

(<http://medicine.uiowa.edu/eye/>)

Ophthalmology and Visual Sciences
(<http://medicine.uiowa.edu/eye/>)

Eye  ROUNDS.org

([../index.htm](http://medicine.uiowa.edu/eye/..//index.htm))

Successive Presentation of Arteritic and Non-arteritic Anterior Ischemic Optic Neuropathy

Authors: Matthew Benage MD ([../bio/authors/Benage-Matthew.htm](http://medicine.uiowa.edu/eye/..//bio/authors/Benage-Matthew.htm)), Anthony T. Chung MD ([../bio/authors/Chung-Anthony.htm](http://medicine.uiowa.edu/eye/..//bio/authors/Chung-Anthony.htm)), Randy H. Kardon MD, PhD ([../bio/authors/Kardon-Randy.htm](http://medicine.uiowa.edu/eye/..//bio/authors/Kardon-Randy.htm))

Posted July 23, 2018

INITIAL PRESENTATION

Chief Complaint

Headache with scalp pain

Initial History of Present Illness

An 84-year-old gentleman was referred to the Veteran's Affairs eye clinic for a temporal artery biopsy in the setting of possible giant cell arteritis (GCA). He had a 2-year history of polymyalgia rheumatica (PMR) and a long history of seronegative rheumatoid arthritis (RA). At a recent rheumatology appointment, he complained of a headache for the past two months. He reported the pain was located behind his left eye and was associated with scalp tenderness while combing his hair. He denied jaw claudication, fever, weight loss, or vision changes, including diplopia.

Given his symptoms, his rheumatologist elected to draw inflammatory markers and both returned elevated. Erythrocyte sedimentation rate (ESR) was 48 mm/hr (normal range 0-15 mm/hr) and C-reactive protein (CRP) was 0.52 mg/dL (normal range 0.0-0.5 mg/dL). His symptoms and elevated inflammatory markers were concerning for GCA, and he was placed on 60 mg prednisone.

At his appointment in the eye clinic that week, he stated that his acuity was at baseline. He reported mild improvement in his headache since starting systemic steroids.

Past Ocular History

- Refractive error

- Cataract surgery, both eyes (OU) 2003
- Mild primary open angle glaucoma on Latanoprost every night OU

Past Medical History

- Hypertension
- Hyperlipidemia
- Diabetes mellitus type 2
- Polymyalgia rheumatica (PMR)
- Seronegative rheumatoid arthritis (RA)
- Obstructive sleep apnea (OSA); he was not using a continuous positive airway pressure (CPAP) machine at bedtime

Medications

- Hydroxychloroquine 400 mg/day for 10+ years
- Amlodipine 5 mg daily
- Losartan 100 mg daily

Allergies

None

Family History

Grandfather with glaucoma

Social History

Retired factory inspector. No known smoking history

Review of Systems

- As per the above HPI, all other symptoms negative

OCULAR EXAMINATION

- Visual acuity (VA) with correction (Snellen)
 - Right eye (OD): 20/20
 - Left eye (OS): 20/40
- Extraocular motility
 - OD: Full
 - OS: Full
- Intraocular pressure (IOP) by Tonopen
 - OD: 20 mmHg
 - OS: 20 mmHg
- Pupils
 - OD: 4 mm in dark, 3 mm in light, no relative afferent pupillary defect (RAPD)
 - OS: 4 mm in dark, 3 mm in light, no RAPD
- Confrontational visual fields

- OD: Full to count fingers
- OS: Full to count fingers
- Slit lamp examination
 - External: Normal OU
 - Lids: Normal OU
 - Conjunctiva/sclera: Normal OU
 - Cornea: Normal OU
 - Anterior chamber: Normal OU
 - Iris: Normal OU
 - Lens: Posterior chamber intraocular lens (PCIOL) OU
- Dilated fundus examination
 - Disc: Peripapillary atrophy, no disc edema or pallor OU
 - Cup to disc: 0.3 OD, 0.15 OS
 - Macula: Small, hard drusen OU
 - Vessels: Normal OU
 - Periphery: Normal OU

Additional testing

- Temporal artery biopsy was performed, and there was no histological evidence of inflammation.

INTERVAL CLINICAL COURSE

Given his benign ocular exam without evidence of arteritic anterior ischemic optic neuropathy (A-AION), he returned to rheumatology for continued management of his steroids and inflammatory diseases.

Seven months after original presentation the patient returned to the ophthalmology clinic complaining of decreased vision in the left eye for 1 week. He stated the lower half of vision in the left eye was "cloudy." He denied floaters, photopsias, diplopia, scalp tenderness, or jaw claudication. He had been tapered to 10 mg of prednisone per day over the previous 7 months.

OCULAR EXAMINATION

- Visual acuity with Correction
 - OD: 20/20
 - OS: 20/60
- Extraocular Motility
 - OD: Full
 - OS: Full
- Intraocular pressure (IOP) by Tonopen
 - OD: 18 mmHg
 - OS: 19 mmHg
- Pupils
 - OD: 4 mm in dark, 3 mm in light, no RAPD
 - OS: 4 mm in dark, 3 mm in light, 1.2 log unit RAPD
- Confrontational visual fields
 - OD: Full to count fingers
 - OS: Inferior altitudinal defect
- Slit lamp examination

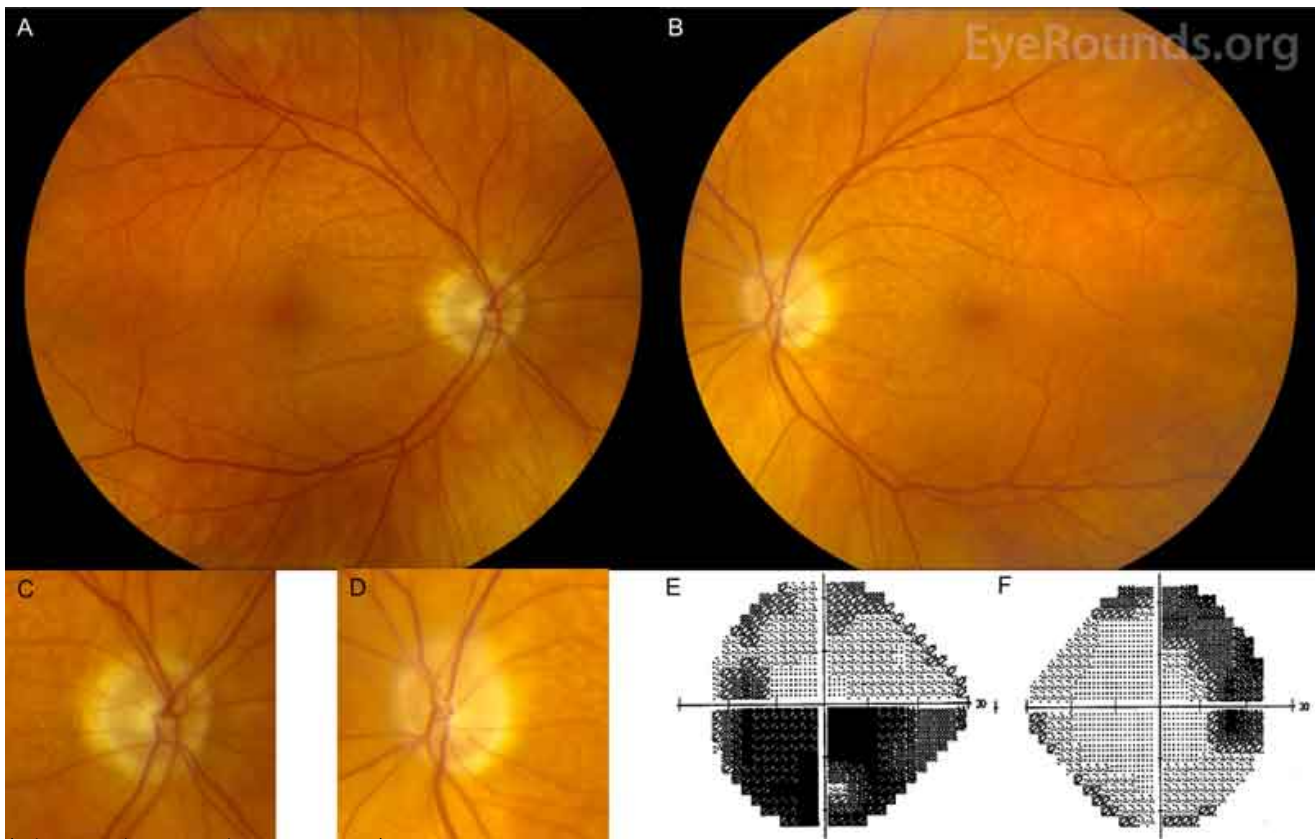
- External: Normal OU
- Lids: Normal, no ptosis OU
- Conjunctiva/sclera: Normal OU
- Cornea: Normal OU
- Anterior chamber: Normal OU
- Iris: Normal OU
- Lens: PCIOL OU
- Dilated fundus examination (Figure 1)
 - Disc: Peripapillary atrophy OU
 - OD 0.3 c/d, no disc edema or pallor
 - OS: 0.15 c/d, superior pallor and edema
 - Macula: Small, hard drusen OU
 - Vessels: Normal OU
 - Periphery: Normal OU

Additional testing

- Humphrey automated visual field test 24-2 (Figure 1)
- Optic nerve head optical coherence tomography (OCT) (Figure 2)
- Fluorescein angiography (Figure 3)
- Laser speckle flowgraphy also showed corresponding significantly reduced choroidal blood flow, further raising the suspicion of arteritic anterior ischemic optic neuropathy.
- ESR was 7 mm/hr, and CRP <0.15 mg/dL.
- Repeat temporal artery biopsy returned positive for healing phase of arteritis, without sign of active inflammation.

Differential Diagnosis for Altitudinal Visual Field Defects

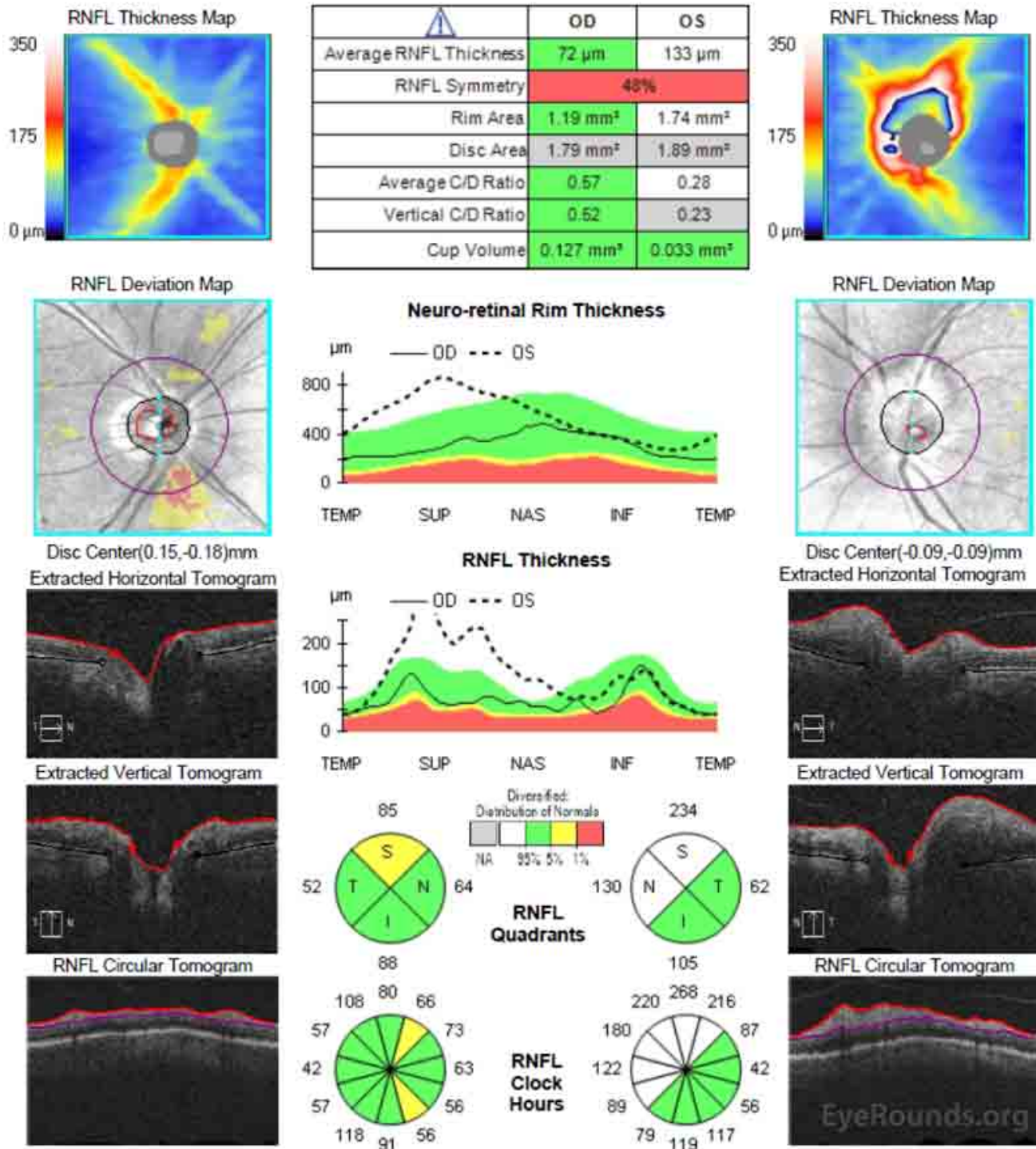
- Arteritic anterior ischemic optic neuropathy (A-AION)
- Non arteritic anterior ischemic optic neuropathy (NAION)
- Idiopathic optic neuritis
- Optic nerve inflammation (i.e. related to syphilis or sarcoidosis)
- Infiltrative optic neuropathy



(../cases-i/case276/Fig1-LRG.jpg)

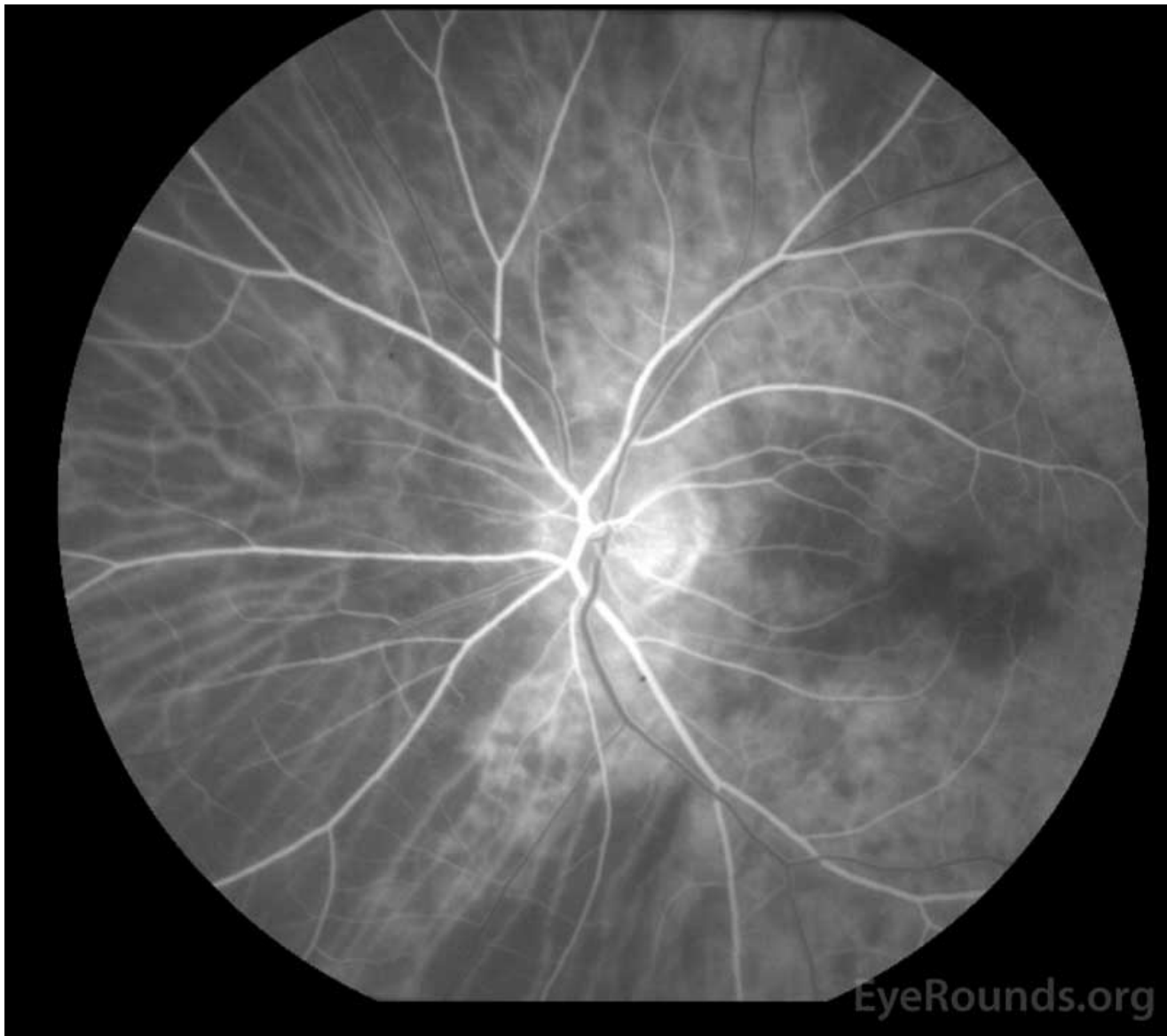
Figure 1: Color fundus photography and Humphrey visual field (HVF) testing, both eyes: A) Right eye has peripapillary atrophy and small macular drusen. B) Left eye had peripapillary atrophy, superior optic nerve pallor with trace edema, and small macular drusen. Higher magnification of the right optic nerve (C) and the left optic nerve (D) are shown. E) HVF testing of the left eye shows an inferior altitudinal defect, which is consistent with an ischemic optic neuropathy. F) HVF testing of the right eye demonstrates a superior arcuate defect extending to blind spot. This is consistent with the patient's previous glaucoma exams.

ONH and RNFL OU Analysis: Optic Disc Cube 200x200 OD ● OS



(./cases-i/case276/Fig2-LRG.jpg)

Figure 2: Optic nerve head optical coherence tomography (OCT): There is normal retinal nerve fiber layer thickness with minimal superior thinning OD. The left optic nerve head has increased thickness in the superior and nasal quadrants, which is consistent with disc edema.



(../cases-i/case276/Fig3-LRG.jpg)

Figure 3: Fluorescein angiography, OS: Left eye fluorescein angiogram shows the lack of choroidal filling in the distribution of the lateral posterior ciliary artery.

DIAGNOSIS

Arteritic anterior ischemic optic neuropathy (A-AION) with posterior ciliary artery occlusion of the left eye

Etiology/Epidemiology

A-AION is almost exclusively due to GCA. It can present in patients over the age of 55 years old but presents most commonly in those over the age of 70 years old. Women are three times more likely than men to suffer from an A-AION, and there is a strong racial predisposition for Caucasians [1].

Pathophysiology

Giant cell arteritis is a systemic, granulomatous vasculitis that primarily affects medium and large arteries. In the case of A-AION, occlusion of the posterior ciliary artery is the most common cause of ischemia.

Signs/Symptoms

Imaging

Fluorescein fundus angiography in A-AION generally displays delayed filling of the optic nerve and choroid due to involvement of the posterior ciliary arteries. This is in contrast to NAION, which may display only nerve early filling defects but not choroidal filling delays beyond those in the choroidal watershed zone. In some cases, magnetic resonance imaging (MRI) with gadolinium contrast may reveal periarterial enhancement around temporal and/or occipital arteries, although this is not routinely obtained. High frequency ultrasound has also demonstrated high reflectivity in the wall of the temporal artery corresponding to areas of active inflammation [5,6].

Treatment/Management/Guidelines

The main goals of treatment of A-AION are to prevent worsening in the involved eye as well as involvement of the contralateral eye. If left untreated, 54-95% of patients present with ischemia in the contralateral eye, typically within four months [7]. The use of steroids reduces contralateral involvement to approximately 15% [4]. Therefore, when clinical suspicion for A- AION is high, prompt high dose prednisone (60-100 mg/day; 1 mg/kg per day) should be instituted with the goal of obtaining a temporal artery biopsy within one to two weeks. Generally, there is clinical improvement after initiation of steroids, thus, if there is no response, an alternative diagnosis should be considered.

In the case of a positive biopsy, steroid tapering strategies in GCA are variable. However, a few general principles can guide therapy. GCA is considered controlled when visual symptoms are stable, constitutional symptoms have resolved, and the ESR and CRP have normalized. The inflammatory response, particularly ESR, may take several weeks on treatment to display improvement. Since GCA is a relapsing condition, there is often a need to increase the steroids during the tapering process. The taper can start with a decrease of 10 mg per month, followed by 5 mg per month, and then to an increment as small as 1 mg per month. This process is individualized, and it may take 1-2 years for a complete taper. In some cases, the patient may require low dose steroids indefinitely [8,9].

Recently, pro-inflammatory cytokines such as interleukin 6 (IL-6) have been shown to play an important role in the pathophysiology of GCA. The Food and Drug Administration (FDA) recently approved Tocilizumab, which is a humanized monoclonal anti-IL-6 receptor antibody that binds both soluble and membrane bound IL-6 receptors. It works by inhibiting IL-6 signal transduction. Tocilizumab is administered as a monthly infusion of either 4 or 8 mg/kg. This has the potential to reduce the steroid dose and duration needed to treat GCA [10,11].

Further discussion of GCA can be found here: <http://EyeRounds.org/article/GCA/> (../article/GCA/)

INTERVAL CLINICAL COURSE

The patient was placed on 60 mg oral prednisone per day and was followed with a slow steroid taper as his symptoms improved and inflammatory markers normalized. At a scheduled appointment 8 months after initiation of steroids, he complained of a "gray bar" in the inferonasal corner of his right eye. He noticed the change upon waking up the previous morning. At this point, he had been tapered to 30 mg of prednisone. He denied any eye or head pain and had no systemic symptoms.

OCULAR EXAMINATION

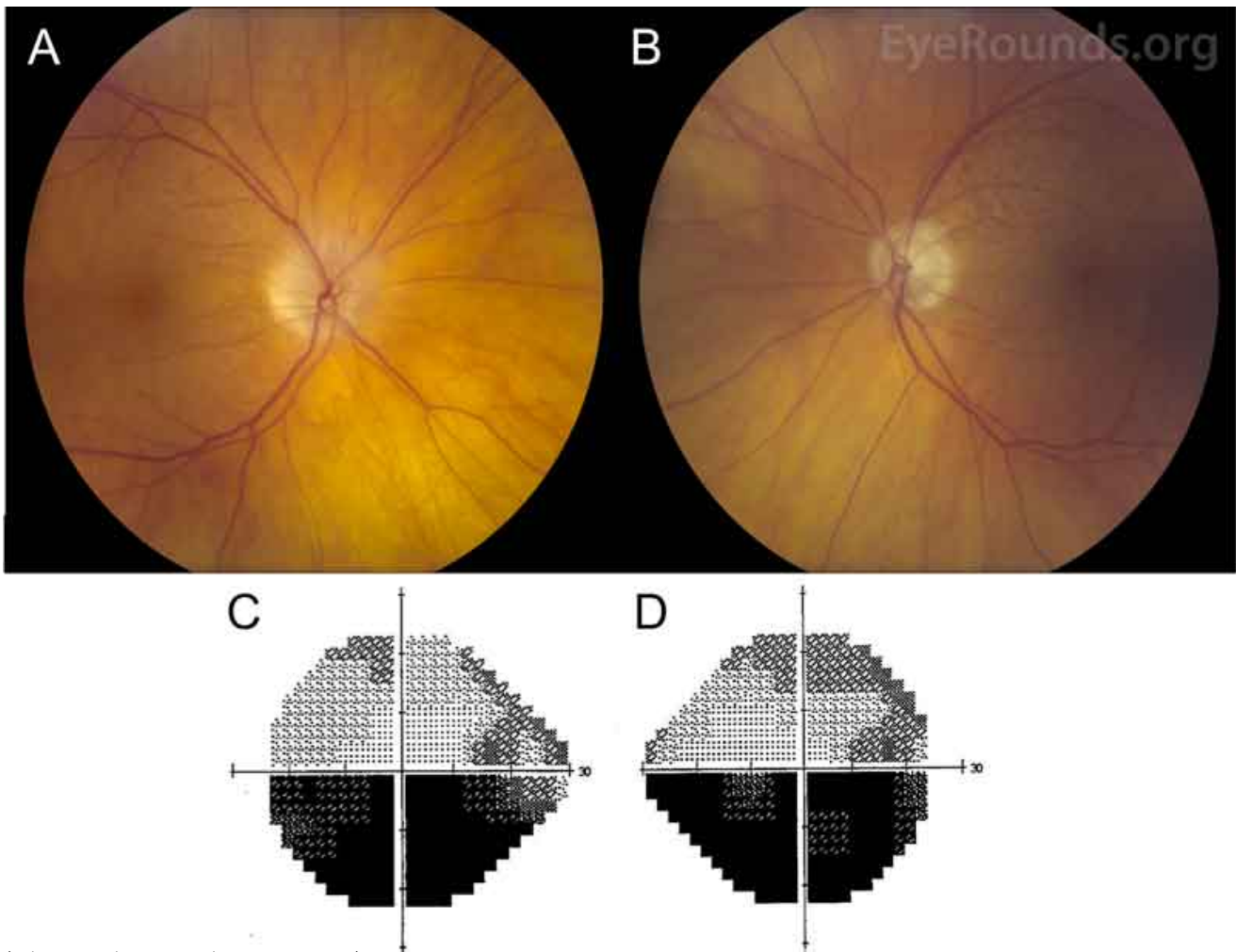
- Visual acuity with correction
 - OD: 20/30
 - OS: 20/50

- Extraocular motility
 - OD: Full
 - OS: Full
- Intraocular pressure (IOP) by Tonopen
 - OD: 16 mmHg
 - OS: 17 mmHg
- Pupils
 - OD: 4 mm in dark, 3 mm in light, no RAPD
 - OS: 4 mm in dark, 3 mm in light, 0.9 log unit RAPD
- Confrontational visual fields (CVF)
 - OD: Inferior altitudinal defect
 - OS: Inferior altitudinal defect
- Slit lamp examination
 - External: Normal OU
 - Lids: Normal, no ptosis OU
 - Conjunctiva/sclera: Normal OU
 - Cornea: Normal OU
 - Anterior chamber: Normal OU
 - Iris: Normal OU
 - Lens: PCIOL OU
- Dilated fundus examination (Figure 4)
 - Disc: Peripapillary atrophy OU
 - OD: Superior hyperemic edema
 - OS: Superior disc pallor; increased cupping
 - Macula: Small, hard drusen OU
 - Vessels: Normal OU
 - Periphery: Normal OU

Additional testing

- HVF 24-2 (Figure 4)
- Repeat fluorescein angiography and laser speckle blood flow both showed no evidence of a posterior ciliary artery occlusion in the right eye, although the tests did demonstrate delay in optic disc filling of the right eye.

ESR was 10 mm/hr and CRP was 0.19 mg/dL, which were both within normal limits



(../cases-i/case276/fig4-LRG.jpg)

Figure 4: Color fundus photography and Humphrey visual field (HVF) testing, both eyes: A) Right eye has peripapillary atrophy with notable superior hyperemic optic nerve edema and small macular drusen. B) Left eye had peripapillary atrophy, superior optic nerve pallor with trace edema, and small macular drusen. C) HVF testing of the left eye shows a stable inferior altitudinal defect. D) HVF testing of the right eye demonstrates superior arcuate defect extending to blind spot as well as a new inferior altitudinal defect.

DIAGNOSIS

Non arteritic anterior ischemic optic neuropathy (NAION), right eye

Etiology/Epidemiology

NAION is the most common ischemic optic neuropathy with an incidence of about 10 per 100,000 [12]. Risk factors for developing NAION can be either systemic or ocular in nature.

Systemic risk factors include [13]:

- Arterial hypertension
- Diabetes mellitus
- Ischemic heart disease
- Hyperlipidemia
- Atherosclerosis and arteriosclerosis
- Obstructive sleep apnea
- Arterial hypotension/Nocturnal arterial hypotension
- Migraines

Ocular risk factors include:

- Absent or small cup of the optic nerve
- Angle closure glaucoma or other causes of markedly raised IOP
- Watershed zone of the posterior ciliary arteries that includes all or a portion of the optic disc
- Optic disc drusen

Pathophysiology [14]

NAION is due to significant ischemia of the optic nerve head secondary to hypoperfusion from the posterior ciliary artery. Considerable investigation has been undertaken to understand the precise mechanism of ischemia. Two mechanisms predominate in our understanding of the pathophysiology [15]. The first is relative hypoperfusion due to drop in blood pressure, which can occur during shock or during sleep (*i.e.*, nocturnal arterial hypotension). This drop in perfusion pressure is thought to lead to relative hypoperfusion of the optic nerve head. Nocturnal hypotension, in particular, is noted as a risk factor. In up to 73% of cases of NAION, the initial vision loss is noted upon waking. The choroidal vascular watershed zone between the lateral and medial posterior ciliary arteries appears to be particularly at risk during these nocturnal hypotensive events. The second and much less common mechanism involves embolic events causing optic nerve head ischemia [14,16].

Signs/Symptoms

NAION generally presents in elderly patients as a sudden, painless, monocular worsening of vision, oftentimes noted upon awakening. At presentation, one third of patients will have vision of 20/20 or better and 51% of patients will have vision of 20/40 or better [17-19]. On presentation of an acute NAION, the optic nerve displays disc edema with hyperemia with splinter hemorrhages commonly found at the margin. The optic nerve begins to display pallor two to three weeks after onset, with resolution of disc edema around two months later. It is common for the fellow eye to have a small or absent cup. This is frequently referred to as a "disc at risk," because upon swelling from ischemia, a compartment syndrome may develop and cause further ischemic damage.

The most common visual field defects at presentation are inferior nasal or inferior altitudinal defects, although superior, central, or cecocentral defects can also be seen. A relative afferent pupillary defect is noted in monocular involvement. The natural history of NAION is that 41-43% of patients have improvement in their visual fields, 15-19% have visual field worsening, and approximately 40% have stability of their visual field at 6 months. The occurrence of NAION in the fellow eye is estimated to be about 20% within ten years, with diabetics having a higher risk for involvement of the second eye [15,18].

Testing/Laboratory work-up

Imaging

Fluorescein fundus angiography displays a filling delay at the optic nerve head or defect in the prelaminar or peripapillary choroid in the early stage of NAION. The delay is noted to be at least five seconds in most cases; however, peripapillary choroidal filling is not delayed, which is in contrast to A-AION [20]. Late in the course of the angiogram, there is optic disc staining, a non-specific finding secondary to disc edema. Laser speckle flowgraphy shows a choroidal watershed zone of decreased blood flow but no significant reduction in blood flow compared to normal eyes measured during waking hours. Following retinal nerve fiber layer atrophy, the blood flow to the optic nerve head is reduced.

Treatment/Management/Guidelines

Currently there is no universally agreed upon treatment for NAION. High dose oral prednisone use during the first two weeks of vision loss has been shown to be of benefit in some patients with a significant improvement in disc edema and visual outcome; however, its use in clinical practice is controversial [18,16]. Avastin and aspirin have been studied as possible treatment interventions with no clear evidence for benefit. However, modifiable risk factors, when applicable, should be addressed. For example, patients should be advised to avoid nocturnal hypotension by moving anti-hypertensive medication administration times to earlier in the day, to appropriately diagnose and treat obstructive sleep apnea, to mitigate diabetes, and to avoid vaso-dilators before bedtime, such as those used for erectile dysfunction disorder, insomnia, or pain.

Further discussion on the pathophysiology of NAION can be found here:

<http://EyeRounds.org/article/AION/AION-part2.htm> (../article/AION/AION-part2.htm)

Characteristic	AAION	NAION
Age	Mean: 70 yrs	Mean: 60 yrs
Sex	F>M	F=M
Associated Symptoms	<ul style="list-style-type: none"> • Headache • Scalp tenderness • Jaw claudication • Transient visual loss 	None
Visual Acuity	<20/200 in >75%	>20/200 in >60%
Visual Field	Diffuse = Altitudinal	Altitudinal most common
Fundus	<ul style="list-style-type: none"> • Pallid edema • No "disc at risk" • Choroidal ischemia 	<ul style="list-style-type: none"> • Hyperemic or pallid edema • Bilateral "disc at risk" • No choroidal ischemia
Natural History	<ul style="list-style-type: none"> • Rarely improves • Fellow eye in 54-95% 	<ul style="list-style-type: none"> • Up to 40% improve • Fellow eye in ~ 20%
Fluorescein angiography	Disc and choroidal filling defect	Disc filling delay

Table 1: Differentiating A-AION and NAION

A-AION

<p>EPIDEMIOLOGY OR ETIOLOGY</p> <ul style="list-style-type: none"> • Occurs almost exclusively in patients > 55 years of age • 3:1 female to male predominance • Systemic vasculitis that primarily affects the medium and large arteries (again, most notably the posterior ciliary artery) 	<p>SIGNS</p> <ul style="list-style-type: none"> • Chalky white optic nerve may or may not be present with circumferential edema in the acute phase • Optic nerve cupping and pallor in the sub-acute phase • Cotton wool spots • Cilioretinal artery occlusion in conjunction with optic disc edema • Central retinal artery occlusion with lateral posterior ciliary artery occlusion • Choroidal ischemia • Fluorescein fundus angiography in arteritic AION generally displays very delayed or loss of choroidal filling in the distribution of one or more of the posterior ciliary arteries • Elevated ESR/CRP
<p>SYMPTOMS</p> <ul style="list-style-type: none"> • Profound vision loss • Amaurosis fugax may precede vision loss, especially with orthostatic drops in blood pressure • Fevers, chills, night sweats • Anorexia • Jaw claudication • Scalp tenderness • Pain of the proximal muscles • General malaise 	<p>TREATMENT/MANAGEMENT</p> <ul style="list-style-type: none"> • The main goal of treatment of A-AION is to prevent contralateral involvement • Treatment should start immediately and should not be delayed by the biopsy • High dose prednisone (60-100 mg/day) with slow taper monitoring for symptoms and lab results • Temporal artery biopsy within 1-2 weeks for definitive diagnosis

NAION

<p>EPIDEMIOLOGY OR ETIOLOGY</p> <ul style="list-style-type: none"> • Incidence: 10 per 100,000 • Acute ischemia of the optic nerve head, most notably via the posterior ciliary artery • Risk factors: hypertension, hypotension (especially nocturnal), diabetes, ischemic heart disease, obstructive sleep apnea, migraines • Ocular risk factors: small optic discs, disc edema of any etiology, optic disc drusen, cataract extraction, elevated IOP 	<p>SIGNS</p> <ul style="list-style-type: none"> • Disc edema, commonly with hyperemia • Contralateral optic nerve with small or absent cup ("disc at risk") • Altitudinal field defect on visual field testing • Fluorescein fundus angiography displays filling delay or defect in the prelaminar or peripapillary choroid
---	--

SYMPTOMS

- Sudden, painless worsening of vision
- Often presents upon awakening

TREATMENT/MANAGEMENT

- No current consensus for treatment, but evidence shows that high dose prednisone started during the first two weeks of vision loss may be beneficial
- Avastin and aspirin use have not been shown to improve outcome or reduce future risk of fellow eye involvement
- Management and reduction of systemic risk factors

References

1. Arnold AC. Ischemic optic neuropathies. *Ophthalmol Clin North Am* 2001;14(1):83-98. <https://PubMed.gov/11370574> (<https://PubMed.gov/11370574>)
2. Hayreh SS, Podhajsky PA, Raman R, Zimmerman B. Giant cell arteritis: validity and reliability of various diagnostic criteria. *Am J Ophthalmol* 1997;123(3):285-296. <https://PubMed.gov/9063237> (<https://PubMed.gov/9063237>)
3. Hayreh SS, Podhajsky PA, Zimmerman B. Ocular manifestations of giant cell arteritis. *Am J Ophthalmol* 1998;125(4):509-520. <https://PubMed.gov/9559737> (<https://PubMed.gov/9559737>)
4. Hayreh SS, Zimmerman B. Management of giant cell arteritis. Our 27-year clinical study: new light on old controversies. *Ophthalmologica* 2003;217(4):239-259. <https://PubMed.gov/12792130> (<https://PubMed.gov/12792130>). DOI: 70631
5. Alberts MS, Mosen DM. Diagnosing temporal arteritis: duplex vs. biopsy. *Qjm* 2007;100(12):785-789. <https://PubMed.gov/18089544> (<https://PubMed.gov/18089544>). DOI: 10.1093/qjmed/hcm103
6. Klink T, Geiger J, Both M, Ness T, Heinzelmann S, Reinhard M, Holl-Ulrich K, Duwendag D, Vaith P, Bley TA. Giant cell arteritis: diagnostic accuracy of MR imaging of superficial cranial arteries in initial diagnosis-results from a multicenter trial. *Radiology* 2014;273(3):844-852. <https://PubMed.gov/25102371> (<https://PubMed.gov/25102371>). DOI: 10.1148/radiol.14140056
7. Liu GT, Glaser JS, Schatz NJ, Smith JL. Visual morbidity in giant cell arteritis. Clinical characteristics and prognosis for vision. *Ophthalmology* 1994;101(11):1779-1785. <https://PubMed.gov/7800356> (<https://PubMed.gov/7800356>)
8. Fraser JA, Weyand CM, Newman NJ, Biousse V. The treatment of giant cell arteritis. *Rev Neurol Dis* 2008;5(3):140-152. <https://PubMed.gov/18838954> (<https://PubMed.gov/18838954>)
9. Collinson N, Tuckwell K, Habeck F, Chapman M, Klearman M, Stone JH. Development and implementation of a double-blind corticosteroid-tapering regimen for a clinical trial. *Int J Rheumatol* 2015;2015:589841. <https://PubMed.gov/25878667> (<https://PubMed.gov/25878667>). DOI: 10.1155/2015/589841
10. Tamaki H, Hajj-Ali RA. Tocilizumab for Giant Cell Arteritis-A New Giant Step in an Old Disease. *JAMA Neurol* 2017;10.1001/jamaneurol.2017.3811. <https://PubMed.gov/29255889> (<https://PubMed.gov/29255889>). DOI: 10.1001/jamaneurol.2017.3811
11. Milman N. Tocilizumab increased sustained glucocorticoid-free remission from giant cell arteritis. *Ann Intern Med* 2017;167(12):Jc63. <https://PubMed.gov/29255853> (<https://PubMed.gov/29255853>). DOI: 10.7326/acpjc-2017-167-12-063
12. Hattenhauer MG, Leavitt JA, Hodge DO, Grill R, Gray DT. Incidence of nonarteritic anterior ischemic optic neuropathy. *Am J Ophthalmol* 1997;123(1):103-107. <https://PubMed.gov/9186104> (<https://PubMed.gov/9186104>)
13. Hayreh SS. Anterior Ischemic Optic Neuropathy: Part II: a discussion for physicians . EyeRounds.org. Posted August 24, 2016; Available from <http://eyerounds.org/article/AION/AION-part2.htm> (./article/AION/AION-part2.htm)

In a study of 363 patients, the most significant symptoms associated with GCA included jaw claudication, neck pain, and anorexia [2]. Additional symptoms include headache, scalp tenderness, postural amaurosis fugax, weight loss, myalgia of proximal girdle muscles, malaise, and diplopia, if the extraocular muscles become ischemic. In this same series by Dr. Sohan Hayreh, it was noted that 21% of patients presented with visual loss due to GCA but had no systemic symptoms. This is termed occult giant cell arteritis. In up to 76% of patients with A-AION, the presenting vision is worse than 20/200. The nerve in acute A-AION may present with chalky white edema. Following the acute phase of edema, the nerve becomes pale with increased cupping, which may be confused with glaucomatous changes. Additional fundoscopic changes may include cotton wool spots, central retinal artery occlusion, and choroidal ischemia [2,3].

Testing/Laboratory work-up

If there is any concern for GCA, a prompt work-up should include the following: ESR, CRP, and CBC with differential (particularly platelets). While ESR has commonly been considered the most sensitive marker for GCA, evidence suggests that CRP may be a more reliable test. A combination of CRP and ESR elevation leads to a diagnostic specificity of 97% in patients where there is concern for GCA [4]. Common associated hematologic abnormalities include thrombocytosis, anemia, and leukocytosis.

Temporal Artery Biopsy

A temporal artery biopsy (TAB) should be obtained for tissue diagnosis. Unfortunately, TABs carry a relatively high rate of both false positives and false negatives. Histological findings consistent with GCA include intimal thickening, internal limiting lamina fragmentation, and chronic inflammatory cells, including giant cells and lymphocytes. In order to have sufficient tissue specimen biopsies should be at least two centimeters in length. The pathologist will perform serial sectioning to avoid a false negative result due to the phenomenon of discontinuous arteriolar involvement, which leads to skip lesions.

from [University of Iowa Ophthalmology](#)

02:49



If video fails to load, use this link: <https://vimeo.com/123352772> (<https://vimeo.com/123352772>)

14. Miller NR, Arnold AC. Current concepts in the diagnosis, pathogenesis and management of nonarteritic anterior ischaemic optic neuropathy. *Eye (Lond)* 2015;29(1):65-79. <https://PubMed.gov/24993324> (<https://PubMed.gov/24993324>). DOI: 10.1038/eye.2014.144
15. Arnold AC. Pathogenesis of nonarteritic anterior ischemic optic neuropathy. *J Neuroophthalmol* 2003;23(2):157-163. <https://PubMed.gov/12782932> (<https://PubMed.gov/12782932>).
16. Hayreh SS. Non-arteritic anterior ischemic optic neuropathy: role of systemic corticosteroid therapy. *Surv Ophthalmol* 2010;55(4):399-400; author reply 400-391. <https://PubMed.gov/20682167> (<https://PubMed.gov/20682167>). DOI: 10.1016/j.survophthal.2010.03.003
17. Norregaard JC, Gerner N, Jensen OA, Prause JU. Malignant melanoma of the conjunctiva: occurrence and survival following surgery and radiotherapy in a Danish population. *Graefes Arch Clin Exp Ophthalmol* 1996;234(9):569-572. <https://PubMed.gov/8880155> (<https://PubMed.gov/8880155>).
18. Hayreh SS. Corticosteroid Therapy in Nonarteritic Anterior Ischemic Optic Neuropathy. *J Neuroophthalmol* 2017;37(3):349-350. <https://PubMed.gov/28759499> (<https://PubMed.gov/28759499>). DOI: 10.1097/wno.0000000000000557
19. Hayreh SS, Zimmerman MB. Nonarteritic anterior ischemic optic neuropathy: natural history of visual outcome. *Ophthalmology* 2008;115(2):298-305.e292. <https://PubMed.gov/17698200> (<https://PubMed.gov/17698200>). DOI: 10.1016/j.opthta.2007.05.027
20. Arnold AC, Badr MA, Hepler RS. Fluorescein angiography in nonischemic optic disc edema. *Arch Ophthalmol* 1996;114(3):293-298. <https://PubMed.gov/8600889> (<https://PubMed.gov/8600889>).

Suggested citation format

Benage M, Chung AT, Kardon RH. Successive Presentation of Arteritic and Non-arteritic Anterior Ischemic Optic Neuropathy. EyeRounds.org. Posted July 23, 2018; available at <http://eyerounds.org/cases/276-AAION-NAION.htm>

last updated: 07/23/2018

Share this page:

University of Iowa Carver College of Medicine
Department of Ophthalmology & Visual Sciences
200 Hawkins Dr.
Iowa City, IA 52242

Web Privacy Policy (<http://www.uiowa.edu/homepage/online-privacy-information>) | Nondiscrimination Statement (<http://opsmanual.uiowa.edu/community-policies/nondiscrimination-statement>)

 THE UNIVERSITY OF IOWA
(<http://www.uiowa.edu>)

Directory (<https://www.dna.its.uiowa.edu/Whitepages/>) | A-Z Search (<http://www.uiowa.edu/a-z>) |
About Iowa (<http://www.uiowa.edu/homepage/about-university>) | Contact Us
(<http://www.uiowa.edu/homepage/about-university>) | Calendars

**(<http://www.uiowa.edu/homepage/calendars>) | Privacy Information
(<http://www.uiowa.edu/homepage/online-privacy-information>)**

copyright ©2016 The University of Iowa.