ANTERIOR ISCHEMIC OPTIC NEUROPATHY

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ANTERIOR ISCHEMIC OPTIC NEUROPATHY

- Anterior ischemic optic neuropathy (AION) is the most common acute optic neuropathy in patients more than 50 years of age.
- Patients experience painless monocular vision loss that develops over hours to days.
- Visual acuity may be diminished, but visual field loss always occurs; altitudinal and other variants of arcuate defects are most common, although any defect may also occur.
- An RAPD is present unless the optic neuropathy is bilateral. Optic disc edema develops at onset and may precede the vision loss.
- AION is classified as either arteritic (AAION), in which case it is associated with vasculitis, most commonly giant cell arteritis (GCA), or nonarteritic (NAION).

Table 4-4 Arteritic Versus Nonarteritic Ischemic Optic Neuropathy

Characteristic	Arteritic Features	Nonarteritic Features
Age	Mean, 70 years	Mean, 60 years
Sex	F > M	F = M
Associated symptoms	Headache, scalp tenderness, jaw claudication, transient visual loss	Usually none
Visual acuity	<20/200 in >60% of cases	>20/200 in >60% of cases
Disc/fundus	Pallid disc edema common Cup normal Cotton-wool spots	Hyperemic disc edema Cup small
Erythrocyte sedimentation rate	Mean, 70 mm/hr	Mean, 20–40 mm/hr
C-reactive protein level	Elevated	Normal
Platelet count	Elevated or normal	Normal
Fluorescein angiography findings	Disc delay and choroid delay	Disc delay
Natural history	Rarely improve Fellow eye involved, 54%–95%	31% improve Fellow eye involved, 12%–19%
Treatment	Systemic steroids	None proven

Arteritic anterior ischemic optic neuropathy

AAION is less frequent (5%–10% of AION cases) than NAION and usually occurs in older patients (mean age, 70 years).

Cause

- It is caused by inflammatory and thrombotic occlusion of the short posterior ciliary arteries.

Symptoms

- Systemic symptoms of GCA are usually present,
- ✓ Headache,
- ✓ Scalp tenderness,
- ✓ Malaise,
- ✓ Anorexia,
- ✓ Weight loss,

- ✓ Fever.
- ✓ Jaw claudication, the most specific symptom, describes masseter muscle pain or fatigue that occurs with prolonged chewing.
- The symptom worsens until chewing stops and resolves over minutes.
- Transient vision loss preceding AION may indicate GCA.
- Vision loss is typically severe (visual acuity is <20/200 in >60% of patients), and no light perception vision should prompt an aggressive evaluation for GCA.

Diagnosis

- Occult GCA, defined as either elevated erythrocyte sedimentation rate (ESR) without systemic symptoms or normal ESR in the presence of systemic symptoms, may occur in up to 20% of patients with AAION.
- Funduscopic clues to a diagnosis of AAION over NAION include the following:
  - ✓ Chalky white optic disc edema (in NAION the disc is often hyperemic)
  - ✓ Cotton-wool spots away from the optic disc indicative of concurrent retinal ischemia (cottonwool spots on or adjacent to the optic disc can be present in NAION)
  - ✓ Delayed choroidal filling on fluorescein angiographic studies (normally, the choroid fills completely within 3–5 seconds, before retinal arteries)
  - ✓ Normal or large cup in the fellow eye (in NAION, a small cup–disc ratio is common)
- Confirmational temporal artery biopsy may be delayed for 7–10 days without compromising test results

Management

- When AAION is suspected, immediate initiation of high-dose corticosteroid therapy is crucial.
- Adjunctive daily aspirin can also be added.
- Intravenous methylprednisolone (1 g/day for the first 3–5 days) is most often recommended, after which oral prednisone (1 mg/kg/d) may be used (up to 100 mg/day, tapered slowly over 3–12 months or more, depending on response).
- Alternate-day corticosteroid therapy is inadequate for AAION.
- The major goal of AAION therapy (apart from avoiding systemic vascular complications) is to prevent contralateral vision loss.
- Untreated, the fellow eye becomes involved in up to 95% of cases, within days to weeks.
- Although the initially affected eye may improve somewhat, recovery does not generally occur.
- The risk of recurrent or contralateral optic nerve involvement during corticosteroid withdrawal has been reported at 7%; thus, tapering must be done slowly and carefully.
- Recurrent symptoms or elevation of ESR should prompt reevaluation for disease activity.

Giant Cell Arteritis

- Giant cell arteritis (GCA), or temporal arteritis, is a systemic inflammatory granulomatous vasculitis that affects large and medium-sized arteries.
- This disease affects individuals aged 50 years or older, and the incidence increases with increasing age.
- Women are 2–4 times more likely to be affected than men.
- The annual incidence of GCA varies according to geographic location, for example, up to 20 cases per 100,000 people older than 50 years of age in Scandinavian countries but only about 1.5 cases per 100,000 people older than 50 years of age in Asian countries.
- Early diagnosis and treatment of GCA can limit or prevent permanent vision loss.

Clinical presentation of giant cell arteritis

- Systemic symptoms of GCA include headache and tenderness of the temporal artery or scalp.
- Jaw or tongue claudication (a cramping pain that develops while chewing or talking) is the symptom most specific for the disorder.
- Other symptoms include malaise, anorexia and weight loss, fever, new neck pain, joint and muscle pain, and ear pain. Systemic sequelae can include cerebrovascular ischemia, myocardial infarction, and aortic aneurysm or dissection.
- Visual symptoms may include transient or permanent visual loss, diplopia, and eye pain.
- Arteritic anterior ischemic optic neuropathy (AAION) is the most common cause of vision loss but central retinal artery occlusion, cilioretinal artery occlusion, posterior ischemic optic neuropathy, and ocular ischemic syndrome also occur.
- Ocular motor cranial nerve palsies can result in transient or constant diplopia.

Diagnosis of giant cell arteritis

- A high level of suspicion of GCA is paramount when evaluating patients over age 50 years with the visual symptoms just described.
- Diagnostic evaluation begins with laboratory tests of the erythrocyte sedimentation rate (ESR), complete blood count (CBC), and C-reactive protein (CRP) level.
- Most cases of GCA show marked elevation of the Westergren ESR (mean 70 mm/hr; often >100 mm/hr), but the level may be normal in up to 16% of cases.
- The ESR rises with anemia and with increasing age; levels above the laboratory's listed upper limit of normal are common in patients over 70 years old without arteritis.
- A more accurate estimate of the upper normal value is obtained by using these formulas: [age]/2 (men) or [age + 10]/2 (women).
- Measurement of CRP level, which may be more specific and less affected by increasing age and anemia, may increase diagnostic accuracy and is currently recommended in conjunction with the ESR.
- Thrombocytosis (increased platelet count) may suggest active disease.
- A normochromic normocytic anemia may be present.
- Temporal artery biopsy is required whenever clinical suspicion or laboratory studies suggest the possibility of GCA; however, a negative result on biopsy does not rule out GCA.
- False-negative results have been estimated at 3%– 9%, relating in part to the possibility of discontinuous arterial involvement ("skip areas") and missed lesions; biopsy segments should be 2–3 cm long to increase the potential yield.
- If an initial biopsy result is negative, a contralateral biopsy should be considered if the clinical picture is highly suspicious.
- Other imaging strategies (color Doppler ultrasonography, positron emission tomography, and magnetic resonance imaging [MRI]) are being investigated for use in the diagnosis of GCA, but no firm conclusions can yet be made regarding their accuracy.

Treatment of giant cell arteritis

- When GCA is suspected, steroid therapy must begin immediately.
- Confirmational temporal artery biopsy, ideally at a site of localized tenderness and/or on the side of vision loss, should be done within 1–2 weeks but in some cases remains positive for months after initiation of therapy.

- Intravenous methylprednisolone (1 g/day for the first 3–5 days) is often recommended when vision loss is present.
- For patients with suspected GCA but without loss of vision, oral prednisone 60–100 mg/day may be sufficient.
- Treatment is also required for patients with severe, bilateral vision loss, as this is a systemic disease.
- The clinical response to steroids is often dramatic, with reduction of symptoms within days.
- The corticosteroids are tapered slowly, depending on response, and patients typically continue therapy for at least 1–2 years, sometimes longer.
- In addition, long-term steroid therapy requires monitoring and treatment for osteopenia and osteoporosis as well as prophylaxis of gastrointestinal effects, and it is recommended that these patients be co-managed with a rheumatologist or internist.
- Corticosteroid treatment does not generally improve vision, but it may prevent new ischemic events from occurring.
- The risk of recurrent or contralateral optic nerve involvement during withdrawal from corticosteroid treatment has been reported at 7%; thus, tapering must be done slowly and carefully.
- Patients must also be monitored for thoracic and aortic aneurysms, which are associated with GCA.
- Additionally, these patients are at risk of increased cerebrovascular and cardiovascular ischemia; thus, low-dose aspirin administration should be considered.

Non-arteritic anterior ischemic optic neuropathy

- The non-arteritic form of AION is more common (accounting for 90%–95% of AION cases) and occurs in a relatively younger age group (mean age, 60 years) than the arteritic form.
- The annual incidence is approximately 80/100,000.

Cause

- NAION presumably relates to compromised optic disc microcirculation in eyes with structural "crowding" of the disc.

Histology

- Histologic studies show the area of infarction is located within the scleral canal alone, supporting the compartment syndrome theory.

Symptoms & signs

- Patients frequently report visual impairment upon awakening.
- The initial course may remain static, in which vision loss is stable from onset, or become progressive, which involves either episodic, stepwise decrements or a steady decline of vision over weeks to months before eventual stabilization.
- The progressive form has been reported in 22%–37% of NAION cases.
- No systemic symptoms are typically associated with NAION.
- Vision loss is usually less severe than in AAION (visual acuity >20/200 in >60% of cases).
- The most common pattern of visual field loss is altitudinal defects, but any pattern may be seen.
- The optic disc edema in NAION may be diffuse or segmental and hyperemic or pale.
- The optic disc in the contralateral eye is typically small in diameter and demonstrates a small or absent physiologic cup ("disc at risk").

- The optic disc usually becomes visibly atrophic within 6–8 weeks; persistence of edema past this point could suggest an alternative diagnosis.
- The 5-year risk of contralateral involvement is 15%.
- Occurrence in the second eye produces the clinical appearance of "pseudo–Foster Kennedy syndrome," in which the previously affected disc is atrophic and the currently involved nerve head is edematous.
- Both eyes show visual field loss characteristic of AION.
- This is in contrast to the true Foster Kennedy syndrome, secondary to intracranial mass, in which 1 optic disc is atrophic because of chronic compression by the mass, whereas the other disc is edematous because of elevated ICP.
- NAION is associated with the following factors:
  - ✓ Structural crowding of the disc (disc at risk),
  - ✓ Systemic hypertension,
  - ✓ Diabetes mellitus (particularly in young patients),
  - ✓ Hyperlipidemia.
- Neither carotid occlusive disease nor prothrombotic disorders is a proven risk factor.
- Hypercoagulable disorders, sleep apnea, and nocturnal hypotension may be associated but remain currently unproven.
- One suggested association is with phosphodiesterase inhibitors (eg, sildenafil), presumably because of their hypotensive effect, although causation has not been proven.
- NAION must be differentiated from optic neuritis.

Table 4-5 NAION Versus Optic Neuritis: Typical Features

	NAION	Optic Neuritis
Age	>50 years	<40 years
Associated pain	Unusual	With eye movement, 92%
Pupil	+ RAPD	+ RAPD
Visual field defect	Altitudinal	Central
Optic disc findings	Edema 100%; may be pale	Edema 33%; hyperemic
Retinal hemorrhage	Common	Unusual
Fluorescein angiographic findings	Delayed disc filling	No delayed disc filling
MRI findings	No optic nerve enhancement	Optic nerve enhancement

MRI = magnetic resonance imaging; NAION = nonarteritic anterior ischemic optic neuropathy; RAPD = relative afferent pupillary defect.

Diagnosis

- In unclear cases, contrast enhanced MRI can help in the differentiation.
- The affected optic nerve appears normal in NAION (95%) but enhances with use of gadolinium contrast in optic neuritis (90%).
- Untreated NAION generally remains stable after reaching the low point of visual function, but improvement of at least 3 lines of Snellen visual acuity was reported in 31% of patients after 2 years in the Ischemic Optic Neuropathy Decompression Trial (IONDT).
- Recurrent episodes of vision loss in the same eye after 3 months are unusual in NAION (up to 6.4%), occurring most often in young patients.

Treatment

- There is no proven therapy for NAION.
- The IONDT showed no benefit of ONSF for NAION, and this surgical option has therefore been abandoned as a treatment modality.

- Neuroprotective drugs have demonstrated beneficial effects against secondary neuronal degeneration in animal models of ischemic retinal ganglion cell damage and optic nerve crush injury; however, clinical studies have been unsuccessful in recruiting sufficient numbers of patients in a timely manner.
- One study examined treatment of patients with oral prednisone, 80 mg daily for 2 weeks, followed by a slow taper over several weeks.
- This retrospective evaluation of almost 700 patients showed improvement in visual acuity and visual fields only among patients with visual acuity of 20/70 or worse.
- This treatment remains controversial given the potential biases in the study methodology.
- There is also no proven prophylaxis for the fellow eye.
- Although aspirin has a proven effect in reducing the risk of secondary stroke, its role in reducing the incidence of fellow eye involvement after the initial episode remains unproven.

Posterior ischemic optic neuropathy

- Acute ischemic damage to the retrobulbar portion of the optic nerve is characterized by abrupt, often severe, vision loss, an RAPD, and initially normal-appearing optic discs.
- Posterior ischemic optic neuropathy (PION) is considered rare and is a diagnosis of exclusion.
- It occurs in 3 distinct scenarios:
  1. Perioperative (most commonly in spine, cardiac, and head or neck procedures);
  2. Arteritic, especially from giant cell arteritis; and
  3. Nonarteritic (with risk factors and clinical course similar to those of NAION).
- Vision loss in perioperative PION is more commonly bilateral and profound.
- Perioperative PION has gained more widespread scrutiny because of the increasing number of reports and the potential medicolegal implications.
- Nearly all patients with perioperative PION have incurred substantial blood loss, hypotension, and prolonged anesthesia time.
- However, several case control studies do not show that any one factor clearly occurs more commonly in patients with PION than in patients who undergo similar surgeries but do not sustain perioperative vision loss.
- Additionally, reports have been published of patients with PION who lost less than 500 mL of blood and had systolic blood pressure >90 mm Hg.
- More than likely, the cause of vision loss is multifactorial and results from a combination of patient and perioperative factors.
- Patients with no history of previous surgery should undergo neuroimaging.
- Also, a careful evaluation for symptoms and laboratory evidence of GCA should be undertaken in older individuals.
- High-dose corticosteroid treatment is indicated for cases of proven GCA.

Prognosis for vision recovery in PION is poor.