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Research Article

ISCHEMIC OPTIC NEUROPATHIES

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A R T I C L E I N F O

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A B S T R A C T

Ischemic optic neuropathy represents as an acute ischaemic disorder of the optic nerve. It is of two very distinct types: anterior ischemic optic neuropathy (AION) involving the ONH, and posterior ischaemic optic neuropathy (PION) involving a segment of the rest of the optic nerve posteriorly. Arteritic anterior ischaemic optic neuropathy (AAION) results from short posterior ciliary artery (SPCA) vasculitis and the resultant optic nerve head infarction. The rapid onset, stable course with generally poor recovery, association with vasculopathic risk factors, and similarity to AAION implies a vascular cause for nonarteritic anterior ischaemic optic neuropathy (NAION) as well, but the direct evidence remains limited. AAION is caused by giant cell arteritis (GCA). NAION has been reported in association with a number of diseases that could predispose to reduced perfusion pressure or increased resistance to flow within the optic nerve head. Typically, AAION develops in elderly patients, with a mean age of 70 years, with severe visual loss. Pallor of the optic disc, which may be severe, chalky-white, is associated with AAION. The nonarteritic form of the disease occurs in a somewhat younger age group (mean age of 60 years) and usually is associated with less severe visual loss. Ischemia of the optic nerve that does not involve the optic nerve head is termed posterior ischemic optic neuropathy (PION). It presents with acute visual loss associated with signs of optic neuropathy (afferent pupillary defect and visual field loss) in one or both eyes, with initially normal appearance of the optic disc, which subsequently becomes atrophic.

INTRODUCTION

Optic nerve ischemia most frequently occurs at the optic nerve head, where, in susceptible individuals, structural crowding of nerve fibers and the relative reduction of the vascular supply combine to impair perfusion to a critical degree and produce optic disc edema[1]. The optic nerve head (ONH) is almost entirely supplied by the posterior ciliary artery (PCA) circulation and the rest of optic nerve posterior to the ONH is supplied from several other sources. In view of that, pathogenetically, as well as clinically, ischaemic optic neuropathy is of two very distinct types: anterior ischemic optic neuropathy (AION) involving the ONH, and posterior ischaemic optic neuropathy (PION) involving a segment of the rest of the optic nerve posteriorly[2,3]. Generally, AION is categorized as either arteritic (associated with temporal arteritis) or nonarteritic.

Anterior Ischemic Optic Neuropathy (AION)

Epidemiology and Pathogenesis

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Nonarteritic anterior ischemic optic neuropathy (NAION) is the most common acute optic neuropathy in patients over 50 years of age. No gender predisposition exists, but the disease occurs with significantly higher frequency in white than in black or Hispanic populations.[4,5] The incidence of arteritic anterior ischemic optic neuropathy (AAION) is lower.[4]

Arteritic anterior ischemic optic neuropathy (AAION)

AAION results from short posterior ciliary artery (SPCA) vasculitis and the resultant optic nerve head infarction. Human autopsy studies of acute AAION demonstrate optic disc edema with ischemic necrosis of the prelaminar, laminar, and retrolaminar portions of the nerve and infiltration of the SPCAs by chronic inflammatory cells. Segments of these vessels in some cases were occluded by inflammatory thickening and thrombus.[6]

Nonarteritic anterior ischemic optic neuropathy (NAION)

The rapid onset, stable course with generally poor recovery, association with vasculopathic risk factors, and similarity to AAION implies a vascular cause for NAION as well, but the direct evidence remains limited.[7] The most commonly proposed pathogenic theory states that insufficiency of the optic disc circulation, exacerbated by structural crowding of nerve fibers and supporting structures at the nerve head,

eventually reaches a point at which inadequate oxygenation produces ischemia and swelling of the disc.[1] These features may be mild and subclinical (no visual loss), reversible to some degree, or irreversible (infarction). Periodic nocturnal systemic hypotension and that the optic disc is in a watershed zone between distributions of lateral and medial SPCAs may be contributing factors.[8]

Systemic Associations

Arteritic anterior ischaemic optic neuropathy (AAION) is caused by giant cell arteritis (GCA). GCA is systemic granulomatous arteritis affecting large and medium sized arteries (internal elastic lamina) which mainly affects temporal artery, posterior ciliary artery, ophthalmic artery and vertebral artery. About 50% of patients with GCA have polymyalgia rheumatica (PMR) at diagnosis, while around 20% of PMR patients will develop GCA. PMR is characterized by pain and stiffness in proximal muscle groups, typically the shoulders and biceps, that is worse on waking. The causative relationship between GCA and PMR remains uncertain, though many suspect them to be different presentations of the same underlying entity.[9]

NAION has been reported in association with a number of diseases that could predispose to reduced perfusion pressure or increased resistance to flow within the optic nerve head. Systemic hypertension has been documented in up to 47% of patients who have NAION and diabetes in up to 24%. Diabetics in particular show a predisposition to NAION at a young age.[10] Also, NAION has been reported in association with multiple forms of vasculitis, acute systemic hypotension, migraine, optic disc drusen, and idiopathic vaso-occlusive diseases. Other risk factors, such as hyperopia, smoking, the presence of human lymphocyte antigen A29, and hyperlipidemia have been proposed.[1]

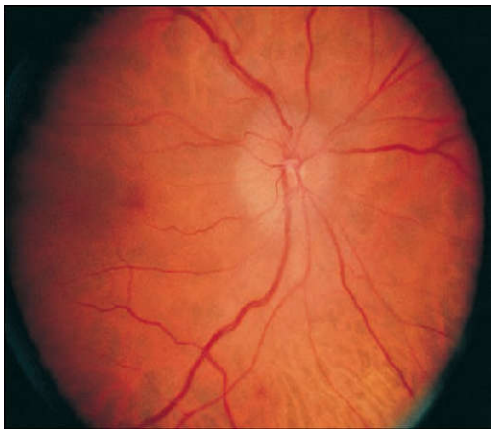
Ocular Manifestation

AION presents with rapid onset of painless, unilateral visual loss manifested by decreased visual acuity, visual field, or both. The level of visual acuity impairment varies widely, from minimal loss to no light perception. An altitudinal field defect is most common, but generalized depression, broad arcuate scotomas, and cecocentral defects also are seen. A relative afferent pupillary defect is present with monocular optic neuropathy. The optic disc is edematous at onset, and edema occasionally precedes visual loss by weeks to months.[13]

Arteritic anterior ischemic optic neuropathy

Typically, AAION develops in elderly patients, with a mean age of 70 years, with severe visual loss (visual acuity < 20/200 (6/60) in the majority). It may be preceded by transient visual loss similar to that of carotid artery disease; this finding is extremely unusual in the nonarteritic form and, when present, is highly suggestive of arteritis.[14] Patients with the arteritic form note other symptoms of the disease - headache (most common), jaw claudication, and temporal artery or scalp tenderness are those aligned most frequently with a final diagnosis of temporal arteritis. Malaise, anorexia, weight loss, fever, proximal joint arthralgia, and myalgia also are noted commonly; however, the disease rarely manifests with visual loss in the absence of overt systemic symptoms, so-called occult giant cell arteritis. In GCA patient, amaurosis fugax is a very important symptom. Pallor of the optic disc, which may be severe, chalky-white, is associated with AAION. The disc of the fellow eye is of normal diameter in AAION.[15]

Although papilledema has been described as the hallmark of AAION, it is common to see hyperemic swelling, particularly in the nonarteritic form. The disc most often is swollen diffusely, but a segment of more prominent involvement may be present, and either focal or diffuse surface telangiectasia is not unusual and may be quite pronounced.[1]

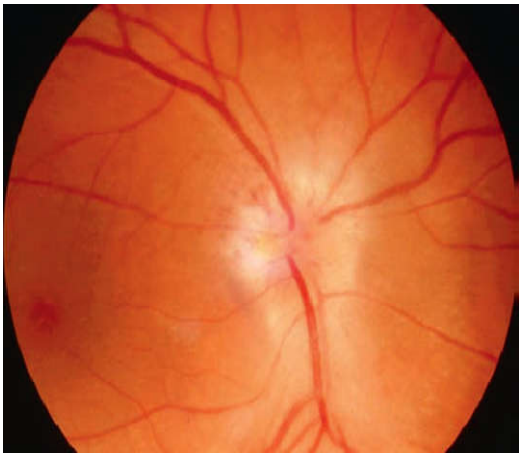


Fundus view, anterior ischemic optic neuropathy-The optic disc demonstrates pale, diffuse edema.[1]

Nonarteritic anterior ischemic optic neuropathy

The nonarteritic form of the disease occurs in a somewhat younger age group (mean age of 60 years) and usually is associated with less severe visual loss. Frequently, visual impairment is reported upon awakening, possibly related to nocturnal systemic hypotension.[8] The progressive form has been reported in up to 37%[16] of NAION cases. Usually, no associated systemic symptoms occur, although periorbital pain is occasionally described. Fellow eye involvement is estimated to occur in 12-19% by 5 years after onset.[17]

The optic disc edema in NAION may be diffuse or segmental, hyperemic or pale, but pallor occurs less frequently. A focal region of more severe swelling is often seen and typically display an altitudinal distribution, but it does not correlate consistently with the sector of visual field loss.[18] The optic disc in the contralateral eye typically is small in diameter and demonstrates a small or absent physiological cup. The disc appearance in such fellow eyes has been described as the disc at risk, with postulated structural crowding of the axons at the level of the cribriform plate, associated mild disc elevation, and disc margin blurring without overt edema.

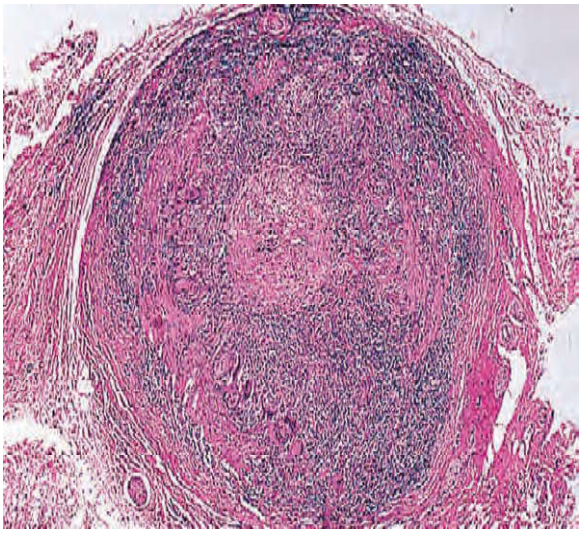


Fundus view, nonarteritic anterior ischemic optic neuropathy. The hyperemic disc edema is more prominent superiorly. Focal surface telangiectasia of disc vessels is seen superotemporally (arrows).[1]

Diagnosisnd Ancilliary Testing

The most important early step in the management of AION is the differentiation of the arteritic from the nonarteritic form of the disease. Measurement of the erythrocyte sedimentation rate (ESR) remains the standard of care. Active temporal arteritis usually is associated with an elevation of ESR to 70-120 mm/hour, and this finding suggests the arteritic form; in most cases, it should prompt immediate corticosteroid therapy and confirmatory temporal artery biopsy. Hayreh *et al*.[19] reported 97% specificity for temporal arteritis in cases of AION in which both ESR > 47 mm/ hour and CRP > 2.45 mg/dL were found.

Positive biopsy findings, such as intimal thickening, internal limiting lamina fragmentation, and chronic inflammatory infiltrate with giant cells, provide support for long-term systemic corticosteroid therapy. A negative biopsy result, however, does not rule out arteritis; both discontinuous arterial involvement ('skip lesions') and solely contralateral temporal artery inflammation may result in false-negative results.

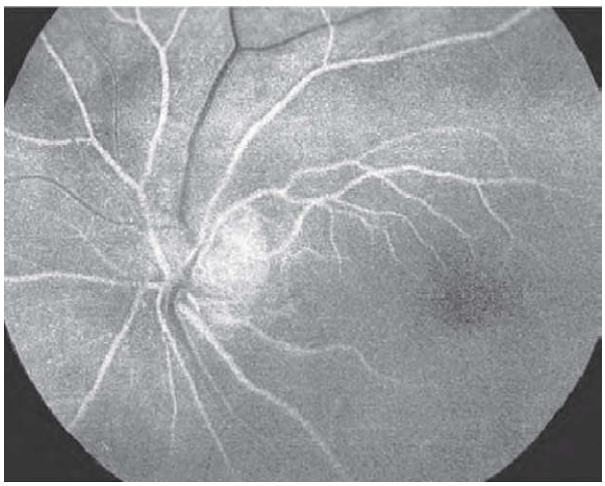


Typical temporal arteritis. Histological section shows a vasculitis involving all coats of the temporal artery.(Courtesy of Dr. M. M. Rodrigues.)[1]

Perimetry shows a relative or an absolute field defect which may be sectorial or altitudinal or there may be a central scotoma. The most common field defect in NA-AION is inferonasal sectoral field defect.

Optic Coherence Tomography (OCT) is a useful tool in the assessment of AION. Sectoral disc edema, retinal nerve fiber layer (RNFL) thickness, and then resolution to normal or atrophy can be well documented.[20] The extent and pattern of RNFL loss can be correlated to visual field loss.[21]

Fluorescein angiographic data shows delayed filling of the optic disc and choroid. Extremely poor or absent filling of the choroid has been depicted as a characteristic of AAION and has been suggested as one useful factor by which to differentiate AAION from NAION. Delayed completion of choroidal fluorescein filling that averages 30-69 seconds has been reported in AAION, compared with a mean of 5-13 seconds in NAION.[22]



Fluorescein angiogram, early arteriovenous phase, in nonarteritic anterior ischemic optic neuropathy. The temporal portion of the optic disc fills normally, but the remaining sectors demonstrate markedly delayed filling approximately 10 seconds later.[1]

Differential Diagnosis

The differential diagnosis of AION includes idiopathic optic neuritis, particularly in patients under 50 years of age; other forms of optic nerve inflammation, such as those related to syphilis or sarcoidosis; infiltrative optic neuropathies; anterior orbital lesions that produce optic nerve compression; and diabetic papillopathy.

Treatment

Arteritic anterior ischemic optic neuropathy

Treatment is aimed at preventing blindness of the fellow eye, as visual loss in the index eye is unlikely to improve even with immediate treatment; the second eye may still become involved in 25% despite early steroid administration.[9] The regimen is as follows:

Intravenous methylprednisolone, 500 mg to 1 g/day for 3 days followed by oral prednisolone 1-2 mg/kg/day. After 3 more days the oral dose is reduced to 50-60 mg (not less than 0.75 mg/kg) for 4 weeks or until symptom resolution and ESR/CRP normalization. A typical subsequent regimen consists of reducing the daily dose by 10 mg/day every 2 weeks until 20 mg/day is reached, with tapering afterwards titrated against ESR/CRP and symptoms, e.g. a 2.5 mg reduction every 2-4 weeks to 10 mg then a 1 mg reduction every 1-2 months.[9] A positive response is so typical that if it does not occur, an alternate disease process should be considered.[1]

Nonarteritic anterior ischemic optic neuropathy

There is no proven effective therapy for NAION. Oral corticosteroids at standard dosage (1 mg/kg per day) are not beneficial, and megadose intravenous therapy has not been evaluated systematically.

The Ischemic Optic Neuropathy Decompression Trial compared ONSD surgery in 119 patients with no treatment in 125 controls.[4] The study revealed no significant benefit for treatment and a possible, harmful effect; it was recommended that ONSD not be performed for NAION.

A controlled clinical pilot study of hyperbaric oxygen in 22 patients who had acute NAION, however, has shown no beneficial effect.[23] The effect of aspirin in reducing risk of fellow eye involvement is also unproven.[24]

Course and Outcome

Arteritic anterior ischemic optic neuropathy

Prognosis for visual recovery in the affected eye that has treatment generally is poor, but recent reports suggest a 15-34% improvement rate,[25] which is higher with intravenous than with oral therapy. Worsening of vision in spite of therapy has been reported in 9-17% of cases.

Nonarteritic anterior ischemic optic neuropathy

Reports indicate that 24-43% of cases demonstrate spontaneous improvement of visual acuity by three Snellen lines or more.[5] Even in the progressive form, improvement has been reported to occur in roughly 30% of cases. Whether NAION is static or progressive, visual acuity and field stabilize after several months.

Posterior Ischemic Optic Neuropathy

Ischemia of the optic nerve that does not involve the optic nerve head is termed posterior ischemic optic neuropathy (PION). It presents with acute visual loss associated with signs of optic neuropathy (afferent pupillary defect and altitudinal visual field loss) in one or both eyes, with initially normal appearance of the optic disc, which subsequently becomes atrophic.

The diagnosis of PION is most often made in one of the following settings:

Vasculitis, most importantly giant cell arteritis (GCA), systemic lupus erythematosus, collagen vascular disease, shock optic neuropathy, combination of *systemic hypotension and anemia*, usually related to blood loss either from surgery (coronary artery bypass and lumbar spine procedures most commonly), gastrointestinal bleed, or trauma.[1]

The diagnosis of PION should be made only after other causes of retrobulbar optic neuropathy, such as compression or inflammation, have been excluded. Initially, the optic disc appears normal but pallor develops over weeks.[9] Although the PION is typically more abrupt in onset, neuroimaging is indicated to rule out.

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