

## CASE REPORT

# A Case of Bilateral Non-Arteritic Anterior Ischemic Optic Neuropathy with Bilateral Optic Nerve Atrophy in Coexistence of Optic Nerve Disc Drusen

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## ABSTRACT

Optic nerve head drusen (ONHD) are calcified bodies in the optic nerve which are diagnosed incidentally during the ophthalmic examination. Patients with ONHD are usually asymptomatic, yet can be presented with blurry vision and transient visual obscurations. ONHD diagnosis can be challenging for ophthalmologists because clinically it mimics optic nerve disc swelling and can be misdiagnosed with papilledema or optic neuritis. For a proper diagnosis to be made, clinical examination supported by diagnostic tests as ultrasound B scan, optical coherence tomography (OCT), and computed tomography (CT) orbits are required. The presented case is a 38 year old healthy lady with ONHD presented with asymmetrical vision loss. Following clinical assessment and investigations, non-arteritic anterior ischemic optic neuropathy (NAION) secondary to ONHD was diagnosed.

## INTRODUCTION

Drusen of the optic nerve head (ONHD) are acellular, hyaline-like bodies composed of glycoprotein which gradually calcify within optic nerve head [1, 2]. The reported prevalence is 3.4 24/1000 with more than 70% bilaterality [3, 4]. Patients with ONHD generally are asymptomatic but can present occasionally with visual disturbances. The most common reported symptoms are transient visual obscuration, painless vision loss and visual field loss [5, 6]. Visible optic disc drusen are associated with visual field defects with an incidence up to 71% [5, 6]. ONHD is believed to be a risk factor for developing anterior ischemic optic neuropathy due to its crowding and compressive effect on the nerve axons [7]. The reported case has ONHD with bilateral anterior ischemic optic neuropathy complicated by bilateral optic atrophy with asymmetrical visual outcome.

## CASE PRESENTATION

A 38-year-old lady presented to the clinic with a history of decreased vision in her left eye of one-month duration. The patient had attacks of headache associated with her visual complaints.

The past medical history of the patient was not significant. Her vision in the right eye was 20/25 (refractive error of -0.5 spherical - 1.00-cylinder axis 170) and the left eye vision was 20/40 (-0.75 spherical - 1.00-cylinder axis 170) at the time of presentation. Slit-lamp biomicroscopy of the anterior segment was unremarkable in both eyes. She had a relative afferent pupillary defect in her left eye. Color vision was 11 of 11 Ishihara plates in the right eye and 4 of 11 in the left eye. Dilated pupillary examination showed visible extensive optic disc drusen in both eyes.

Ultrasound B scan confirmed the presence of Optic nerve head drusen (ONHD) in both eyes

(Figure 1). Spectral-domain optical coherence tomography also confirmed the presence of ONHD with severe retinal nerve fiber layer (RNFL) loss (Figure 2). A reliable Humphrey central 24-2 visual field test of the right eye demonstrated decreased sensitivity and inferior altitudinal defect with a mean deviation of -15.6 dB,

whereas the left-field showed generalized constriction and inferior altitudinal defect involving the central 5 degrees with a mean deviation of -21.6 dB (Figure 3, 4).

The patient was referred to neurologist for a complete workup and lab work was unremarkable. Magnetic resonance imaging (MRI) of the brain and orbit, with and without contrast showed atrophy of the optic nerves including the intraorbital segments, optic chiasm, and tracts (Figure 5). Yet, the cause of retrobulbar optic atrophy, couldn't be addressed. The patient was seen again after 4 weeks, and with refraction, the visual acuity was at 20/20 in the right eye and remained at 20/40 in the left eye with no improvement. A diagnosis of ONHD with anterior ischemic optic neuropathy in both eyes was made.

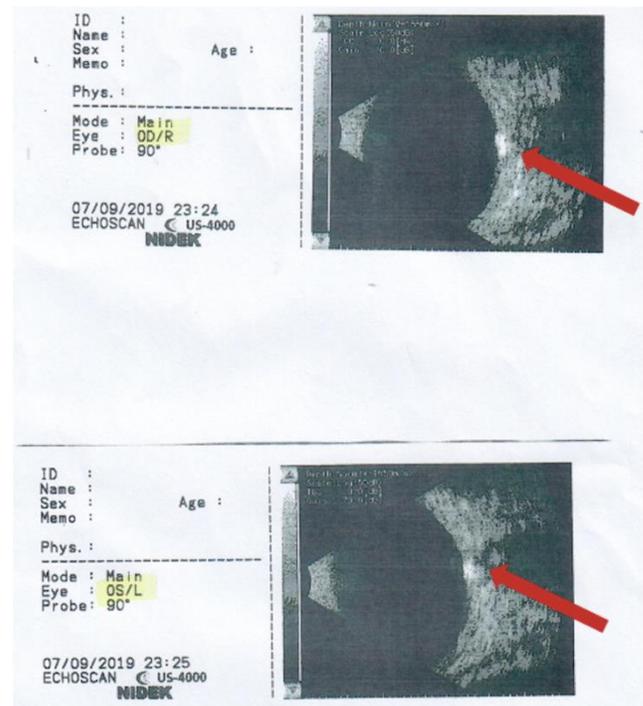


Figure 1: Ultrasound B scan showing drusen

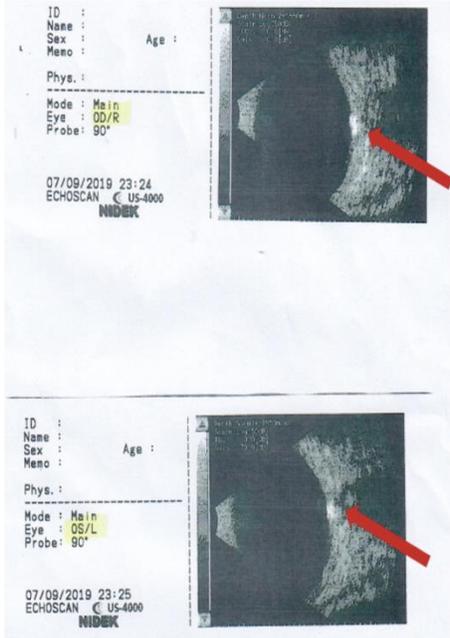


Figure 1: Ultrasound B scan showing drusen

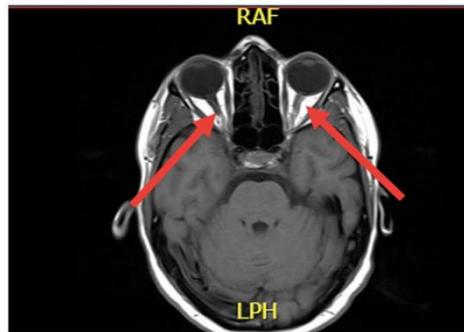


Figure 2: Optical coherence tomography retinal nerve fiber layer thickness shows optic disc drusen and significant retinal nerve fiber layer loss in both eyes

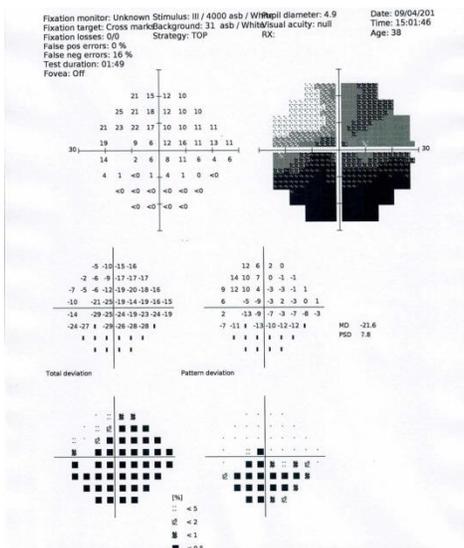


Figure 3: Visual field showing inferior altitudinal defect in the left eye

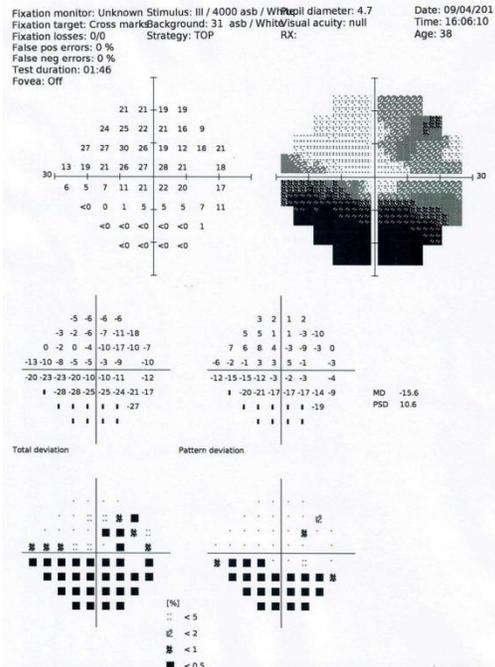


Figure 4: Visual field showing inferior altitudinal defect in the right eye

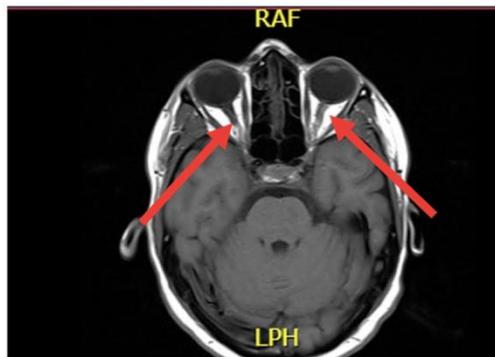


Figure 5: Magnetic resonance imaging showing optic nerve atrophy in both eyes

## DISCUSSION

ONHD are diagnosed usually as incidental findings during routine fundus exam. Drusen are asymptomatic, yet, patients may observe transient visual obscuration which could happen due to transient ischemia of the optic nerve head. Calcified drusen are believed to increase the tissue pressure of the optic disc and may cause transient impairment of optic disc circulation [8, 9].

ONHD may cause visual field defects similar to other ophthalmological disorders such as glaucoma, and NAION [7]. Moreover, it has been reported to be gradual and shows different visual field defects such as arcuate defects, enlarged blind spot, general depression, and altitudinal defects [7, 10-16]. The compressive effect on nerve axons by drusen is assumed to be the cause [7, 10-16].

There are several conditions which result in significant vision loss in the elderly where Nonarteritic anterior ischemic optic neuropathy (NAION) is considered to be one

of the common causes. However, NAION has been reported to occur in a younger age group in patients with optic nerve drusen [7, 11, 13, 17]. The patient's examination, with a relative afferent pupillary defect and retinal nerve fiber layer loss in OCT and bilateral altitudinal visual field defect combined with disc drusen, is most consistent with NAION. In accordance with the presented case, NAION patients have different visual outcomes in which drusen related usually have better visual outcomes than the elderly [7-17]. Optic disc drusen can mimic edematous optic nerve and therefore the patient was referred to neurology and imaging was done to rule out intracranial hypertension. In the absence of other neurological disorders that can cause optic atrophy, the presence of bilateral optic nerve atrophy in MRI could be related to NAION in which drusen bodies cause infarction to optic disc vessels as proposed in Purvin et al report [7]. Moreover, the patient had no other systemic condition that could result in developing NAION. This report contributes to the literature as a new case of association between ONHD and NAION in a middle-aged adult with good visual outcome in detailed documentation using modern techniques.

## CONCLUSION

ONHD can occasionally cause NAION. Ophthalmologists should highly consider this association since no current prevention or treatment for NAION is identified. Yet, ONHD can mimic other neurological disorders, detailed investigation should be performed in patients with suspected possible systemic disorders. Therefore, patients with ONHD should be followed on a regular basis with serial visual fields, nerve fiber layer analysis, and fundus photography. Hence, the management protocol is to monitor for progressive vision loss or to detect possible complications that may develop and any other associated risk factors. This is in order to provide low vision rehabilitation therapy to assist patients with vision impairment and maintain daily life activity.

## REFERENCES

1. 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.
2. Tso MO. Pathology and pathogenesis of drusen of the optic nervehead. Ophthalmology. 1981;88(10):1066-80. doi:10.1016/s0161-6420(81)80038-3
3. Auw-Haedrich C, Staubach F, Witschel H. Optic disk drusen. Survey of ophthalmology. 2002;47(6):515-32. doi:10.1016/s0039-6257(02)00357-0
4. Friedman AH, Gartner S, Modi SS. Drusen of the optic disc. A retrospective study in cadaver eyes. The British journal of ophthalmology. 1975;59(8):413-21. doi:10.1136/bjo.59.8.413
5. Lee AG, Zimmerman MB. The rate of visual field loss in optic nerve head drusen. Am J Ophthalmol. 2005;139(6):1062-6. doi:10.1016/j.ajo.2005.01.020
6. Wilkins JM, Pomeranz HD. Visual manifestations of visible and buried optic disc drusen. Journal of neuro-ophthalmology : the official journal of the North American Neuro-Ophthalmology Society. 2004;24(2):125-9. doi:10.1097/00041327-200406000-00006
7. Purvin V, King R, Kawasaki A, Yee R. Anterior ischemic optic neuropathy in eyes with optic disc drusen. Archives of ophthalmology (Chicago, Ill : 1960). 2004;122(1):48-53. doi:10.1001/archoph.122.1.48
8. Sadun AA, Currie JN, Lessell S. Transient visual obscurations with elevated optic discs. Annals of neurology. 1984;16(4):489-94. doi:10.1002/ana.410160410
9. Tan DK, Tow SL. Acute visual loss in a patient with optic disc drusen. Clinical ophthalmology (Auckland, NZ). 2013;7:795-9. doi:10.2147/oph.s42233
10. Farah SG, Mansour AM. Central retinal artery occlusion and optic disc drusen. Eye (London, England). 1998;12 ( Pt 3a):480-2. doi:10.1038/eye.1998.112
11. Gittinger JW, Jr., Lessell S, Bondar RL. Ischemic optic neuropathy associated with optic disc drusen. Journal of clinical neuro-ophthalmology. 1984;4(2):79-84.
12. Michaelson C, Behrens M, Odel J. Bilateral anterior ischaemic optic neuropathy associated with optic disc drusen and systemic hypotension. The British journal of ophthalmology. 1989;73(9):762-4. doi:10.1136/bjo.73.9.762
13. Newman WD, Dorrell ED. Anterior ischemic optic neuropathy associated with disc drusen. Journal of neuro-ophthalmology : the official journal of the North American Neuro-Ophthalmology Society. 1996;16(1):7-8.
14. Liew SC, Mitchell P. Anterior ischaemic optic neuropathy in a patient with optic disc drusen. Australian and New Zealand journal of ophthalmology. 1999;27(2):157-60. doi:10.1046/j.1440-1606.1999.00176.x
15. Kamath GG, Prasad S, Phillips RP. Bilateral anterior ischaemic optic neuropathy due to optic disc drusen. European journal of ophthalmology. 2000;10(4):341-3. doi:10.1177/112067210001000414
16. Ayhan Z, Yaman A, Söylev Bajin M, Saatci AO. Unilateral Acute Anterior Ischemic Optic Neuropathy in a Patient with an Already Established Diagnosis of Bilateral Optic Disc Drusen. Case reports in ophthalmological medicine. 2015;2015:730606. doi:10.1155/2015/730606
17. Bandyopadhyay S, Singh R, Gupta V, Gupta A. Anterior ischaemic optic neuropathy at high altitude. Indian journal of ophthalmology. 2002;50(4):324-5