

**Case Report**

# Hickam's Dictum: "Post-Cataract Surgery" Ischemic Optic Neuropathy in a Setting of Atypical Chronic Inflammatory Demyelinating Polyneuropathy and Acute Angle Closure Glaucoma

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**ABSTRACT**

Non-arteritic ischemic optic neuropathy (NAION) is associated with crowded optic discs, hypertension, dyslipidemia, diabetes, and smoking [1]. In the past, cataract surgery was considered a potential risk factor. Recent reports have suggested that the frequency of NAION after cataract surgery is comparable to that of the general population risk [1-5], although this is still contested within the literature [4]. While NAION following acute angle closure glaucoma (AACG) has been described, there are no reports of NAION in the setting of Atypical Chronic Inflammatory Demyelinating Polyneuropathy (CIDP) [6-9]. In this report, we describe a case of a 60-year old male who developed NAION following lens extraction for AACG while being treated for atypical CIDP. This report documents an unusual case of NAION in association with acute angle closure glaucoma (AACG) and Atypical Chronic Inflammatory Demyelinating Polyneuropathy (CIDP) [6-9].

**Introduction**

Non-arteritic ischemic optic neuropathy (NAION) is the most common acute optic neuropathy with an incidence of 2-10 per 100,000 in adults over the age of 50 [1]. Post-cataract surgery ischemic optic neuropathy (PCION) is something that has been debated in the literature [1-5]. Recently, Moradi, et al. [1] reported that PCION occurs at a level comparable to that of NAION in the population, suggesting that modern cataract surgery is not a risk factor. Although NAION following acute angle closure glaucoma has been previously documented, it has not yet been seen in association with Atypical Chronic Inflammatory Demyelinating Polyneuropathy (CIDP). This report reviews a case of NAION following lens extraction for Acute Angle Closure Glaucoma (AACG) in the setting of atypical CIDP.

Potential associations and pathophysiological mechanisms will be discussed.

**Case Report**

A 60-year old Caucasian male was admitted to the intensive care unit for atypical CIDP with diaphragmatic, lower, and upper limb paralysis. Ophthalmology was consulted for severe right eye pain, redness, and decreased vision. Snellen acuity was 20/200 and 20/20 in the right and left eyes, respectively. The right pupil was mid-dilated and non-reactive, while the left pupil maintained good reactivity. The intraocular pressure via Reichert Tono-Pen was 55mmHg in the right eye and 20mmHg in the left eye. Bedside gonioscopy was performed using a portable slit lamp, and confirmed closed angles

in the right eye and narrow angles in the left eye. Ophthalmoscopy revealed a cup-to-disc ratio of 0.3 and 0.4 in the right and left eyes, respectively. His angle closure attack was medically treated given an inability to transfer and adequately position for laser peripheral iridotomy. He was undergoing treatment with monthly intravenous immunoglobulin G, tacrolimus, prednisone, pregabalin, and rituximab. Patient demographics and medical course are summarized in Table 1.

Following successful medical treatment of his AACG, the patient agreed to undergo lens extraction to minimize the risk of recurrent angle closure. The patient's upper and lower limb paralysis precluded him from undergoing laser peripheral iridotomy. Preoperative evaluation revealed a Snellen visual acuity of 20/20 in each eye. Intraocular pressures were 13mmHg in the right eye and 14 mmHg in the left eye via Tono-Pen tonometry while on timolol 0.5% twice a day in the right eye, brimonidine 0.1% twice a day in the right eye, and pilocarpine 2% twice daily in the right eye. Axial length was 22.64mm and 22.53mm in the right and left eyes, respectively. Anterior chamber depth was 1.93mm in the right eye and 1.92mm in the left eye. Measurements were obtained with ultrasound biomicroscopy. Sequential phacoemulsification and intraocular lens implantation were successfully performed under topical anesthesia first in the right eye, followed by the left. There were no intraoperative complications in either eye. Surgery in the right eye occurred 4 months following the AACG attack.

Post-operative examination of the right eye one week after surgery revealed a normal intraocular pressure (12 mmHg) and open angles on gonioscopy. At the one-month post-operative visit, the patient reported an inferior "shadow" in the visual field of his right eye. Visual acuity had decreased from 20/20 to 20/40 in the right eye and intraocular pressures were 12 mmHg and 14 mmHg in the right and left eyes, respectively. There was a right relative afferent pupillary defect. Ophthalmoscopy revealed right optic nerve swelling superiorly with papillary hemorrhages (Figure 1A and B). Spectralis optical coherence tomography confirmed right optic nerve edema (Figure 2A and B). Humphrey 30-2 visual field analysis demonstrated an inferonasal visual field deficit in the right eye (Figure 3). Repeat visual field analysis 8 months later showed a stable visual field defect.

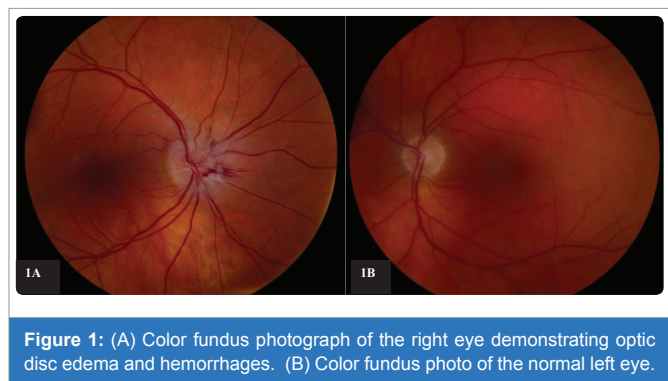
**Discussion**

This case is interesting in that it highlights the concurrent presence of three potential risk factors for NAION that have been reported in the literature. Perhaps the most interesting risk factor is cataract surgery, given its recent prevalence within the literature. While earlier studies reported an association between cataract extraction and NAION – the so-called "post-cataract surgery ischemic optic neuropathy" (PCION) – Moradi, et al. [1] have suggested the frequency of this is comparable to the general population, and is therefore not a real entity [1-3,5]. McCulley, et al. (2017) responded to this by stating that while incidence of PCION may be declining with modern surgery and anesthesia techniques, it would be reckless to disregard this as a potential complication based on a single retrospective study [1-4].

Acute angle closure has been implicated as a potential risk factor for NAION [6-9]. In fact, a recent case study has suggested that sudden IOP lowering in AACG with laser iridotomy is temporally related with optic disc swelling [10]. In cases of AACG, hemodynamic instability and/or ocular hypoperfusion secondary to acutely raised

**Table 1: Clinical Course and Patient Demographics.**

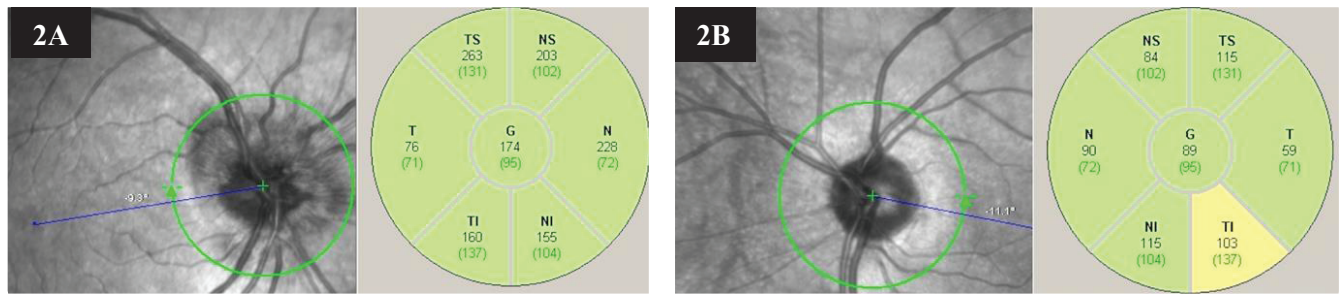
Age (Years)	60
Gender	Male
Admission to hospital for Atypical Chronic Inflammatory Demyelinating Polyneuropathy (CIDP)	7-Jul-17
Medical treatment for CIDP	Intravenous immunoglobulin G monthly
	Tacrolimus 0.10mg/Kg/day BID
	Prednisone 60 mg daily
	Pregabalin
	Rituximab
Onset of right eye acute angle closure glaucoma	15-Jul-17
Medical therapy for angle closure glaucoma	Timolol 0.5% twice a day in the right eye
	Brimonidine 0.1% twice a day in the right eye
	Pilocarpine 2% twice a day in the right eye
Right Lens Extraction	24-Aug-17
Onset right eye non-arteritic ischemic neuropathy (Post-cataract)	28-Sep-17



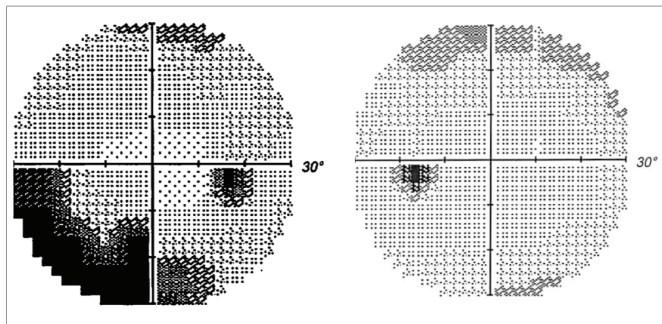
intraocular pressure are thought to contribute to subsequent ischemic optic neuropathy. However, to the authors' knowledge, AACG has never been associated with PCION.

Another consideration in this patient was his active treatment with IVIG for atypical CIDP. While CIDP in itself is not considered a risk factor for NAION, this patient was actively receiving intravenous immunoglobulin (IVIG) treatments. IVIG is generally considered safe, although there are increasing reports of adverse events as its use becomes more common. Specifically, IVIG therapy has been implicated with increased blood viscosity and a 3-5% risk of thromboembolic events secondary to pre-existing risk factors [11]. It is possible that the ongoing IVIG therapy for CIDP predisposed our patient to PCION.

An additional consideration is the predisposing role of the patient's ocular anatomy; his axial lengths were 22.64 mm and 22.53 mm in the right and left eyes respectively while his cup-to-disc ratios were 0.3 and 0.5 in the right and left eyes. While it has been previously suggested that a small optic disc area predisposes



**Figure 2:** (A) Optical coherence tomography analysis of the right eye optic nerve demonstrating edema as indicated by the optical coherence tomography retinal nerve fiber layer quadrant map (Micrometers) (B) Optical coherence tomography analysis of the left eye optic nerve demonstrating edema as indicated by the optical coherence tomography retinal nerve fiber layer quadrant map (Micrometers). Comparison between A and B demonstrated significant thickening in the superior temporal, superior nasal, nasal, inferior nasal, and inferior temporal in the right eye, consistent with nerve head edema.



**Figure 3:** The left figure shows a right eye Humphrey visual field 30-2 with an inferior nasal field defect (Visual field index 85%, mean deviation -7.09 dB, pattern standard deviation 11.39 dB) and right figure demonstrates the left eye with normal visual field (Visual field index 98%, mean deviation -3.32 dB, pattern standard deviation 2.22 dB).

patients to circulatory compromise, a large portion of the existing literature indicates that CDR and hyperopia are the major possible contributors to NAION risk [12-14]. This suggests that this patient's ocular anatomy may have contributed to this patient's PCION.

This case is interesting given the number of variables to consider. Classical teaching would suggest that one diagnosis explains all – Occam's razor. However, it is hard to ignore the number of suggested and potentially new risk factors for NAION presented in this case – Hickam's dictum. Based on Moradi, et al. [1] PCION is not a true entity and this case is due to chance alone [1]. However, it is hard to ignore that this patient had AACG, was receiving IVIG treatment, and had ocular anatomy that all posed risk factors for NAION [6-14]. These factors all placed the patient at greater risk for ocular vascular compromise which ultimately could have been acutely precipitated by the intraocular inflammation associated with IOL placement.

While this is merely a case report with no ability to imply causality, the authors would like to suggest several considerations: (1) PCION may be a true entity that has decreased in incidence with modern anesthesia and surgical techniques; (2) acute angle closure may be a risk factor for NAION; (3) IVIG therapy may rarely be associated with thromboembolic events and might be a risk factor for NAION; (4) ocular anatomy, in combination with other risk factors for ocular vascular compromise may put the patient at risk for PCION. It may be prudent for the cataract surgeon to consider

IVIG and AACG as potential risk factors for PCION. Additionally, the neuro-ophthalmologist may wish to enquire for any previous episodes of AACG or IVIG treatments when assessing their NAION patients. Further studies are required to assess the causal relationship between cataract surgery and NAION, as well as the potential risk of NAION following IVIG treatment.

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