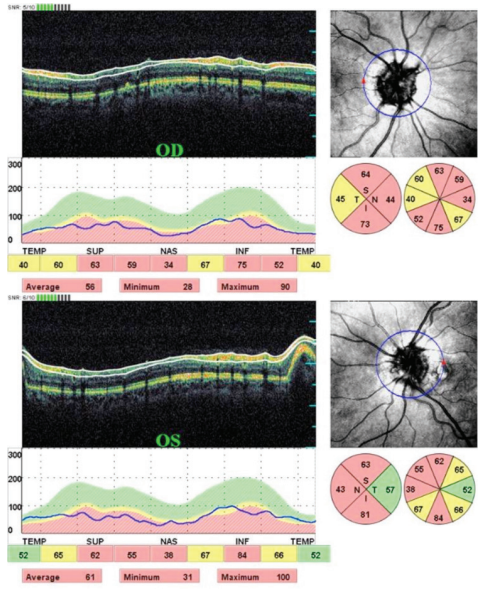

Figure 4. Optical coherence tomography shows advanced nerve fiber thinning in both eyes

retinal or optic nerve head pathologies. In an overview reported by Salinas-Garcia and Smith, it is stated that 75% of the binasal hemianopia cases originated from ocular pathologies.[5] Optic nerve pathologies presenting with binasal hemianopia were glaucoma (most common), ischemic optic neuropathy, congenital optic nerve pits, and optic nerve drusen in those patients.

Most large series report that 75% of patients with disc drusen have visual field defects.[6] The described ODD-associated field defects were nerve fiber bundle defects, arcuate defects, enlargement of the blind spot, and concentric narrowing. These defects were reported to affect the inferonasal quadrant most frequently.

Dramatic visual field loss in ODD is most often due to vascular complications, but it can also be caused by ODD alone when proposed pathogenetic mechanisms are considered.[7] These mechanisms may be described as: 1) impaired axonal transport in an eye with a small scleral canal leading to gradual attrition of optic nerve fibers, 2) direct compression of prelaminar nerve fibers by drusen, 3) ischemia within the optic nerve head. Although no effective treatment of ODD has been established, a prophylactic topical antiglaucomatous treatment was given to our patient as it had been suggested in the presence of visual field loss in the previous reports.[8]

A careful ocular examination must be done for all patients with binasal hemianopia. Although disc drusen are commonly diagnosed with ophthalmoscopy alone, their appearance may change throughout the patient's lifespan from buried to superficial in location; thus it might be confirmed by ancillary tests. The correct diagnosis is critical as they can mimic a number of serious pathologies presented with binasal hemianopia.

In conclusion, the ophthalmologists should consider ODD in the differential diagnosis of advanced visual field loss and binasal hemianopia.

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