CASE REPORT

Keeping NAION visual loss: discriminating urgent versus emergent visual loss in an elderly female

Sarah Lubner,¹ Richard Alweis^{1,2}

¹Department of Medicine, Reading Health System, West Reading, Pennsylvania, USA
²Jefferson Medical College, Philadelphia, Pennsylvania, USA

Correspondence to

Dr Richard Alweis, Richard.Alweis@readinghealth.org

Accepted 9 March 2014

SUMMARY

A 72-year-old woman presented with sudden, monocular vision loss and a temporal headache without eye pain. Examination revealed complete loss of vision in the right eye in bilateral superior visual fields and hyperaemic oedema with haemorrhages of the optic disc. She had significant vasculopathic risk factors, including age greater than 50 years, hypertension, recent coronary artery bypass graft and hyperlipidaemia. Atypical features were investigated, including simultaneous vision changes in contralateral eye, prodrome, jaw claudication, scalp tenderness and headache. The patient indicated persistent temporal headaches since the onset of the visual changes, prompting investigation with temporal biopsy and inflammatory markers with initiation of steroids. Diagnostic studies excluded inflammatory disease and retinal vascular thrombosis. The patient was diagnosed with non-arteritic anterior ischaemic optic neuropathy (NAION) and the steroids were tapered off. At 6 months, the patient has maintained altitudinal visual loss, but her central vision in the affected eye has remained 20/25.

Physical examination revealed an alert, obese female with stable vital signs, including blood pressure of 134/76 mm Hg and a regular heart rate of 72 bpm. Her visual acuity with correction was 20/25 on the right and 20/20 on the left eye. Intraocular pressure was 16 mm Hg on the right and 17 mm Hg on the left eye. Extraocular motility was fully intact. The anterior segment examination showed a well-positioned posterior chamber intraocular lens bilaterally. A dilated funduscopic examination showed 1–2+ hyperaemic oedema of the optic disc with splinter haemorrhages and dilated telangiectatic capillaries in the right eye. The left eye examination revealed a normal optic nerve and retina with a cup-to-disc ratio of 0.3. Visual field examination revealed dense superior altitudinal loss on the right with no deficits on the left eye. Nerve fibre layer optical coherence tomography (NFL OCT) was obtained and showed significant nerve fibre layer oedema in the right eye, more so in the inferior nasal segment. The left side was within normal limits. Her temporal arteries were non-tender, and she had no evidence of inflammatory arthritis of the hips or shoulders.

BACKGROUND

Non-arteritic anterior ischaemic optic neuropathy (NAION) is the most common form of ischaemic optic neuropathy seen in 2–10/100 000 persons per year and has the best prognosis.¹ Alternatively, a similar counterpart, arteritic ischaemic optic neuropathy (AION), is less common and has very poor vision prognosis if left untreated. Given the significant differences in outcome, it is important for the clinician to understand the features of NAION as well as atypical 'red flag' features that should initiate further testing and prompt treatment.

CASE PRESENTATION

A 72-year-old woman with a medical history including coronary artery disease status after bypass surgery 5 months prior to presentation, hypertension, hyperlipidaemia, migraines, bilateral pseudophakia and obstructive sleep apnoea on continuous positive airway pressure (CPAP) therapy presented with monocular visual disturbance. The visual change was characterised by a sudden development of diffuse blurriness of the right eye that persisted and progressively worsened for 1 week prior to presentation. She denied associated ocular pain, halos, discharge, photosensitivity, scalp tenderness, jaw claudication or prodromal symptoms. However, she did describe severe intermittent generalised headaches since the onset of the visual change.

INVESTIGATIONS

Laboratory evaluation, including complete blood counts, electrolytes and renal function were within normal limits. Most notably, her erythrocyte sedimentation rate (ESR) was 10 mm/h and C reactive protein (CRP) was 0.39 mg/dL. Lyme disease titre was negative. CT of the brain/head was obtained and showed chronic small vessel ischaemic disease as well as atherosclerotic calcification of internal carotid and vertebral arteries. There was no evidence for a compressive or infiltrative aetiology. The patient was sent for urgent right temporal artery biopsy which showed non-specific mild intimal proliferation, negative for arteritis.

DIFFERENTIAL DIAGNOSIS

The differential diagnosis for NAION includes optic neuritis, Leber's hereditary optic neuropathy, neuroretinitis and AION. Of these diagnoses, differentiation of NAION from AION is of particular importance. The primary differentiating factors are prodromal symptoms and appearance of disc oedema. Giant cell arteritis is typically associated with proximal muscle weakness, headaches, temporal scalp tenderness and jaw claudication. The optic disc in AION is typically pallid rather than hyperaemic as seen in NAION. The prognosis of untreated AION is quite poor and could lead to

CrossMark

To cite: Lubner S, Alweis R. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2013-202262

rapid blindness, making the differentiation of arteritic and non-arteritic diseases paramount.

TREATMENT

On initial evaluation, there was concern for giant cell arteritis due to elderly age, headache and optic nerve oedema. She was started on prednisone therapy at 50 mg once daily. Two days after initiation of steroid therapy, the temporal artery biopsy was performed. The steroids were tapered off when the biopsy returned clear for inflammation. She was diagnosed with NAION due to the patient's age, presence of vasculopathic risk factors, the appearance of optic neuropathy, relatively good vision despite dense visual field loss and the negative biopsy result. The risk factors associated with NAION were discussed with the patient and she was encouraged to continue aspirin 81 mg as well as her CPAP therapy.

OUTCOME AND FOLLOW-UP

At 6 months of follow-up, the patient continues to have her altitudinal visual loss, but her central vision in the affected eye has remained 20/25. The unaffected eye has remained normal, with vision of 20/20 and no evidence of optic neuropathy.

DISCUSSION

In patients greater than 50 years of age, ischaemic optic neuropathy is the most common optic nerve disorder, with the most common of these neuropathies being NAION.² While there are multiple proposed mechanisms of NAION pathogenesis, the aetiology remains unclear. However, a unifying theory suggests that small vessel vascular insufficiency leads to an ischaemic insult generating swelling of the optic nerve. At the crowded optic nerve head and in the right clinical milieu, this progresses to infarction.^{3 4} Pathogenesis, while controversial, appears to be related to non-thrombotic hypoperfusion from transient hypotension. This is in clear differentiation of retinal vascular occlusion secondary to thrombosis. NAION fluorescein fundus angiography shows delayed but transient filling of the peripapillary choroid whereas thromboembolic disease would demonstrate complete occlusion. Additionally, NAION does not have an association with smoking and there is no response to aspirin in direct contrast with thromboembolic disease.⁵ In NAION, as in our patient, there is sudden, unilateral painless visual loss

with examination revealing optic disc swelling, loss of visual acuity, afferent pupillary defect and splinter haemorrhages. The unaffected eye is often reported to have a small cup-to-disc ratio. In two-thirds of patients, vision loss is often first noticed on awakening in the morning, and described as a diffuse blurriness that persists and slowly progressive over several days. The average peak vision loss is thought to occur at 5 days.⁶ Generally visual acuity loss is to a lesser extent than other optic neuropathies with residual central vision and better than 20/60 in more than half of the cases reported in some studies, but there can be complete loss of light perception depending on the region of infarction.^{2 6 7} Classically, the defect is altitudinal with absolute inferior nasal defect being most prevalent.⁸ A key discriminator between NAION and the more severe AION is the optic disc oedema that tends to be hyperaemic with splinter haemorrhages in NAION rather than pallid, as is more common in AION.⁴ NAION is most commonly described as painless; however, retrospective studies have demonstrated approximately 12% of patients to experience headache or ocular pain.⁵

NAION is a clinical diagnosis, with age, presence of vasculopathic risk factors (refer to [box 1](#)), vision defect pattern and the appearance of oedematous optic disc being the key factors in making the diagnosis. Since as many as 60% of patients with NAION have diabetes, hypertension or use tobacco, particular attention needs to be paid to these historical risk factors.⁹ Additionally, identifying those patients with sleep apnoea may be of significance, particularly in risk reduction, as studies have also shown that sleep apnoea potentially carries three times the risk for NAION than either diabetes or hypertension and may be causative due to nocturnal hypoperfusion.^{10–12}

While the clinical features and diagnosis of NAION is of importance, it is of further significance to recognise atypical features ([box 2](#)) that should alert to more urgent alternative diagnoses, particularly AION (eg, giant cell arteritis). In patients older than 55 years of age, evaluation should include ESR and CRP. Additionally, a temporal artery biopsy should be considered if these inflammatory markers are elevated or if there is a high clinical suspicion, particularly if the patient describes systemic symptoms typical of polymyalgia rheumatica.⁷ If there is a likely delay in obtaining a temporal artery biopsy, oral steroids should be initiated, as this is controversial but beneficial in NAION (though may worsen diabetes mellitus or hypertension), and also may save vision in AION. Lack of prompt treatment in AION may lead to rapid blindness and vision recovery is poor once this loss has occurred.⁴

NAION is a clinical diagnosis based on elderly age, presence of vasculopathic risk factors, pattern of vision defect, an

Box 1 Risk factors for non-arteritic anterior ischaemic optic neuropathy

Risk factors:

Major (>10%)

- Atherosclerosis risk factors, particularly hypertension and diabetes
- Sleep apnoea syndrome
- Nocturnal hypotension

Minor (<10%)

- Prothrombotic conditions
- Small optic cup-to-disc ratio
- Renal failure
- Surgery (particularly cardiopulmonary bypass and spine)
- Medications (phosphodiesterase-5 inhibitors, interferon- α , amiodarone and sympathomimetics)
- Migraines
- Carotid dissection

Box 2 Features suggestive of alternative diagnosis to non-arteritic anterior ischaemic optic neuropathy

Risk factors

- Age <50
- Lack of vasculopathic risk factors
- Bilateral vision loss
- Prodromal symptoms
- Systemic symptoms (jaw claudication, proximal myalgias or arthralgias, scalp tenderness, headache and fatigue)
- Intermittency of symptoms
- Recurrence in the same eye
- Pallid optic disc

oedematous optic disc, and most importantly, the exclusion of arteritic disease. After the initial visual insult, most patients will stabilise without further visual loss. Worsening of vision after the initial period should lead to further investigation. Recurrence of NAION in the affected eye is rare. In cases of optic neuropathy, it is paramount to recognise atypical features that may require further testing and early treatment.

Learning points

- ▶ Non-arteritic anterior ischaemic optic neuropathy diagnosis is based on clinical findings and most importantly, the exclusion of arteritic disease.
- ▶ Atypical features should prompt consideration for inflammatory marker investigation, steroid initiation and temporal artery biopsy.
- ▶ Lack of prompt treatment in arteritic ischaemic optic neuropathy may lead to permanent visual defects.

Contributors SL and RA write and developed the report and also drafted and revised the work.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

REFERENCES

- 1 Kerr NM, Chew SS, Danesh-Meyer HV. Non-arteritic anterior ischaemic optic neuropathy: a review and update. *J Clin Neurosci* 2009;16:994–1000.
- 2 Rucker JC, Biouesse V, Newman NJ. Ischemic optic neuropathies. *Curr Opin Neurol* 2004;17:27.
- 3 Arnold AC. Pathogenesis of nonarteritic anterior ischemic optic neuropathy. *J Neuroophthalmol* 2003;23:157.
- 4 Levin LA, Danesh-Meyer HV. Hypothesis: a venous etiology for nonarteritic anterior ischemic optic neuropathy. *Arch Ophthalmol* 2008;126:1582.
- 5 Hayreh SS. Ischemic optic neuropathy. *Prog Retin Eye Res* 2009;28:34–62.
- 6 Rizzo JF III, Lessell S. Optic neuritis and ischemic optic neuropathy. Overlapping clinical profiles *Arch Ophthalmol* 1991;109:1668.
- 7 Lee AG, Lin DJ, Kaufman M, *et al.* Atypical features prompting neuroimaging in acute optic neuropathy in adults. *Can J Ophthalmol* 2000;35:325.
- 8 Hayreh SS, Zimmerman B. Visual field abnormalities in nonarteritic anterior ischemic optic neuropathy: their pattern and prevalence at initial examination. *Arch Ophthalmol* 2005;123:1554.
- 9 [No authors listed]. Characteristics of patients with nonarteritic anterior ischemic optic neuropathy eligible for the Ischemic Optic Neuropathy. Decompression Trial. *Arch Ophthalmol* 1996;114:1366.
- 10 Bilgin G, Koban Y, Arnold AC. Nonarteritic anterior ischemic optic neuropathy and obstructive sleep apnea. *J Neuroophthalmol* 2013;33:232–4.
- 11 Palombi K, Renard E, Levy P, *et al.* Non-arteritic anterior ischaemic optic neuropathy is nearly systematically associated with obstructive sleep apnoea. *Br J Ophthalmol* 2006;90:879–82.
- 12 Arda H, Birer S, Aksu M, *et al.* Obstructive sleep apnoea prevalence in non-arteritic anterior ischaemic optic neuropathy. *Br J Ophthalmol* 2013;97:206–9.

Copyright 2014 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit <http://group.bmj.com/group/rights-licensing/permissions>.
BMJ Case Report Fellows may re-use this article for personal use and teaching without any further permission.

Become a Fellow of BMJ Case Reports today and you can:

- ▶ Submit as many cases as you like
- ▶ Enjoy fast sympathetic peer review and rapid publication of accepted articles
- ▶ Access all the published articles
- ▶ Re-use any of the published material for personal use and teaching without further permission

For information on Institutional Fellowships contact consortiasales@bmjgroup.com

Visit casereports.bmj.com for more articles like this and to become a Fellow