CASE REPORT

Bilateral Simultaneous Nonarteritic Anterior Ischaemic Optic Neuropathy: Case Report

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ABSTRACT

Bilateral simultaneous nonarteritic anterior ischaemic optic neuropathy (NAION) is extremely rare. A 57-year-old woman presented with bilateral optic disc oedema and peripapillary splinter haemorrhages. Initial visual acuities were hand movements in the right eye and light perception in the left eye. The patient had a mildly elevated diastolic blood pressure and glucose intolerance. Erythrocyte sedimentation rate and C-reactive protein levels were within normal limits. Temporal artery biopsy was negative for temporal arteritis. Marked visual improvement occurred in both eyes (0.8 in the right eye, 0.6 in the left eye) after systemic steroid therapy in the 16th month of follow-up.

Keywords: nonarteritic anterior ischaemic optic neuropathy treatment, simultaneous nonarteritic anterior ischaemic optic neuropathy

INTRODUCTION

Anterior ischaemic optic neuropathy (AION) causes sudden visual loss and visual field defects, which can be associated with various systemic disorders, such as giant cell arteritis, diabetes, atherosclerosis, and systemic connective tissue diseases.¹ Arteritic AION (AAION) is caused by giant cell arteritis, and nonarteritic AION (NAION) is the commonly occurring type and not due to giant cell arteritis.² Bilateral AION occurs more often in arteritic patients and bilateral simultaneous NAION is extremely rare.^{3–5} There are only a few reports of bilateral simultaneous NAION, which is usually caused by sudden systemic arterial hypotension.⁶ In this report, we present a patient with simultaneous NAION treated with systemic corticosteroids.

CASE REPORT

A 57-year-old woman with sequential vision loss within 12 hours was referred for definitive diagnosis

and treatment. At presentation, visual acuity (VA) was hand movements in the right eye (OD), and light perception (LP) in the left (OS). A relative afferent pupillary defect (RAPD) was detected in the left eye. Direct light responses were weak bilaterally. Anterior segment examination was unremarkable in both eyes. Fundoscopy revealed swollen optic discs and peripapillary splinter haemorrhages (more distinctive in the left eye) in both eyes (Figure 1A, B). Intraocular pressure was 14 mm Hg bilaterally. Optic disc horizontal diameters were 1.6 mm, bilaterally. Temporal artery biopsy was performed although both temporal arteries were evaluated as normal on palpation.

Fat-suppressed, contrast-enhanced magnetic resonance imaging showed the absence of inflammatory changes (Figure 2). Colour Doppler ultrasound examination of both carotid arteries showed no significant obstruction. The patient did not have any systemic problem and did not report weight loss, fever, headache, or depression. Blood pressure was 134/90 mm Hg without any medication. Serum glucose level (85 mg/dL), erythrocyte sedimentation rate (ESR; 18/mm/h), and C-reactive protein (CRP;

Received 29 April 2013; revised 29 May 2013; accepted 4 June 2013; published online 19 September 2013

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FIGURE 1. Fundus photos during the initial examination (A, B) and the 16th-month examination (C, D). *Note:* Figure 1 of this article is available in colour online at www.informahealthcare.com/oph.

FIGURE 2 Coronal and axial magnetic resonance images. Fat-suppressed, contrast-enhanced T1-weighted image (A); fat-suppressed T2-weighted image (B).

2.5 mg/dL) were all in normal limits. However, serum haemoglobin A1c (HbA1c) level was 6.8% (normal: 4–6.5%). Internal medicine consultation revealed glucose intolerance. Serum total cholesterol, low-density lipoprotein (LDL) cholesterol, and triglyceride levels were 271 (normal: 125–240), 161 (normal: ≤ 150), and 180 (normal: 50–150) mg/dL, respectively. Serum aspartate aminotransferase (AST) and alanine aminotransferase (ALT) levels were 57 (8–40) and 121 (10–40) mg/dL, respectively. Anti-nuclear antibodies, anti-DNA, cytoplasmic and perinuclear anti-neutrophil cytoplasmic antibodies (c-ANCA and p-ANCA), and

anti-phospholipid antibodies were negative. Thyroid function tests, angiotensin-converting enzyme, rapid plasma reagin, chest radiography, and thorax contrast tomography were all in the normal range or unremarkable. Full-field electroretinogram showed normal rod and cone responses. Temporal artery biopsy was negative for inflammation in the vascular wall.

Upon initial examination, systemic steroid (methylprednisolone 80 mg/day) and aspirin (100 mg/day) was initiated. At the end of the 1st week, visual acuities (Vas) in both eyes progressively increased to counting fingers from 4 and 1 m in OD/OS,

FIGURE 3 Humphrey visual field examinations during the 2nd-week (A, B), 6th-month (C, D), and 16th-month (E, F) examinations.

FIGURE 4 Retinal nerve fibre layer analysis at the 16th-month examination.

respectively. At the end of the 2nd week, VAs were 0.5 (OD) and 0.4 (OS). Meanwhile, optic disc oedema resolved substantially, and both optic discs were mildly pale. Visual field (VF) testing showed overall sensitivity loss in both eyes, with a dominant inferior altitudinal defect in the left eye (Figure 3A, B).

Visual acuities were 0.6 (OD) and 0.5 (OS) at the 6th month. The morphology of the visual field defects at the 6th month was similar to the 2nd-week VF, with a lower mean deviation (Figure 3C, D). VAs increased to 0.8 (OD) and 0.6 (OS) at the 16th-month examination. Both optic discs were pale (Figure 1C, D). Relative afferent pupillary defect in the left eye persisted. Retinal nerve fibre layer analysis showed pathologic thinning in temporal, superior, and inferior quadrants in both eyes (Figure 4). However, normal visual fields were observed (Figure 3E, F). Pattern visual evoked potentials of the patient showed normal morphologies but delayed latencies and reduced amplitudes in both eyes. Colour Doppler ultrasonography showed diastolic restriction of the right retinal artery.

DISCUSSION

AION is caused by diminished blood supply to the optic disc and anterior portion of the optic nerve by the posterior ciliary arteries. Sudden, often catastrophic, "stroke-like" vision loss in elderly patients with vasculopathic risk factors is typical of this condition. AION may have an arteritic (AAION) and nonarteritic (NAION) origin. AAION occurs

in patients with temporal arteritis (giant cell arteritis). Immediate systemic steroid therapy should be initiated in patients with AAION in order to prevent fellow eye involvement. In contrast to AAION, NAION usually have diabetes and/or hypertension, and are particularly at risk if they have a small, crowded optic nerve head. Other systemic conditions such as antiphospholipid syndrome, previous radiation therapy, juvenile diabetes, shock, severe hypertension, hyperlipidaemia, nocturnal hypotension, sleep apnoea, and migraine have been reported in association with NAION. Our case had a mildly elevated diastolic blood pressure (90 mm Hg), hyperlipidaemia and glucose intolerance. In contrast to hypertension, nocturnal hypotension is one of the frequently suggested risk factors in NAION.⁷⁻¹⁰ This explains the high rate of vision loss upon awakening in these patients. NAION in patients with sleep apnoea may also be related to nocturnal hypotension. Hypertension treatment may cause nocturnal hypotension, which may cause NAION in these patients who already have vasculopathy.

A study reported a lifetime risk of 30-40% of second eye involvement in NAION.⁵ Five-year risk was 14.7% in the ischaemic optic neuropathy decompression trial.¹¹ The cumulative incidence rates are even higher in diabetic and hypertensive patients.¹² However, bilateral simultaneous NAION cases are extremely rare, except in cases with severe arterial hypotension during cardiopulmonary or extensive surgery with massive blood loss, haemodialysis, etc.¹³

Yamauchi et al. reported bilateral simultaneous AION in a patient with Behçet's disease¹⁴.

Recently, bilateral simultaneous NAION cases were reported in association with sildenafil use.^{15–17} However, the incidence of development of NAION among millions of person using phosphodiesterase-5 inhibitors is so small that a possible relationship has been regarded as impossible to prove by sceptical ophthalmologists. In contrast, Hayreh is in favour of a cause-and-effect relationship mostly because of a nocturnal hypotensive effect of phosphodiesterase-5 inhibitors.¹⁸ Shibayama et al. reported a case of bilateral and almost simultaneous NAION in a diabetic patient with small discs and small optic cups.³

Hayreh and Zimmerman¹⁹ introduced the term “incipient NAION” in 2007. The authors presented 55 patients (60 eyes) with optic disc oedema without any visual loss attributable to classic NAION. However, 55% of the fellow eyes had classic NAION, 25% of the eyes progressed to NAION after a median time of 5.8 weeks. The authors concluded that optic disc oedema was a pre-symptomatic stage of NAION. Patients with “incipient” compared with classic NAION had greater prevalence of diabetes mellitus. We do not know whether our patient had that pre-symptomatic stage or optic disc oedema before the clinical onset of NAION. However, the patient reported sudden visual loss in both eyes.

Bilateral simultaneous NAION is extremely rare. For us, the most critical point is to decide whether it has an arteritic or nonarteritic origin. Immediate systemic steroid treatment carries prognostic and vital significance in AAION, it may be better to initiate this treatment in doubtful cases. However, there is no known effective therapy for NAION. Steroids were thought to be ineffective for decades; however, Hayreh and Zimmerman reported better visual acuity and visual field results with systemic corticosteroid therapy.²⁰ The significance of these findings is unclear, for this reason, their use is not common in clinical practice. Hyperbaric oxygen and aspirin were also shown to be ineffective.^{21–23} Because of the recent reports on the effects of systemic corticosteroid treatment in NAION, it seems better to initiate this therapy in patients with simultaneous AION in elderly patients.

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

Note: Figure 1 of this article is available in colour online at informahealthcare.com/oph

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