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Vessels in full view: preserved vascular clarity in acute arteritic anterior ischaemic optic neuropathy

Stephen C. Pollock,[1] Anthony C. Arnold[2] and Neil R. Miller[3]

[1]Duke University Health System, Durham, North Carolina, USA
[2]Neuro-Ophthalmology Division, Department of Ophthalmology, University of California, Los Angeles, California, USA
[3]Neuro-Ophthalmology Division, Departments of Ophthalmology, Neurology & Neurosurgery, Johns Hopkins University School of Medicine, Baltimore, Maryland, USA

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Anterior ischaemic optic neuropathy (AION) due to giant cell arteritis (GCA) is a true ophthalmic emergency, requiring prompt diagnosis and treatment to prevent further catastrophic loss of vision. The purpose of this report is to share our observation of a heretofore unrecognized ophthalmoscopic sign of this condition: preserved clarity of retinal vessels overlying the acutely swollen optic disc. We conclude that the 'clear vessel sign' is a specific marker for arteritic AION, that it helps to distinguish arteritic from non-arteritic AION, and that its presence should alert clinicians to the likelihood of GCA and the need for urgent intervention. It also adds to the clinical picture of arteritic AION, thereby expanding and strengthening the diagnostic armamentarium of neuro-ophthalmologists and general ophthalmologists alike.

Over the last forty years, a consensus has developed among investigators that the pathogenesis of optic disc swelling in patients with non-arteritic AION involves interruption of axonal transport at the level of the distal optic nerve. Disruption of transport results in the accumulation of mitochondria, cytoskeletal proteins and other cytoplasmic elements within the prelaminar segments of ganglion cell axons. The pathologic distension of these axons leads to an increase in optic disc volume and to loss of the normal transparency of fibres overlying the disc. This latter change accounts for one of the defining ophthalmoscopic features of acute disc swelling in patients with non-arteritic AION: partial or complete obscuration of retinal vessels as they course across the disc and traverse its margins (Fig. 1).

We retrospectively reviewed photographic records of patients with biopsy-proven GCA who presented with acute visual loss and optic disc swelling. Ten representative images (Fig. 2) were selected for this report. In each case, the retinal vessels overlying the disc exhibit remarkable clarity. No obscuration is present, indicating preserved transparency of prelaminar ganglion cell axons. This finding is not a feature of acute non-arteritic AION or any other cause of acute optic disc swelling.

Among patients with arteritic AION who were evaluated by the authors, only a fraction underwent fundus photography. This, in conjunction with the limited size of our image databases and the retrospective nature of our review, makes it difficult to determine with accuracy the prevalence of the clear vessel sign among affected individuals. We nevertheless have been impressed by the ubiquity of the sign since becoming aware of it. Colour photographs of discs with a virtually identical appearance have been published previously (De Smit et al. 2016), though without reference to the clarity of the overlying retinal vessels.

We offer a preliminary hypothesis to account for this distinctive ophthalmoscopic feature, with the understanding that additional investigation will be needed to assess the validity of our proposed mechanism.

Giant cell arteritis can involve any branch of the ophthalmic artery, including the central retinal artery. Adequate perfusion of the retina is necessary to support anterograde axonal transport, a kinesin-mediated energy-dependent process. We suggest that in some individuals with acute arteritic AION, concurrent retinal hypoperfusion results in a shutdown of axonal transport within the retinal nerve fibre layer, specifically within those axon segments extending from ganglion cell bodies to the optic disc. Arrested transport of cellular organelles and other cargo to the disc would account for: (a) absence of distension of the prelaminar nerve fibres, and (b) retention of the normal

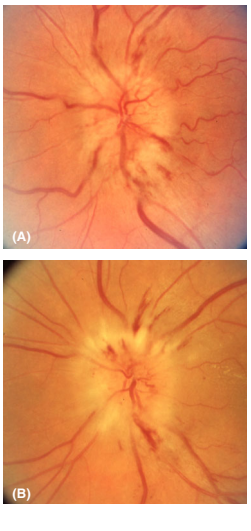

Fig. 1. (A,B) Images of acute optic disc swelling in two patients with non-arteritic AION. As is typical of disc swelling in this context, many of the superficial retinal vessels are indistinct and/or focally obscured.

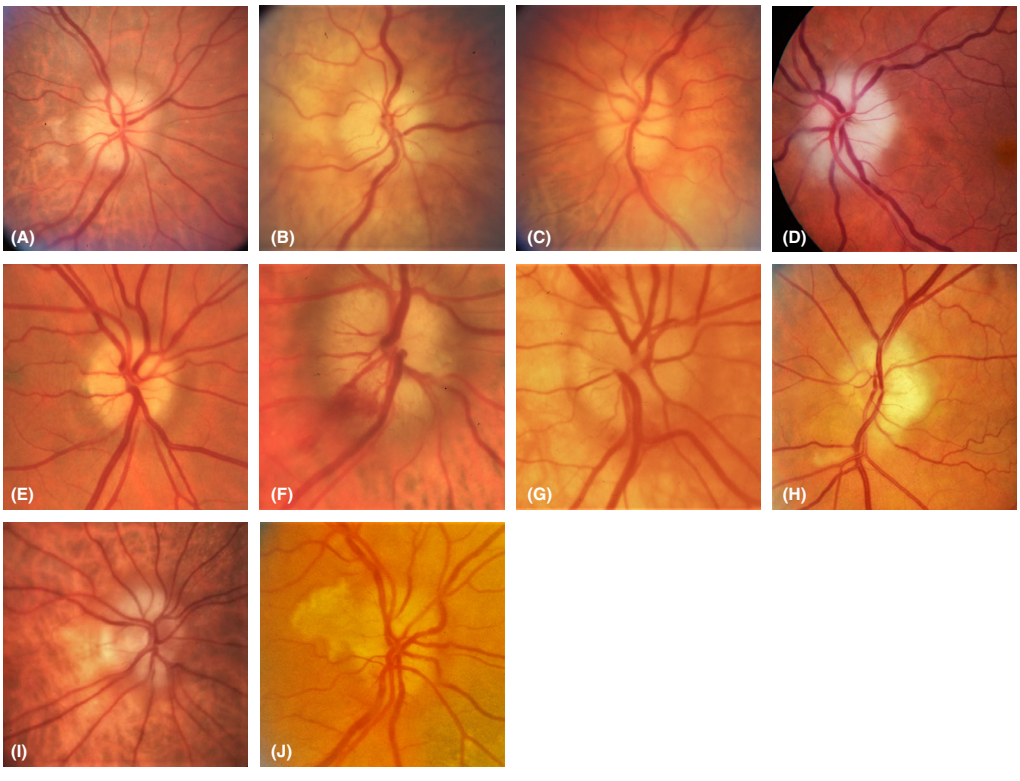

Fig. 2. (A–J) Images of acute optic disc swelling in ten eyes of patients with GCA and arteritic AION. Note that in each case, the retinal vessels coursing over the surface of the disc and its margins exhibit pristine clarity. In addition, no coarsening or blurring of the peripapillary nerve fibre layer is evident. The indistinct yellowish patches deep to the retina in images B and C likely represent areas of choroidal ischaemia. The disc in image E (reprinted with permission from Wolters Kluwer Health, Inc.) was photographed 1 day after the onset of acute visual loss to the level of hand motions. Fundus examination revealed rouleaux formation in the arteries and veins in combination with slow pulsatile flow. Fluorescein angiography demonstrated delayed appearance of dye in the retinal arterioles, a prolonged arteriovenous transit time, and absent fluorescence of the right optic disc. In image J, the whitish, cauliflower-shaped patch just temporal to the disc represents a cilioretinal artery occlusion.

transparency of those fibres. This, in turn, would explain the pristine view of retinal vessels traversing the disc and its margins.

In their seminal paper on the role of axonal transport in the pathophysiology of ischaemic disc swelling, McLeod et al. (1980) emphasized that the development of disc swelling after posterior ciliary artery occlusion is dependent on continued anterograde transport in the retinal nerve fibre layer and that such transport is itself contingent on sufficient retinal blood flow. In support of this, they pointed out that disc swelling is not observed in cases of combined occlusion of the posterior ciliary and central retinal vessels.

None of our cases was associated with a central retinal artery occlusion (CRAO). However, the absence of a CRAO should not be interpreted to mean that flow through the central retinal artery was normal. Retinal hypoperfusion exists along a spectrum. Indeed, virtually all patients with ocular ischaemic syndrome who undergo fluorescein angiography manifest evidence of profoundly reduced retinal perfusion, yet only 12% of these individuals ever develop a CRAO (Mendrinos et al. 2010). We hypothesize that a reduction in retinal blood flow insufficient to produce infarction may still exert a deleterious effect on axonal transport, particularly given the exquisite sensitivity of energy-dependent cellular processes to ischaemia and tissue hypoxia.

Assuming for the moment that axonal contents are *not* being delivered to the optic disc and that prelaminar ganglion cell fibres are *not* abnormally distended in patients with acute arteritic AION and concurrent retinal hypoperfusion, one might reasonably ask why the optic discs in Fig. 2 are swollen at all. We speculate that most or all of the visible disc swelling relates to the infarct itself. As axons within the ischaemic zone become vacuolated, leak cellular contents and undergo necrosis, the infarct expands to some degree. If a portion of the infarct is located anterior to the lamina cribrosa, expansion of that portion will be intraocular and will manifest as a pale swollen disc overlaid with strikingly distinct retinal vessels.

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Correspondence:
Stephen C. Pollock, MD
3005 Timberlyne Court
Mebane, NC 27302
USA
Tel: +1 336 578 2579
Email: spollock373@gmail.com

Dr. Pollock served as Chief of Neuro-Ophthalmology at Duke University and is now retired from academic practice.