Non-Arteritic Anterior Ischemic Optic Neuropathy with a Rapidly Progressive Bilateralization: Case Report

Nooh Abutabeekh; Youssef Harrar
Malik Alkteish; Walid Berramdan
Asmae Maadane; Fatima Zahra Mabrouki
Rachid Sekhsoukh
Ophthalmology Department, University Hospital Center Mohammed VIth Oujda, Morocco

Abstract
We present a case of nonarteritic anterior ischemic optic neuropathy (NA-AION) with rapidly progressive bilateralization and unclear etiology.
We report the case of a 58-year-old man with no specific past medical history who was consulted for a sudden, non-painful, unilateral visual defect in his left eye after waking from sleep. His visual acuity was 10/10 in both eyes. Anterior segment examination was unremarkable in both eyes, but a positive Marcus Gunn signal in the left eye.
Fundoscopic examination, OCT examination, fluorescein and indocyanine green angiography (ICG) confirmed papilledema in the left eye. The octopus visual field (OVF) showed an inferior altitudinal scotoma. Thus, after ruling out other diagnoses, a diagnosis of NAION was made in the left eye, the patient was consulted for the same symptoms in the right eye Vision was limited to counting fingers at 4 meters in the right eye and 10/10 in the left eye Anterior segment examination was unremarkable in both eyes, but OCT examination of the fundus (RAPD) in the right eye Fluorescein and indocyanine green (ICG) angiography confirmed papilledema in the right eye OVF showed tunnel vision in both eyes blood tests and imagery were negative The diagnosis of non-arteritic AION in the right eye was retained.
A therapeutic trial was performed; the patient received a corticosteroid bolus of 1g /day for 3 days, followed by oral treatment
1 month later, visual acuity was 0.4/10 in the right eye and 10/10 in the left eye. Fundus examination and OCT scan showed regression of the edema in the right eye and papillary atrophy in the left eye with tunnel vision in both eyes with OVF.

Keywords: Papillary Edema, Optic Disc Edema, Non-Arteritic Ischemic Optic Neuropathy (NA-AION), bilaterlization ,Visual Field, Optic Nerve Atrophy, Optic Nerve Inflammation.

Introduction

Ischemic optic neuropathy represents an acute ischemic insult to the optic nerve and is classified into two major types: anterior and posterior. Non-arteritic anterior ischemic optic neuropathy (NAION) is the most prevalent acute optic neuropathy in individuals over the age of 50, with an estimated annual incidence in the United States ranging from 2.3 to 10.2 cases per 100,000 population [1,2].
Ischemic optic neuropathies are anatomically categorized into:Anterior Ischemic Optic Neuropathy (AION): Resulting from ischemia of the anterior portion of the optic nerve, primarily supplied by the short posterior ciliaryarteries.Posterior Ischemic Optic Neuropathy (PION): Involving ischemia of the posterior segment of the optic nerve,

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which receives a more complex vascular supply.

AION is further subdivided based on etiology into:Arteritic AION (A-AION): Primarily caused by giant cell arteritis (GCA), also known as Horton disease. Non-Arteritic AION (NA-AION): Attributed to a variety of non-inflammatory causes [3,4].

NAION occurs due to hypoperfusion of the optic disc's anterior segment, supplied by the short posterior ciliary arteries. Multiple systemic and local risk factors contribute to this compromised perfusion, including advanced age, systemic hypertension, diabetes mellitus, hyperlipidemia, nocturnal hypotension, smoking, and a structurally small optic disc with a crowded cup [5,6].

Due to the multifactorial etiology, there is currently no definitive therapy proven to halt disease progression in the affected or contralateral eye.

NAION typically presents as painless, acute vision loss developing over several hours to days. Patients often describe their symptoms as blurring, dimming, or clouding of vision, frequently affecting the inferior visual field. Although the condition is usually painless, periocular discomfort has been reported in 8–12% of cases [5,6].

Visual acuity in NAION varies widely, ranging from 20/20 (logMAR 0) to no light perception. However, vision loss in NAION is generally less severe than in A-AION, with more than 50% of patients retaining visual acuity better than 20/200 (logMAR 1) [7].

Color vision loss in NAION tends to correlate proportionally with the degree of visual acuity impairment, unlike in optic neuritis, where color vision may be disproportionately affected. Visual field defects in NAION may follow various patterns, though inferior altitudinal defects are most common, reported in 55–80% of cases [7,8].

Case Report

We report the case of a 58-year-old French man with no particular history who was consulted for a sudden, non-painful, unilateral visual defect on the left side after waking from sleep.

At presentation, the visual acuity (VA) of the left and right eyes was 10/10. The anterior segment examination was unremarkable in both eyes, but the left eye showed a positive Marcus Gunn signal. OCT examination showed a swollen left optic disk with (Figure 1). Fluorescein and indocyanine green (ICG) angiography confirmed papilledema in the left eye and the absence of signs of delayed choroidal perfusion or choroidal ischemia (Figure 2). Octopus visual field (OVF) showed an inferior altitudinal scotoma (Figure 3) Computed tomography of the brain and orbit ruled out compressive lesions. Therefore, the diagnosis of NAION in the left eye was made. Three months later, the patient consults for rapidly progressive decline in visual acuity in the right eye since a week. VA was limited to counting fingers at 4 meters in the right eye and 10/10 in the left eye Examination showed right eye relative afferent pupillary defect (RAPD). His left optic disc was pale; however, the right was hyperemic and swollen (Figure 4). Papillary Optical coherence tomography (OCT) of the optic nerve showed papilledema in the right eye, RNFL at 254 µm and a nasal RNFL deficit in the left eye. (Figure 5). Fluorescein angiography confirmed papilledema in the right eye and the absence of ocular signs of Horton's disease. (Figure 6) .OVF showed tunnel vision in both eyes (Figure 7), Emergency laboratory tests were requested. CBC vs. CRP returned normal. An orbitocerebral and supra-aortic artery CT angiography was requested, with unremarkable results. The patient was hospitalized in internal medicine.The patient was further investigated in order to establish the etiological diagnosis and the course of treatment.The diagnosis of non-arteritic AION in the right eye was

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retained. A therapeutic trial was carried out; the patient was placed on a corticosteroid bolus of 1g/day for 3 days, followed by oral treatment, with subsequent gradual reduction of the dosage, all while knowing that the clinical picture was suggestive of non-arteritic anterior ischemic optic neuropathy (NAION).

Unfortunately, the patient retained a limited visual acuity of 0.4/10 in the right eye and 10/10 in the left eye, with bilateral concentric visual field constriction (tubular vision) as shown in Figure 7, one month post-onset. There was regression of the papilledema with inferior defect in the right eye and nasal optic disc atrophy in the left eye, as illustrated in (Figure 8).

Discussion

NA-AION primarily affects older adults and is typically characterized by a sudden, painless reduction in vision, most commonly noticed upon awakening [9]. At initial presentation, the condition usually affects only one eye; however, involvement of the fellow eye is not uncommon, occurring in approximately 15% of patients within five years [10,11]. This bilateral manifestation is thought to be due to similar optic disc anatomy in both eyes and exposure to shared systemic vascular risk factors [12,13].

The pathogenesis of NA-AION is multifactorial. In approximately 72% of cases, patients present with predisposing systemic or local risk factors [14]. A nearly universal feature among those affected is the presence of a "disc-at-risk"a small, crowded optic disc that predisposes the eye to ischemic damage [15]. Histopathological evidence from two studies supports the hypothesis that the anatomical configuration of the optic disc may contribute to a compartment syndrome, which in turn compromises axonal function and perfusion [16,17].

Disruption of the autoregulatory mechanisms that maintain optic disc perfusion is believed to result in ischemia. However, current clinical methods do not allow for reliable measurement of blood flow in the posterior ciliary arteries that supply the optic nerve head. Among the numerous contributing factors, nocturnal hypotension is one of the most strongly associated, with up to 73% of cases reporting visual loss upon awakening [9]. Furthermore, phosphodiesterase type 5 (PDE5) inhibitors, commonly taken at night, have been implicated in exacerbating nocturnal hypotension and potentially contributing to NA-AION pathogenesis [18].

Patients with diabetes mellitus and those who experience significant vision loss during the initial episode of non-arteritic anterior ischemic optic neuropathy (NA-AION) are at an elevated risk for sequential bilateral involvement [11]. Clinically, NA-AION is often characterized by the presence of a relative afferent pupillary defect (RAPD) and optic disc edema, which may be either diffuse or sectoral in distribution.

The most commonly observed visual field defect in NA-AION is an altitudinal loss, particularly affecting the inferior field [19]. However, in many cases, the visual field may be profoundly depressed or entirely non-recordable due to the extent of optic nerve dysfunction [20].

Fluorescein fundus angiography (FFA) serves as a valuable diagnostic tool by demonstrating delayed filling of the optic disc, indicative of hypoperfusion and subsequent ischemia. The pathophysiology involves compromised perfusion of the optic nerve head, further exacerbated by structural crowding of axons and surrounding tissue, leading to reduced oxygenation, ischemia, and disc edema. A self-perpetuating cycle may ensueischemia induces axonal swelling, which causes microvascular compression, further worsening ischemia and potentially resulting in progressive optic nerve damage. Such progression has been

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reported in approximately 37% of NA-AION cases [21].

Although there is no universally accepted treatment for NA-AION, evidence from Hayreh et al. suggests that systemic corticosteroids administered during the acute phase,while optic disc edema is still presentmay lead to improvements in both visual acuity and visual field outcomes compared to untreated patients [22]. Additionally, topical brimonidine has been explored for its potential neuroprotective properties; however, clinical trials have not demonstrated statistically significant improvements in visual acuity compared to placebo [23].

The proposed mechanism by which corticosteroids may enhance outcomes in patients with non-arteritic anterior ischemic optic neuropathy (NA-AION) involves interruption of a self-perpetuating cycle of secondary damage [22]. In this model, ischemic axons swell within a structurally crowded optic disc—characterized by a small scleral canal—exerting mechanical pressure on surrounding tissue. While corticosteroids do not prevent the initial ischemic insult, they may theoretically mitigate secondary injury by reducing inflammation and mechanical compression. Another potential mechanism involves corticosteroid-mediated reduction of capillary permeability in the optic disc, thereby limiting fluid leakage and edema formation [23].

It is hypothesized that preserving the optic nerve from secondary inflammatory and mechanical damage may be more effectively achieved through high-dose intravenous corticosteroids rather than oral administration. This approach is supported by findings from the Optic Neuritis Treatment Trial (ONTT), which demonstrated superior efficacy of intravenous corticosteroids in the treatment of optic neuritis [25].

Currently, there is no established prophylactic therapy to prevent NA-AION recurrence in the same or fellow eye. Although some studies suggest that aspirin may reduce the incidence of fellow-eye involvement following an initial episode [26,27], others have reported no long-term benefit [28]. Despite the lack of conclusive evidence for its efficacy in NA-AION prevention, aspirin is frequently recommended by clinicians—primarily for its cardiovascular protective effects, including reduction in the risk of stroke and myocardial infarction, in this vasculopathy-prone population.

Conclusion

Progressive NA-AION affecting the same or the contralateral eye is relatively uncommon but warrants thorough investigation when it occurs. The pathophysiology of NA-AION remains incompletely understood, and no definitive treatment has been identified. Therefore, proactive management of modifiable systemic and local risk factors is essential in reducing the likelihood of further ischemic events.

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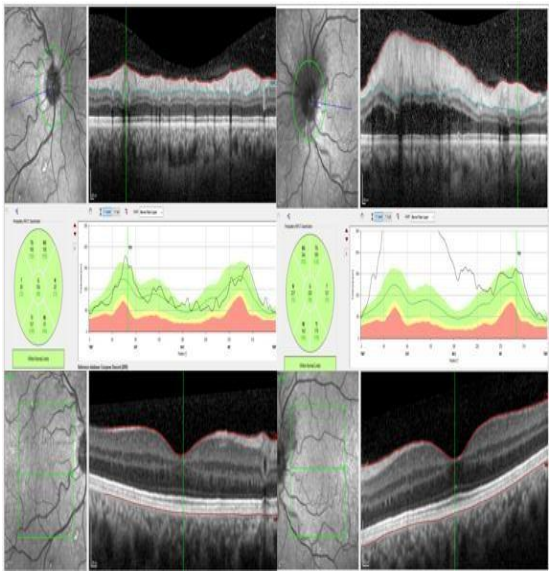

FIGURE 1: OCT scans heidelberg revealed swollen left optic disc with normal right optic disc and normal macular OCT

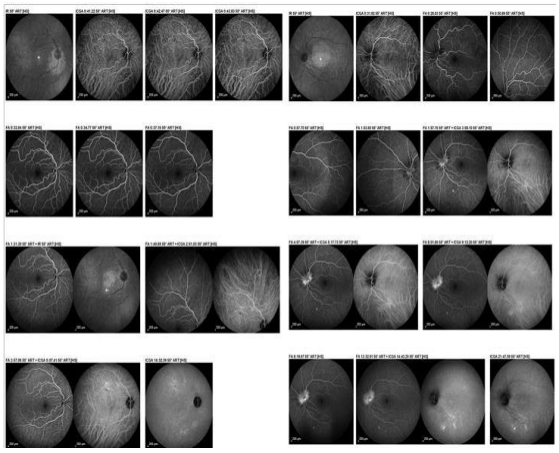

FIGURE 2: Fluoresce in and indocyanine green (ICG) angiography confirmed papilledema in the left eye and the absence of signs of delayed choroidalperfusion or choroidalischemia.

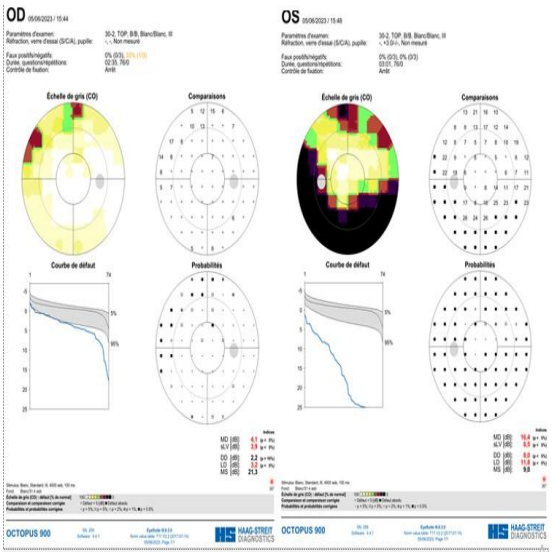

FIGURE 3: Octopus visual field (OVF) showed inferior altitudinal scotoma in the left eye.

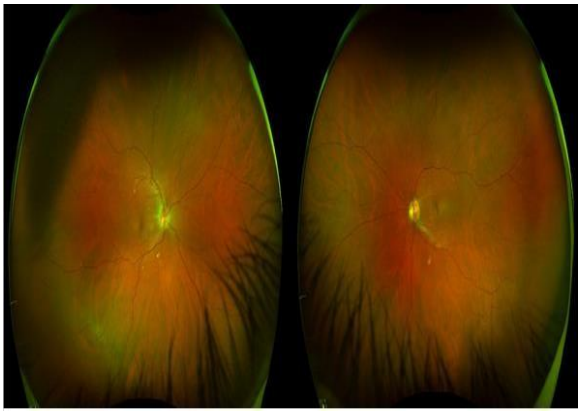

FIGURE 4 : Color fundus photography showsRight eye's swollen optic disc while the left eye's optic disc is pale .

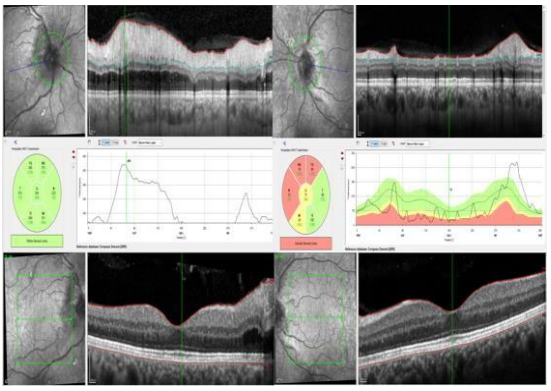

FIGURE 5 :Papillary Optical coherence tomography (OCT) of the optic nerve shows papilledema in the right eye RNFL defect in left eye and normal macular OCT in both eyes

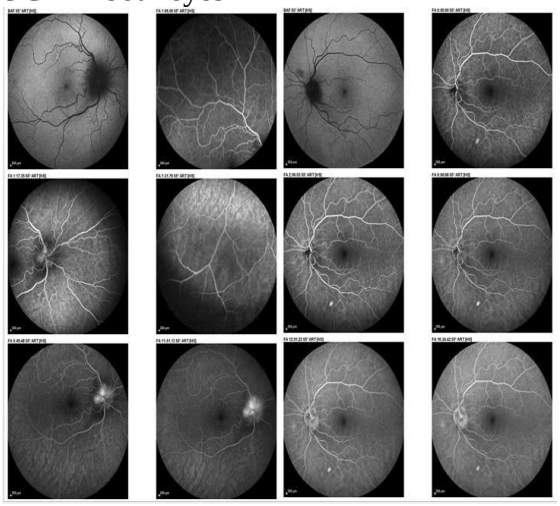

FIGURE 6: Fluorescein angiography confirmed papilledema in the right eye and the absence of ocular signs of Horton's disease.

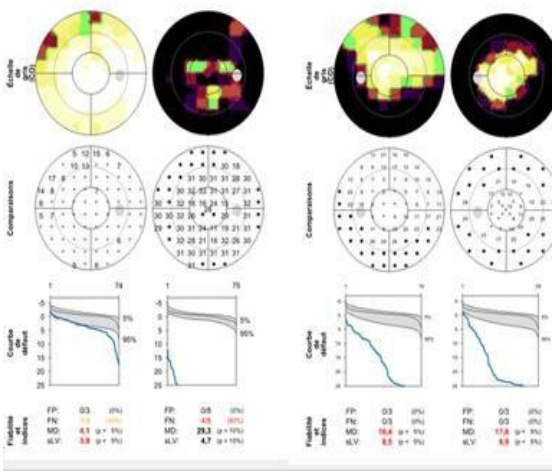

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FIGURE 7:OVF shows tunnel vision in both eyes

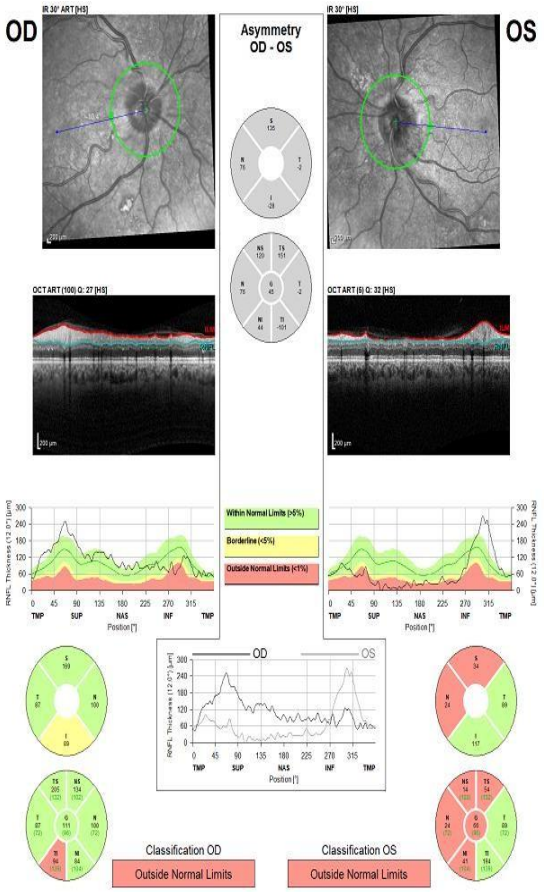

FIGURE 8: Papillary OCT shows regression of edema in the right optic disc with defect in inferior RNFL and nasal optic disc atrophy in the left eye

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