Title:
A Case of Unilateral Optic Nerve Head Drusen with Severe Visual Field Loss

Abstract (35 word max)
Optic nerve head drusen is an abnormal finding usually found first on routine examination. Typically benign and asymptomatic, the following details an abnormal case of unilateral drusen causing symptomatic field loss.

Case History
- Demographics: 71 year-old white, male
- Chief complaint: Longstanding subjective side vision loss in OD, longstanding watering OS.
- Medical History: Non-insulin dependent diabetes mellitus (dx 2007), Hypertension, hyperlipidemia, Coronary Artery Disease (dx 2008)
- Medications: Aspirin 81mg, furosemide 40 mg, simvastatin 20mg, glipizide 30mg, metoprolol 100mg, ompeprazole 40mg, diclofenac 50mg, saxagliptin 2.5mg, nitroglycerin 0.3mg

Examination
- VA cc: OD: 20/20, OS:20/40 PH: ni
- Pupils: PERRL-No APD
- Fields: Restriction OD: sup and inf temp, sup nasal, OS FTFC
- Intraocular pressure: OD 11 mmHG, OS 13 mmHG
- Conjunctiva: Tr injection OU
- Cornea: Clear OU
- Anterior chamber: Deep and quiet OU
- Lens: Tr NS OU
- Fundus:
  Optic nerve - OD: Blurred disc margins with visible white round lesions superior, nasal and temporal on the disc. OS: Distinct rim tissue, small cupping
  C/D Ratio: OD: non-existent, OS: 0.10
- Macula-
  OD: Mild hard drusen
  OS: RPE pigment clumping with surrounding hypopigmentation

Pertinent findings
- Humphrey Visual field 24-2: 2010
  o OD: Unreliable (FN 42%). MD: -18.42, PSD: 12.44, GHT ONL
    -Dense inferior arcuate scotoma, mild superior nasal step
  o OS: Reliable. MD: -2.09, PSD: 4.97, GHT ONL
    -Superior nasal cluster defect
- Humphrey Visual field 24-2: 2014
- OD: reliable. MD: -22.34, PSD: 10.40 GHT: ONL
  - Dense inf->sup arcuate
- OS: unreliable (FL 7/10), MD: -0.48, PSD: 1.65 GHT: WNL
  - Mild sup nasal, inf cluster of defect

**Humphrey Visual Field 24-2: 2016**
- OD: MD -23.00, PSD 10.68, GHT ONL.
  - Dense inferior arcuate scotoma with moderate superior arcuate nasal
- OS: MD +0.29, PSD 1.71, GHT Borderline.
  - Few non-repeatable nasal defects

**B-scan ultrasound 2016:**
- OD: Hyper reflectivity at optic nerve
- OS: Flat and clear retina, no hyper reflectivity noted

**OCT RNFL 2008**
- OD: Thinning superior, inferior and temporal. Average thickness decreased to 55.38.
- OS: Full thickness 360. Average thickness normal at 109.52.

**OCT RNFL 2016**
- OD: sup 77, inf 89, tot 72, small round lesions visible
- OS: sup 140, inf 151, tot 122.

**Differential Diagnosis**
- Optic nerve head drusen
  - Anomalous vascular patterns, central elevation
  - White calcified appearance on surface of the disc developing over lifetime
- Optic Nerve Head Hypoplasia
  - Abnormally small optic nerve head due to a low number of axons
  - Pale or greyish appearance with surrounding peripapillary halo
- (Non)-arteritic Ischemic Optic Neuropathy
  - Occlusion of the short posterior ciliary arteries
  - Hyperemic disc swelling with associated flame hemorrhages
  - Typically altitudinal field defects
  - + APD and typically decreased VA
- Diabetic Papillitis/ Optic Neuritis
  - Typically in insulin dependent diabetics
  - Swelling of the optic nerve similar to Non-arteritic ischemic optic neuropathy
  - Uncontrolled blood glucose and diminishing vision
- Optic pit
  - White or grey depression in optic nerve, typically temporal
- Papilledema
  - Typically bilateral and symmetric elevation of optic nerve head and hyperemia
  - Appears with Paton’s lines, flame shaped hemorrhages, cotton wool spots and tortuous retinal vessels
- **Myelinated NFL**
  - White and feathery patches that follow along the nerve fiber layer lines
  - Commonly obscure blood vessels and the margins of the disc
- **Primary-open angle glaucoma**
  - Progressive damage to optic nerve head leading to peripheral vision loss
  - Typically bilateral in the presence of increasingly large, flat optic nerve cupping

**Diagnosis and Discussion**

Optic nerve head drusen are globular, calcified, hyaline bodies that form within the optic nerve head. While the etiology is generally unknown, the current theory is a believed lack of space through the scleral foramina leads to a stasis of axoplasmic flow and development of drusen. The incidence is between 0.3-2 percent of the population and many times are hereditary appearing in multiple generations.

Seventy-five percent of presentations are bilateral, starting as buried drusen in children, that slowly move forward with age. This anterior displacement leads to exposure and changes in the appearance of the retinal nerve fiber layer sometime within the teenage years. These retinal nerve fiber layer disruptions may cause visual field defects that are susceptible to change over time with the movement of the drusen and can mimic glaucomatous damage.

Initial presentation of optic nerve head drusen can be similar to papilledema, so ancillary testing to confirm the diagnosis is necessary. Fluorescein angiography and fundus autofluorescence show hyperfluorescence at the nerve due to staining of the drusen, and B-scan ultrasonography will show hyper-reflectivity at or deep to the disc. Computer Tomography can also identify drusen, but is less sensitive and more time consuming.

Recent innovations with optical coherence tomography make the test more useful diagnostically. Studies show it can be used to differentiate between disc edema and drusen based on the appearance of the scan within the nerve and in the peripapillary area. When looking at the scan of the nerve, optic nerve head drusen are visible as round bodies within the elevated RNFL, while disc edema will present with smooth elevation. Additionally, differentiation can be made in the paripapillary area. Drusen have elevation at the area of the nerve with an abrupt return to the expected orientation of sensory retina to retinal pigment epithelium. In disc edema, the space between these layers is larger and extends further from the nerve. This diagnostic differentiation can be helpful in primary care settings where more expensive diagnostic tests are not available and the use of optical coherence tomography is already employed.

Our patient has a rare unilateral presentation, with ancillary tests confirming no presence of buried drusen in the fellow eye. The patient has a longstanding history of exposed drusen with visual field testing showing severe visual field
defects in the affected eye. The patient, while initially asymptomatic, has become symptomatic for diminishing peripheral vision over time.

**Treatment and Management**

Optic Nerve head drusen is associated with ocular diseases such as retinitis pigmentosa, angiod streaks and Alagille syndrome. There are rare, yet significant, complications with optic disc drusen such as juxtapapillary choroidal neovascular membranes, disc neovascularization, central retinal arterial and central vein occlusion, Non-arteritic ischemic optic neuropathy and progressive visual field loss due to loss of nerve fiber layer bundle patterns. Patients should be screened yearly for risk of asymptomatic complication and any changes in the structure or function of the optic nerve. Appointments should include visual field testing, dilated fundus examination and optical coherence tomography of the retinal nerve fiber layer.

Currently there are no treatments for optic nerve head drusen. Recently, management has included the use of brimonidine twice daily in the effected eye or eyes to reduce chances of additional complications such as a non-arteritic ischemic optic neuropathy and damage from an unknown comorbidity of glaucoma. Typically used for glaucoma treatment, this drop can decrease intraocular pressure, increasing optic nerve head perfusion providing neuroprotection. Regular assessment of visual function along with dilated examination provides the best prognosis for long-term management of patients with optic nerve head drusen.

Our patient was made aware of the risks associated with his diagnosis along with demonstrated the peripheral vision loss from the changes in the retinal nerve fiber layer. The patient was given the option of brimonidine management for neuroprotective purposes, but chose to defer at this time. Patient is aware of importance of regular examination with visual field to determine any functional changes.

**Conclusion**

Optic nerve head drusen has potential to be a serious detriment to a patient's vision. Our patient is fortunate that the nature of his disease that causes significant vision loss is only present in one eye. Regular screening of ocular health and management of comorbidity is essential in this somewhat common optic nerve head anomaly. Fortunately, innovation continues to allow for effective in-office screening for differentiation and management.

**Bibliography**
