A rare case of simultaneous bilateral non-arteritic anterior ischemic optic neuropathy (NAAION) with retinal edema

Tyler Sommerfeld, O.D.

An 84-year-old male with asymmetric bilateral NAAION reports to the eye clinic due to a sudden onset of a black spot in the central vision of his left eye upon awakening.

- **Case History**
  - **Patient Demographics**
    - 84 year old white male living near Boston, MA
  - **Chief Complaint**
    - Sudden onset black spot in central vision OS 1 week prior upon waking
  - **Ocular History**
    - Mild epiretinal membrane OD
    - Meibomian gland dysfunction
    - Cataract extraction OD 06/2003, OS 10/2004
    - Compound Hyperopic Astigmatism, PCIOL OU
    - Last eye examination: 6 months prior
  - **Medical History**
    - Malignant neoplasm of the prostate
    - Impaired fasting glucose
    - Hyperlipidemia
    - Tendinitis
    - Gout
    - Tinnitus
    - Carpal tunnel syndrome
    - Osteoarthritis
    - Personal history of colonic polyps
    - Trigger finger
  - **Medications**
    - Allopurinol 300mg tablet PO qAM
    - Bicalutamide 50mg tablet PO every day
    - Calcium 250mg/Vitamin D 125 Unit 2 tablet PO every day
    - Finasteride 5mg tablet PO every day
    - Loperamide HCL 2mg capsule PO q6hr PRN
    - Nabumetone 500mg tablet PO BID PRN

- **Pertinent Findings**
  - **Clinical**
    - OS – fundus photos and HVF printout on file
      - BCVA 20/400 (20/20 9 months prior)
      - Positive 2-3+ left APD
      - Swollen disk, margins not distinct 360 degrees
      - Various drance hemes
      - Peripapillary edema surrounding nerve 360 degrees extending through nasal macula including fovea
- Pinpoint exudates throughout macula
- 1-2+ cells in anterior vitreous
- ? cells posterior vitreous above ONH
- HVF 30-2 low reliability, significant constriction with absolute defect greatest superior and nasal; sparing temporal to physiologic blindspot
- Historical C/D: 0.15R reported 9 months prior
  - OD - fundus photos and HVF printout on file
    - BCVA 20/20
    - Swollen disk greatest superior, margins not distinct sparing temporal ONH
    - (+) SVP
    - Peripapillary edema extending to within ¾ DD of fovea
    - 1-2+ cells in anterior vitreous
    - HVF 30-2 reliable, relative inferior arcuate scotoma
    - Historical C/D: 0.30R reported 9 months earlier
  - Physical
  - Laboratory studies
    - ESR: 28mm/hr (high-normal)
    - CRP: 0.32mg/L (normal)
    - Cerebrospinal fluid: clear
    - Lyme screen: negative
    - Toxoplasmosis: negative
    - FTA-ABS: nonreactive
    - RPR: non-reactive
    - Hepatitis C: negative
    - Hepatitis B: negative
    - PPD: negative
    - ACE: normal
    - ANA
  - Radiology Studies
    - Chest X-Ray: negative
    - CT scan: normal
- Differential Diagnosis
  - Primary
    - Simultaneous bilateral NAAION with peripapillary edema OS>OD
  - Others
    - Neuroretinitis
      - Cellular reaction is positive finding
      - Patient feels well, no recent history of malaise, fever
      - Infectious causes ruled out
      - FANG no disk leakage, no macular leakage
      - No macular star formation
    - Bilateral AAION
      - ESR high-normal
• CRP normal, ran twice
• No headache, no jaw claudication, no temple/scalp tenderness

- Papilledema
  • (+) SVP OD makes very unlikely
  • Peripapillary edema makes very unlikely
  • CT scan, MRI with and without contrast of head normal

- Bilateral Central Retinal Vein Occlusion
  • Visual fields do not agree
  • Positive APD does not agree

• Diagnosis and Discussion
  o Elaborate on the condition
    - Due to ischemia of the ONH, whose main blood supply is from the posterior ciliary arteries.
    - Subclinical ischemia leads to axoplasmic flow stasis of the optic nerve fibers. This leads to optic disk edema which compresses nearby capillaries leading to more ischemia.
    - Those with small C/D are at higher risk of precipitating the disease process.
    - Other risk factors are nocturnal hypotension, hypertension, diabetes mellitus, ischemic heart disease, hyperlipidemia, and atherosclerosis.
    - NAAION is more common in men than women, and more common in Caucasians than other racial groups
    - Patients often first notice painless decrease in vision after waking due to decreased blood pressure while sleeping.
    - Visual acuity ranges from 20/20 to NLP though most usually better than 20/40.
    - An APD will be present when NAAION is unilateral or asymmetric.
    - Studies have shown that visual field results vary, but NAAION is commonly associated with altitudinal defects
    - Optic disk edema is present at the onset of visual loss with possible associated splinter hemorrhages.
    - Pallor of the rim tissue develops 2-3 weeks after onset with overall resolution being about 6-12 weeks.
    - Visual acuity improves as often as it stays the same, and sometimes gets worse. Visual acuity and visual fields stabilize around 6 months after onset.
  o Expound upon unique features
    - NAAION is almost exclusively unilateral or sequential. The chances of the fellow eye ever developing NAAION after a unilateral NAAION is reported to be only as high as 25%. Very few case reports can be located regarding bilateral simultaneous NAAION.
NAAION may rarely also present with peripapillary and macular subretinal fluid. In this case, both eyes were found to have thickening at onset OS>OD.

The patient only reported visual symptoms in his left eye due to the asymmetry of disease between his eyes.

- Treatment, Management
  - Treatment
    - Extensive lab and imaging analysis performed to rule out differential diagnoses mentioned above
      - All results and images on file including fundus photos, FANG, CT scan, MRI with and without contrast, blood test results, cerebrospinal fluid analysis
    - No active, intervening treatment was initiated
    - Made sure patient was not suffering low blood pressure, nocturnal hypotension.
    - The patient was monitored closely for multiple weeks after onset with gradual resolution of retinal thickening and ONH edema.
    - Nerve appearance OU gradually became pale – photos on file**
    - Currently stable visual acuity OD 20/20, OS 20/40
    - The patient is now scheduled for yearly eye examination including HVF testing.
  - Refer to research where appropriate
    - There are studies that suggest treatment with oral steroid may improve the visual acuity and visual field outcome (Hayreh, 2008).
    - Common patient education is to decrease risk factors when possible (Hayreh, 2009).
  - Bibliography, literature review encouraged

- Conclusion
  - Clinical pearls, take away points
    - Classic presentation for NAAION painless loss of vision, positive APD, optic disk swelling with associated visual field defect.
    - Take fundus photos, like in this case, so that they may be referred to during the disease process.
    - Perform OCT around nerve and macula to detect any subretinal fluid
    - A work-up must be completed to rule out giant cell arteritis/temporal arteritis.
    - If bilateral, other causes more likely and must be investigated via imaging, blood, and cerebrospinal fluid analysis.
    - Corticosteroid therapy may be beneficial to visual outcome when administered in the acute phase of disease.
    - It is important to decrease manageable risk factors when possible: hypotension, hypertension, diabetes mellitus, hyperlipidemia, and atherosclerosis.