CASE REPORT

Two Cases of Segmental Disc Oedema and Normal Visual Acuity in Giant Cell Arteritis

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ABSTRACT

Giant cell arteritis (GCA) frequently involves the ocular circulation. Arteritic anterior ischemic optic neuropathy (AAION) is the most common presentation, producing severe vision loss with a characteristic waxy pallor of the optic disc. Optic disc ischemia in AAION is related to underlying systemic vasculitis, which must be treated aggressively with systemic steroids in order to minimize the risk of fellow eye involvement. In contrast, non-arteritic AION (NAION) is thought to result from localized hypoperfusion of the posterior ciliary arteries and mainly presents with segmental optic disc oedema. This condition is usually seen in individuals with crowded optic discs as well as predisposing vascular risk factors and does not require specific treatment. Distinguishing these two entities is important and often challenging. We describe two cases of biopsy-confirmed AAION presenting as segmental disc oedema with the absence of pallor, suggesting specific and isolated involvement of the posterior ciliary arteries in GCA affecting the ophthalmic circulation. This emphasizes the importance of maintaining a high index of suspicion for GCA, particularly when atypical features are present or systemic findings accompany acute vision loss.

ARTICLE HISTORY

Received 21 March 2018
Revised 15 June 2018
Accepted 15 June 2018

KEYWORDS

Giant cell arteritis; arteritic optic neuropathy; optic disc oedema; differential diagnosis

Giant cell arteritis (GCA) involves ophthalmic circulation in up to 70% of cases with arteritic anterior ischemic optic neuropathy (AAION) being the most common clinical manifestation. It is very important to distinguish non-arteritic AION (NAION) from AAION as AAION requires immediate initiation of high dose steroids to minimize the risk of fellow eye involvement.

The two entities can be differentiated based on a several clinical features. The pathophysiology of NAION is presumed to be hypoperfusion of the short posterior ciliary arteries causing ischemia of the optic nerve head. Most patients that develop this condition have minimal or no optic nerve cupping (“disc at risk”) with resulting crowding of the ganglion cell axons as they pass through the scleral canal.¹ AAION in contrast is caused by systemic vasculitis that affects medium- and large-sized arteries with predilection for extracranial branches of the carotid arteries but often involves ophthalmic branches of internal carotid artery including the short posterior ciliary arteries.^{2,3} The evidence for systemic vasculitis in

these patient is reflected by elevated serum inflammatory markers and systemic symptoms (temporal headache, jaw claudication, and scalp tenderness) in up to 75% of cases.² Diagnosis of AAION can be confirmed by temporal artery biopsy.

Pallid optic nerve oedema is a classic finding of AAION, while in NAION diffuse or segmental optic nerve head oedema with hemorrhages is seen acutely evolving to pallor over 4–6 weeks with resolution of swelling. Visual acuity at presentation is typically much worse in AAION with 38–54% of patients presenting with vision of hand motion or worse.^{3,4} AAION is also associated with subsequent development of optic disc cupping which is generally not seen in NAION⁵ and may be mistaken for glaucomatous optic neuropathy.

Here we describe two unusual cases of biopsy-confirmed AAION secondary to GCA presenting with preserved visual acuity and segmental disc oedema without pallor with corresponding nerve fibre bundle defect on formal visual field testing. These cases are unique in that likely only a subset of short posterior ciliary arteries supplying the optic

nerve head was involved by the arteritis process thus mimicking typical fundoscopy and visual findings of NAION. This serves as a reminder that atypical NAION deserves a thorough investigation.

In the first case, a 78-year-old man presented to an optometrist for “blurred vision” in the right eye and found to have an inferior visual field defect. Ophthalmological referral was made; patient was noted to have central acuity of 20/20 in each eye with the presence of right segmental optic nerve head swelling. Inflammatory markers demonstrated elevated CRP only (70 mg/dl, normal range 0–10 mg/dl) and normal erythrocyte sedimentation rate (ESR) and platelet count. Patient was started on 60 mg of oral prednisone a day. He was seen by our service 2 weeks later. He admitted to the presence of cheek and jaw pain at rest as well as after eating. On examination, visual acuity was 20/20 in each eye. There was right relative afferent pupillary defect (RAPD). There was moderate cupping of the right optic nerve head with cup-to-disc ratio of 0.7 and a superior neuro-retinal rim notch. There was no disc oedema or pallor. The left cup-to-disc ratio was 0.4 with no focal thinning or notching. Visual field testing (Humphrey 24-2 algorithm) demonstrated right inferior altitudinal defect (Figure 1).

In the second case, a 61-year-old woman presented to an emergency department with 1 week history of a sudden onset of right inferior altitudinal visual field defect upon waking and was referred to neuro-ophthalmology clinic. History was positive for 3 months of diffuse headache, neck and shoulder pain, jaw claudication, and 1 month of scalp tenderness. There was no fatigue,

malaise, or fever. On examination, visual acuity was 20/30 and 20/20. There was a subtle RAPD. There was a single cotton wool spot near the optic nerve on the right as well as segmental hyperemic superior optic nerve head oedema. Fundoscopy was normal on the left with cup to disc ratio of 0.4 (Figure 2). Visual field testing (Humphrey 24-2 algorithm) demonstrated right inferior altitudinal defect (Figure 3). CRP was mildly elevated at 11.1 mg/dl, ESR and complete blood count were normal. Contrast-enhanced MRI of the brain and orbits was performed and was unremarkable.

The differential diagnosis for both patients included NAION; however, neither demonstrated a typical “disc at risk” in the contralateral eye and more importantly both had some non-ocular symptoms of GCA indicating a systemic process. Both patients were started on oral prednisone (1 mg/kg dosing) and experienced a rapid resolution of their systemic symptoms including headache and jaw claudication. The visual field defects and central acuity remained stable over time, with no further deterioration and no involvement of the fellow eye. The diagnosis of GCA in both patients was confirmed after positive temporal artery biopsies (Figure 4).

We postulate that in both cases short posterior ciliary arteries were the only part of the ophthalmic circulation involved by GCA, which led to segmental hyperemic optic nerve head oedema and nerve fibre bundle defect on visual fields almost undistinguishable from NAION. While fluorescein angiography was not obtained in these cases, we speculate that choroidal perfusion would be normal as the central acuity was preserved in both patients. Since the

Figure 1. (A) 24-2 Humphrey visual field testing showing a right inferior nerve fibre bundle defect. (B) Optical coherence tomography (OCT) of the optic discs shows marked asymmetry, with superior thinning in the right eye.

Figure 2. Fundus photographs of the right and left eye. The right optic nerve head shows segmental hyperemic oedema superiorly. The absence of a “disc at risk” is notable in the left eye.

Figure 3. (A) 24-2 Humphrey visual field testing showing a right inferior field defect. (B) OCT of the optic discs shows segmental oedema in the superior region of the right optic nerve.

Figure 4. Temporal artery biopsies confirming giant cell arteritis. H&E (A) and elastic trichrome stain (black, B) in Case 1. Intimal thickening, stenosis and inflammatory infiltrate with giant cells (A, inset) are seen with disruption of the internal elastic lamina (B, inset). Case 2 (C, H&E) exhibited similar patchy inflammatory infiltrate and stenosis with severe disruption of the internal elastic lamina (inset). H&E: haematoxylin and eosin.

choroidal circulation supplies photoreceptors the acuity would be decreased if it had been involved by vasculitis. These two cases emphasize that fundoscopy and visual field abnormalities typically seen in

NAION are not specific in differentiating it from AAION.

This is a reminder that GCA of the ophthalmic circulation can selectively involve the short

posterior ciliary arteries thus mimicking NAION. Systemic symptoms should be inquired about and inflammatory markers always checked in patients with a presumed diagnosis of NAION.

Declaration of interest

The authors have no conflict of interest to disclose.

Funding

There are no funding sources to disclose.

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