Title: Advanced Visual Field Loss Secondary to Optic Nerve Head Drusen and Analysis Using Optical Coherence Tomography

Abstract: Optic nerve head drusen is usually a benign condition but can cause significant visual loss in advanced cases. Optical coherence tomography may be beneficial in diagnosing optic nerve head drusen compared to the current standard.

I. Case History
   A. Demographics: A 68-year-old white male
   B. Chief complaint: vision slightly reduced at near greater than at distance
   C. Ocular History:
      • Optic nerve head drusen
      • Vitreomacular traction OU
      • Strabismic amblyopia OD s/p surgery
   D. Medical history:
      • Post traumatic stress disorder
      • Allergic rhinitis
      • Chlamydia
      • Prostatic hypertrophy
   E. Medications:
      • Citalopram, Finasteride, Sildenafil, Terazosin, and Acetaminophen

II. Pertinent findings
   A. Clinical
      • BCVA: 20/40-2 OD, 20/20-2 OS
      • Pupils: PERRL, -APD
      • CVF: FTFC OD/OS
      • Alignment: constant right exotropia and hypotropia
      • Extraocular motility: right inferior oblique underaction OD
      • SLE: unremarkable
      • Applanation tonometry: 10 mmHg OD, 11 mmHg OS
      • DFE: 0.1 x0.1 C/D in 2.0mm VDD with superficial drusen 360° OU
         i. Irregular disc margins OU
         ii. Macula/Vessels/Periphery: Normal OU
   B. Physical
• Alert and oriented
• Mood/affect appropriate

C. Other testing
• 30-2 visual fields:
  o Dense superior arcuate defect > inferior nasal defect OD>OS; repeatable and stable
  o OD: MD -18.91 DB, PSD 13.19 DB
  o OS: MD -10.56 DB, PSD 10.62 DB
• Spectralis OCT:
  o RNFL:
    ▪ OD: Global thickness 48um, thin all quadrants except superior temporal
    ▪ OS: Global thickness 57um, thin globally, inferior temporal and inferior nasal
  o Optic nerve head with enhanced depth imaging: reflective areas consistent with intra-papillary drusen
• Pachymetry: 564um OD, 571um OS

III. Differential diagnosis
A. Primary: superficial optic nerve head drusen OU
B. Others: disc edema

IV. Diagnosis and discussion
A. Elaborate on the condition
  • Optic nerve head drusen (ONHD) are a collection of calcium phosphate crystals buried underneath the optic nerve head anterior to the lamina cribosa, which can become more superficial and clinically visible over time.
  • Eyes with ONHD also present with abnormal blood vessel patterns, including vessel trifurcations, tortuosity, and optociliary shunt vessels.
  • The condition often presents bilaterally, has no gender predilection and is more common in Caucasians.
  • Optic nerve head drusen tends to be benign, and patients are usually asymptomatic. Therefore, it is most often diagnosed clinically and has traditionally been confirmed with B scan ultrasonography when the diagnosis is in question.
  • Clinical prevalence of the optic nerve head drusen is reported as approximately 0.3% but autopsy reports have shown a higher prevalence of 2.4%. This may be due to the asymptomatic nature of the condition.

B. Unique features:
ONHD rarely affect visual acuity, but 24-87% of cases have been reported to have measurable visual field defects ranging from enlarged blind spot to concentric narrowing.

V. Treatment, management

A. Treatment and response to treatment
- There is currently no treatment for this condition but lowering intraocular pressure (IOP) has been shown to slow progressive field loss when IOP is elevated. With drusen confounding the nerve appearance, field loss, and nerve fiber loss, it is difficult to assess the optic nerve for other ocular diseases, such as glaucoma. Therefore, prompt initiation of treatment should be considered if the patient has other risk factors for vision loss, such as high intraocular pressure.
- Serial visual fields should be performed periodically to assess for progressive field loss
- OCT-RNFL can assess the general health of the optic nerve but has poor spatial correlation with drusen associated field loss

B. Refer to research where appropriate
- Multiple theories exist on how and why ONHD develops. Sacks et al attributed it to the congenital vascular abnormalities that enable extracellular material deposits. Tso proposed that drusen formation is associated with axonal degeneration of the optic nerve head secondary to abnormal axonal metabolism that causes calcium crystals to deposit in the mitochondria. Kapur et al supported this idea by histologically identifying calcium phosphate, thought to be associated with cell death, in surgically removed ONHD material. Other theories suggest that patients with ONHD have smaller scleral canals that cause abnormal axonal metabolism.
- OCT may be a beneficial diagnostic tool for detecting drusen in suspicious nerves versus the standard B scan.
- EDI-OCT has been reported to be able to quantify drusen dimensions and assess integrity of neighboring retinal structures. Therefore it may be beneficial in assessing the relationship among ONHD, visual field loss, and retinal nerve fiber loss.

VI. Conclusion

A. Patients with ONHD should be routinely monitored to detect visual field loss and counsel patients appropriately.
B. OCT should be considered in the work-up of ONHD as a diagnostic tool and to assess the health of the optic nerve.
References


