A Rare Presentation of Cystoid Macular Edema Due to Acute Posterior Multifocal Placoid Pigment Epitheliopathy
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Abstract: A rare case of Acute Posterior Multifocal Placoid Pigment Epitheliopathy with an atypical presentation highlights the careful evaluation, diagnosis and treatment in patient’s with this disease.

I. Case History
- Patient demographics: 34 year old white male
- Chief complaint: sudden distortion of vision OD x1day with longstanding distortion OS first noticed while on the computer. Squinting does not help. Patient also complains of issues with depth perception and has been tripping while walking.
  - mostly central vision affected with stable blur at distance
  - wants to know if glasses would help
  - also complains of difficulties with glare x2months
  - Denies diplopia, eye pain, headaches, floaters or any redness. Denies any recent or current illness.
- Ocular history: previously seen 3 months prior at UEC for evaluation of intermittent photopsia with constant peripheral blur following his grandmother’s funeral 1 week ago. Last comprehensive eye exam x4 years ago. WNL per patient.
  - Medical History: LME: 3 years ago; borderline HTN, Alopecia.
  - Medications: Propecia
  - TBI 3 years ago during a bar fight. Patient was intoxicated and does not know many details, but was hospitalized and did have surgery for resultant hematoma.
- Occupation: CPA
- FHx: GLC (MGM), MS (Mother)

II. Pertinent findings
- Clinical:
  - BCVA 20/25+2 OD 20/400 OS PHNI OD, OS
  - EOMs, Pupils, CVF, Anterior Segment wnl
  - IOP: 14/14 OD, OS
  - Vitreous: clear OD, OS
  - Posterior pole: OD C/D: 0.2/0.2, NFL wnl OS C/D: 0.2/0.2, slightly elevated nerve with irregular borders
  - vessels: normal caliber; 2/3--areas of whitening surrounding vasculature in posterior pole OD and OS
  - OD macula: slight elevation
  - OS macula: large elevation with cystoid macular edema
  - OD periph: inferior and inferior temporal chorioretinal scar
  - OS periph: inferior scarring with pigmentation.
  - Amsler Grid:
    - OD- paracentral and central metamorphopsia, circular pattern
    - OS- paracentral and central metamorphopsia, more pronounced than OD.
• IVue OCT: OD: mild macular edema with small PED OS: severe cystoid macular edema; 754 microns thick with underlying PED within 1mm of fovea
• Fluorescein Angiography(at outside retina specialist): early blockage and late staining in areas of lesions OD, OS; petalloid leakage OS in macula
• Laboratory:
  Blood work was ordered in conjunction with the patient’s primary care doctor and retinal specialist to rule out possible viral, infectious or autoimmune component. Results are pending.

III. Differential diagnosis
• Primary/leading: Acute Posterior Multifocal Placoid Pigment Epitheliopathy (APMPPE)
• Others: Vitelliform Dystrophy, Haranada’s disease, Birdshot Chorioretinopathy, Multifocal Choroiditis, Multiple Evanescent White Dot Syndrome, Cat Scratch Fever, Sarcoidosis, Lyme Disease

IV. Diagnosis and discussion—
• APMPPE is characterized by a rapid, transient loss of visual acuity with acute onset of multifocal discrete yellow-white placoid lesions at the level of the RPE.\(^1\)
• Patient’s usually present with scotomas or metamorphopsia bilaterally (or sequentially typically within 6 months after the initial episode).\(^2,4\)
• The mean age of onset is 26.5 years old. No sex or racial predilection has been demonstrated.\(^2\)
• APMPPE is a rare entity, diagnosed in only five patients over one 10-year period at the Immunology and Uveitis Service of the Massachusetts Eye and Ear Infirmary.\(^3\)
• One-third of patients present with a viral prodrome: fever, malaise, headache, dizziness and myalgia. Less frequently, hearing loss, tinnitus or vertigo.\(^5\)
• The characteristic lesions typically resolve over several weeks leaving depigmentation and pigment clumping with great improvement in visual function.\(^1,6\)
• Reports have linked some cases of APMPPE with cerebral vasculitis, erythema nodosum and sarcoidosis suggesting a possible viral link.\(^2,7-11\)
• Typical fluorescein patterns show early blockage/masking due to overlying edematous patches with mid-to-late diffuse, even staining of lesions.\(^9,12\)
• Typical OCT findings show hyper reflectance of the outer retinal layers without increase in retinal thickness.\(^13\)
• Papillits is an atypical, but possible feature thought to reflect involvement of the choroidal supply of the optic nerve head.\(^14\)
• Serous retinal detachments in patient’s with APMPPE have been commonly described in literature.\(^15\)
• Few cases of cystoid macular degeneration in APMPPE have been reported.\(^16\)

V. Treatment, management
• Treatment and response to treatment: Cystoid macular edema was treated with 4mg/0.05mL injection of Triescence OS. At one week follow-up, the patient achieved 20/70+ BCVA OS with 20/20 stable VA OD. The macular edema and serous detachments had resolved, but some placoid lesions were still present. Patient is due to return to UEC in 2 weeks for additional follow-up.
• Patients have also been successfully treated with systemic corticosteroids in cases of APMPPE without cystoid macular edema with positive outcomes.\(^14\)
VI. Conclusion

While APMPPE is a rare disorder, it can present with cystoid macular edema similar to other diseases. While more research must be done to determine its etiology, intravitreal steroids remain beneficial in cases of APMPPE presenting with cystoid macular edema.

Bibliography: