Anterior Ischemic Optic Neuropathy in a Young Patient – Not as Rare as You'd Think!

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Introduction:

A 42 year old Caucasian male presents with a 'shade' in the superior field of view in the right eye. The patient has no medical problems, no history of medication use and is a non-smoker. Other than visual disturbance, a 12 point review of systems was negative. He denied a history of sleep apnea, use of amiodarone, erectile dysfunction medications, headache, weight loss or jaw claudication symptoms. The patient also denied any exposure to cats, recent vaccinations, or use of tetracyclines or vitamin A preparations.

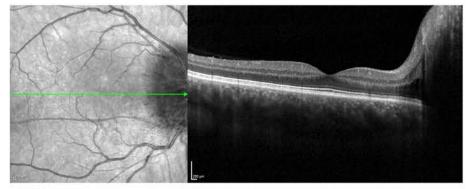
Exam:

Visual acuity was 20/20 in both eyes. A relative afferent pupillary defect was detected in the right eye. Visual field by confrontation revealed an superior

field defect in the right eye, and full in the left. Motility and intraocular pressure were normal. A Humphrey visual field confirmed a superior altitudinal defect of the right eye.

Dilated fundus examination of the right eye revealed an edematous nerve. The inferior edge of the disc was noted to have prominent edema with presence of hemorrhages and obscuration of vessels. The left optic disc appeared to be small and crowded, with a cup-to-disc ration of approximately 0.1. The rest of the examination was normal.

Optical coherence tomography (OCT) of the right eye revealed a normal macula with disc edema (Figure 1A). OCT of the left eye showed an ovoid area of lower



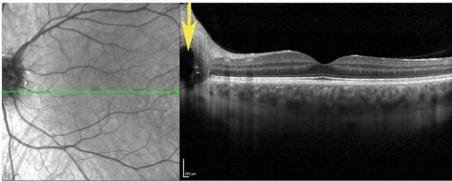


Figure 1A and 1B: OCT of both eyes. 1A (top) demonstrates disc edema. 1B (bottom) reveals a hyporeflective ovoid mass at the level of the optic disc.

reflectivity at the optic nerve (Figure 1B). Figure 2 is an OCT of the right optic nerve using a macular cube algorithm. Fundus autofluorescence (Figure 3) showed hyperautofluorescent structures with irregular borders at the optic disc.

Discussion:

This is an otherwise healthy young male who presents with vision loss secondary to an anterior ischemic optic neuropathy (AION). AION involves the anterior 1 mm segment of the optic nerve (optic disc) and results in disc edema. Two varieties of AION include non-arteritic (NAION) and arteritic (AAION). AAION is typically associated with giant cell arteritis. The majority of NAION cases are idiopathic, however some

associations have been reported. Though not definitively established, there are relationships between sleep apnea, amiodarone and optic disc drusen (ODD)¹.

Though AION is less common in younger patients, it is by no means rare. Preechawat et al report that 23% of AION patients were younger than 50, and 15% of AION patients were younger than 45 in a tertiary care neuro-ophthalmic service². In that series, 95% of patients were Caucasian,

and there was a slight predominance for males. However, if younger than 40, females were affected more frequently.

Compared to the typical AION patients, young patients with AION have better visual prognosis³. However, AION in young patients is more likely to be bilateral, upwards of 41% of cases according to one series2. In comparison, the Ischemic Optic Neuropathy Decompression Trial (IONDT) determined a 15% risk of bilateral involvement in patients older than 50 years⁴.

In young patients, optic disc structure likely plays an important role in developing AION. In patients under 50 with AION, 82% were found to have cup-less or the so-called 'disc at risk'². ODD are thought to be associated with younger age of onset of NAION.

especially ODD, buried, can be challenging to identify on examination. B-scan ultrasonography can effective in diagnosing deep ODD given their high reflectivity⁵. FAF, as demonstrated in our case, can be a convenient method of identifying superficial ODD. They typically appear as round/oval hyperautofluorescent structures

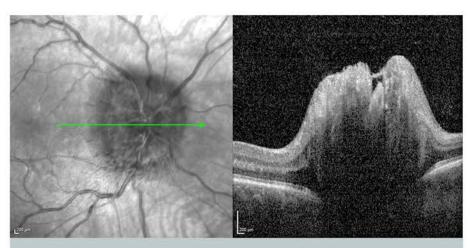


Figure 2: Disc edema of optic nerve of the right eye.

with irregular edges. Fluorescein angiography (FA) is more invasive method of visualizing ODD, which appear as mild hyperfluorescent smooth bodies in the peripapillary region typically noted in the late phase. Compared to optic disc edema, hyperflourescence is noted in early phases due to leakage. Telangiectatic vessels at the optic nerve head are revealed on FA in disc edema, where they are absent in ODD⁶.

Presently, there is no effective treatment for AION, however systemic workup should be performed to rule out other causes of bilateral visual morbidity. In older patients, it is certainly reasonable to perform a GCA workup in the setting of AION. In younger patients, assessing for microvascular risk factors such as hypertension and diabetes, hypercoagulable disorders, and anemia would be considerations.



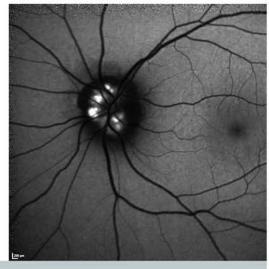


Figure 3: Hyperautofluorescent drusen in both nerves.

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