Acute Zonal Occult Outer Retinopathy: The Role of Auxiliary Testing in Diagnosis and Management
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Abstract
A 25 year old male presents with an acute unilateral scotoma. This case report illustrates the importance of auxiliary testing in diagnosing and monitoring functional changes of the retina in acute zonal occult outer retinopathy.

I. Case History
a. Patient demographics
   25 year old Caucasian male
b. Chief complaint
   Sudden onset of darkened area of vision inferiorly and infra-nasally out of the left eye
c. Ocular, medical history
   Psoriasis
d. Medications
   Currently using a steroid cream as needed for psoriasis
e. No history of ocular trauma, surgery, or infection

II. Pertinent Findings
a. Clinical
   1. BCVA uncorrected:
      • OD: 20/20
      • OS: 20/20
   2. Pupils: Round and reactive to light with a 1+ RAPD OS
   3. Color vision with Ishihara: normal OD, OS
   4. Anterior segment evaluation:
      • OD: unremarkable
      • OS: mild anterior chamber reaction, mild anterior vitritis
   5. Posterior segment evaluation:
      • OD: c/d 0.40 round, flat and evenly pigmented macula
      • OS: c/d 0.50 round, mild granulation of macula, faint multiple white/grey dots nasal to the optic nerve which resolved within a few days
b. Laboratory studies
   1. Complete blood count: within normal limits
   2. Rapid plasma reagin: non-reactive
   3. Tuberculosis: negative
   4. Lyme titer: negative
   5. Toxoplasmosis titer: negative
c. Radiology studies
   ○ MRI: results pending
d. Multifocal Electroretinogram (mfERG)
   - Right eye: Responses are normal with amplitudes for all 103 locations being normal.
   - Left eye: Responses were reduced to below normal limits at all but 5 of the 103 locations. Responses within the fovea tended to be larger, which gives the impression of an annular pattern of retinal dysfunction. The pattern of ERG waves are consistent with AZOOR.

e. Fluorescein Angiography
   - OD: normal
   - OS: normal

f. HVF 30-2
   - Multiple HVF 30-2 were performed in order to monitor the scotoma.
   - All visual fields were reliable, and the right eye remained unaffected with no visual field defects.
   - The left eye initially had a dense inferior scotoma with absolute defects. As the visual fields were repeated and scotomas monitored, the scotoma grew to become a dense ring scotoma with absolute defects in the superior and inferior fields, involving the blind spot.
   - After starting oral corticosteroids treatment, there is resolution of the superior visual field defects, and the inferior scotoma remains.

g. Macula OCT
   - OD: normal foveal contour, intact photoreceptor integrity line
   - OS: slightly irregular foveal pit, disruption of photoreceptor integrity line, elevated intraretinal lesions

III. Differential Diagnosis
   a. Primary/leading: Acute zonal occult outer retinopathy (AZOOR)
   b. Secondary differential diagnoses: other causes of scotomas with a relatively normal looking fundus
      - Autoimmune retinopathy (including cancer associated retinopathy, melanoma-associated retinopathy, and non-neoplastic autoimmune retinopathy)
      - Multiple evanescent white dot syndrome (MEWDS)
      - Idiopathic acute optic neuritis, retrobulbar

IV. Diagnosis and Discussion
   a. Elaborate on the condition
      - Acute zonal occult outer retinopathy (AZOOR) is a rare condition of unknown etiology characterized by degeneration of photoreceptors. This condition is characterized by the presence of sudden photopsia and acute scotomas, which are related to loss of outer retinal function. AZOOR generally occurs unilaterally in young adult, Caucasian, myopic women where the fundus initially appears normal with possible development of retinal pigment epithelial atrophy or pigment clumping.
Prognosis is generally good, as central vision remains 20/40 or better, even with electroretinogram changes and persistent visual field defects.

b. Expound on unique features

   o Signs
     ▪ RAPD present in 21% of AZOOR patients
     ▪ Vitreous cells present in 23% of AZOOR patients
     ▪ Visual field
       • Most common: enlarged blind spot
       • Possible to have ring scotoma, arcuate defects, or multiple isolated scotomas
       • Central vision spared
     ▪ FA usually normal
     ▪ Fundus appearance usually normal or minimally involved
     ▪ OCT usually presents with disruption of the IS/OS junction
     ▪ mfERG usually shows very depressed retinal function

   o AZOOR complex: Idea that AZOOR lies on a spectrum of white dot syndromes, allowing for some overlap or co-existence of two conditions at one given time.

V. Treatment, Management

   a. Treatment and response to treatment
     ▪ Literature contains arguments in support of both autoimmune and infectious processes as the etiology of AZOOR, however there is currently no treatment that has proven to improve visual outcome. Different forms of therapy have been attempted, including antimicrobials, antivirals, NSAIDs and the most commonly attempted therapy is systemic corticosteroids.

     ▪ The patient was treated with a slow taper of oral prednisone, and is currently on the taper schedule. Although AZOOR may be self-limiting, there were no contraindications for oral corticosteroid use, and the patient selected for treatment over observation alone.

     ▪ Although this condition is usually self-limiting to a certain degree, with the current treatment of oral corticosteroids, the patient’s superior visual field defect is resolved. The inferior visual field defect is what remains.

     ▪ A multi-focal ERG is scheduled in the near future to monitor changes in retinal function that corresponds with changes to the visual field.

   c. Bibliography


     ▪ Spaide, R.F., et al. “Photoreceptor Outer Segment Abnormalities as a Cause of Blind Spot Enlargement in Acute Zonal Occult Outer


VI. Conclusion

a. Clinical pearls, take away points

○ AZOOR is a diagnosis of exclusion, but may have some overlap with and even co-exist with white dot syndromes such as MEWDS.

○ In the presence of visual field defects with a normal appearing fundus, we must rule out retrobulbar etiologies.

○ Multi-focal ERG assesses outer retinal layers and photoreceptor function, making it useful for early diagnosis of AZOOR.