CASE REPORT

A Case of Bilateral Optic Nerve Head Drusen-Induced Inferior Altitudinal Hemianopsia

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ABSTRACT

The current case involved a 59-year-old woman who was referred to our hospital with bilateral optic nerve abnormality and gradually progressive bilateral inferior visual field defects. An ophthalmological examination revealed superficial optic nerve head drusen (ONHD) and bilateral inferior altitudinal hemianopsia, but no intracranial lesions. The results of an angiogram revealed no evidence of retinal vascular occlusion or anterior ischaemic optic neuropathy (AION). Prior reports suggested that drusen-induced hemianopsia was slowly progressive but that ONHD-induced AION caused acute altitudinal hemianopsia, which led us to the diagnosis of bilateral drusen-induced altitudinal hemianopsia.

Keywords: Bilateral inferior altitudinal hemianopsia, optic nerve head drusen, visual field defect

INTRODUCTION

Optic nerve head drusen (ONHD) presents with many uncertainties in terms of causation. Pathologically, it has been deduced that abnormal axonal metabolism leads to the deposition of high levels of calcium in the mitochondria, which extrude into the extracellular space.¹ In addition, it has been proposed that the visual field defects associated with ONHD are caused by a mechanism in which the calcified bodies compress adjacent ganglion cell axons, resulting in ganglion cell death, retrograde axonal degeneration, and subsequent loss of visual field.² Although most patients with ONHD are asymptomatic, 71% of cases are identified through the detection of peripheral visual field defects.³ The diverse patterns of visual field defects include pericaecal scotoma, arcuate scotoma, and concentric constriction.^{3–6} Further, some cases demonstrate irregular or advanced visual field defects including inferior altitudinal hemianopsia,^{6–8} but observation of these symptoms are generally rare. Nonetheless, the current report involves a case in which we observed superficial ONHD that resulted in bilateral inferior altitudinal hemianopsia.

CASE REPORT

The patient was a 59-year-old woman with no apparent family history. Sixteen years prior, a health examination indicated morphological bilateral optic disc head abnormalities, and a subsequent fundus examination conducted in our hospital revealed angioid streaks and ONHD. The patient was counselled to undergo a detailed examination, but she declined further testing. However, as time progressed, her visual field gradually narrowed. Following a visit to a local physician, the patient was diagnosed with morphological bilateral optic nerve head abnormalities and bilateral inferior altitudinal hemianopsia and was referred to our hospital for further examination.

During an examination at our hospital, the patient's corrected visual acuity was 20/20 1.0 in both eyes, whereas intraocular pressure in the right and left eyes was 10 and 9 mm Hg, respectively. The detection of a relative afferent pupillary defect was indefinite in both eyes, and we observed no evident abnormalities in the anterior segment or the optic media. In colour fundus photographs, excavation of the optic nerve head had disappeared in both eyes and highly reflective protrusions with a mulberry-like

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FIGURE 1 (A and B) Colour fundus photographs. Optic nerve head drusen (ONHD) and angioid streaks extending radially from the nerve head were observed in both eyes. (C and D) Fundus autofluorescence imaging prior to contrast agent injection. Marked autofluorescence consistent with drusen was observed in both eyes.

appearance had accumulated (Figure 1A and B). The macular area was intact in both eyes, but we observed angioid streaks extending from the optic nerve head and *peau d'orange* fundus. Fundus autofluorescence imaging revealed marked autofluorescence consistent with drusen prior to injection of fluorescein (Figure 1C and D). In vertical-sectional images of the nerve head acquired using swept source optical coherence tomography (SS-OCT; DRI OCT-1 Atlantis; Topcon Corporation, Tokyo, Japan), accumulation of substantial ONHD appeared to extend to the deepest layer of the nerve head (Figure 2A). The protrusions of the ONHD were little different between the upper and lower regions. Measurements obtained using spectral domain optical coherence tomography (SD-OCT; 3D OCT-2000 FA; Topcon) demonstrated bilateral advanced thinning of both the retinal nerve fibre layer (RNFL) surrounding the optic nerve head and the inner retinal layer throughout the retina (Figures 2B, 3A and B). During fundus autofluorescence, the retina surrounding the nerve head demonstrated hyperfluorescence consistent with angioid streaks, but new vessels and altitudinal differences in perfusion were not observed. In Goldmann perimetry, we observed bilateral concentric constriction and inferior altitudinal hemianopsia with multiple overlapping isoptres on the horizontal meridian (Figure 4). In addition, OCT revealed a divergence between the

visual field defects and RNFL thickness and inner retinal layer thickness.

Computed tomography (CT) revealed ONHD limited to the optic discs. Magnetic resonance imaging (MRI) showed that both optic nerves were remarkably thinned. However, no sign of cortical deficit, infarction, or space-occupying lesion was observed.

All clinical examinations of the patient were performed in accord with the guidelines of the Declaration of Helsinki.

DISCUSSION

Generally, altitudinal hemianopsia is a rare manifestation of ONHD. In the current case, four potential mechanisms were considered as explanations for the bilateral inferior altitudinal hemianopsia. The first possible mechanism included intracranial lesions in the form of cerebral infarction and brain tumours.^{9,10} The second possibility included the bilateral occurrence of anterior ischaemic optic neuropathy (AION) induced by ONHD.^{11,12} The third possible mechanism was retinal vascular occlusion caused by ONHD, whereas the fourth possibility involved compression of nerve fibres by drusen. However, cranial imaging and neurological findings did not support the presence of intracranial lesions or cerebral infarction.

FIGURE 2 (A) Vertical-sectional images of the optic nerve heads acquired using swept source optical coherence tomography (SS-OCT) in both eyes. S and I refer to superior and inferior directions, respectively. Solid optic nerve head drusen (ONHD) accumulations extended to the deepest layer of the optic nerve head. (B) Spectral domain optical coherence tomography (SD-OCT) analysis of the thickness of the circum-papillary retinal nerve fibre layer (RNFL). The RNFL demonstrated complete thinning except for on the nasal side in both eyes.

As well, a fluorescein angiogram revealed no evidence of AION, and the patient had no risk factors for AION including arteriosclerotic factors such as diabetes or hypertension. According to previous reports on ONHD-induced AION by Kamath *et al.*¹¹ and Purvin *et al.*,¹² inferior altitudinal hemianopsia normally progresses along a course of acute apoplectic onset.¹⁰ In our case, however, the visual field defects were slowly progressive. Consequently, the second possibility of AION comorbidity was rejected.

In addition to AION, retinal artery occlusion, retinal vein occlusion, and other forms of retinal vascular occlusion are known to occur in cases of ONHD.^{13,14} Nonetheless, these possibilities were all rejected based on the fluorescein angiography and OCT findings. Accordingly, we concluded that the inferior altitudinal hemianopsia was caused by compression of the nerve fibres. In contrast to ONHD-induced AION, previous reports by Morris *et al.*⁶ and Roh *et al.*⁸ indicated that drusen-induced unilateral inferior altitudinal hemianopsia progressed slowly.

To our knowledge, this is the first report of bilateral ONHD-induced altitudinal hemianopsia.

OCT is useful as a supplementary diagnostic tool for assessing changes in RNFL thickness due to ONHD.^{8,15} However, as the present case demonstrated, detecting sites of visual field changes that correspond to morphological changes can be difficult, even when the changes in the retinal morphology can be detected by OCT. In a similar report in which OCT was used to assess superficial ONHD, Schuman *et al.* presented a number of cases with overall thinning of the RNFL surrounding the nerve head in which the thinning sites and the visual field loss sites did not completely match.¹⁶ The RNFL includes not only ganglion cells and axons but also glial cells. Consequently, thinning of the RNFL may not necessarily signify a reduction in axons. Similarly, thinning of the ganglion cell layer does not necessarily signify a reduction in ganglion cells. These phenomena may explain the divergence between retinal thinning sites and visual field defect sites in the present case.

FIGURE 3 (A and B) The inner retinal layer in the macular area by SD-OCT demonstrated even and overall advanced thinning throughout the retina in the right eye (A) and left eye (B).

FIGURE 4 Goldmann perimetry. Although the central visual field was preserved in both eyes, bilateral superior temporal concentric contraction, and inferior altitudinal hemianopsia with multiple overlapping isoptres on the horizontal meridian were observed.

CONCLUSION

To our knowledge, this is the first report of bilateral altitudinal hemianopsia in ONHD. Prior reports suggest that altitudinal hemianopsia caused by drusen compression progressed slowly, whereas ONHD-induced AION caused acute altitudinal hemianopsia, which factored into the current diagnosis. Further, OCT analysis of RNFL or ganglion cell layers may not be useful in chronic nerve fibre damage in ONHD.

Declaration of interest: The authors report no conflicts of interest. The authors alone are responsible for the content and writing of the paper.

Note: Figures 1–4 of this article are available in colour online at <http://informahealthcare.com/oph>.

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