Case Report: Acute Exudative Polymorphous Vitelliform Maculopathy (AEPVM)

Jayson Madriaga, OD
Paul Vejabul, OD
Jennifer Monarrez, OD

Abstract
With about twenty cases reported in the literature since 1988, AEPVM is a rare acute disorder. Similar to previously reported cases, this case demonstrates the decrease in vision within weeks to months without corticosteroid therapy.

I. Case History

- **Patient demographics:** 87 year old Caucasian male
- **Chief complaint:** Gradual decrease in vision OU which started around October 2014. Patient denies any persistent headache, flu-like events, or viral diseases around time of occurrence.
- **Ocular history:** No significant ocular history
- **Medical history:** Vitamin D Deficiency, aortic valve disorder, COPD, UTI, tobacco use disorder, GERD, occlusion and stenosis of carotid artery, CAD, elevated PSA, hypertension, hearing loss, peptic ulcer, hyperlipidemia, hypertrophy of prostate
- **Medications:** Amlodipine, albuterol, atorvastatin, metoprolol, omeprazole, terazosin, aspirin

II. Pertinent findings

- **Clinical**
  - Right Eye:
    - Best corrected visual acuity (BCVA) 20/30; unremarkable dilated fundus exam (DFE), fundus photos, fluorescein angiography (FA), and optical coherence tomography (OCT)
  - Left Eye:
    - BCVA 20/40 at initial visit
    - DFE/fundus photos exam: Macular scar with RPE clumping and large thickened surrounding area
    - OCT Macula: Large serous detachment with a normal foveal contour, underlying pigment epithelial detachment (PED), and scar tissue. No evidence of intraretinal cyst. Remarkable for hypertrophy of the outer retinal segments.
    - FA: Serous fluid leakage/pooling surrounding disciform scar at the macula with minimal evidence of diffusion of dye into the surrounding subretinal exudation
Electrooculogram (EOG): Patient was not a good candidate for EOG due to age and fatigue. He exhibited slow and inaccurate saccades throughout the test and was constantly blinking, rendering the results inconclusive.

III. Differential diagnosis

- **Primary**: Acute Exudative Polymorphous Vitelliform Maculopathy OS
- **Others**: Central serous chorioretinopathy, pigment epithelial detachment, exudative age-related macular degeneration, pattern dystrophy, adult vitelliform dystrophy, Harada’s disease, ocular lymphoma, acute multifocal placoid pigment epitheliopathy

IV. Diagnosis and discussion

- **Diagnosis**: Acute Exudative Polymorphous Vitelliform Maculopathy OS
- **Discussion**:
  - There have been many hypotheses of the causes of AEPVM, but most research has shown the cause to be at the level of the RPE where there is an autoimmune retinal pigment epitheliopathy trigger from a viral infection, a neoplasm, or some other autoimmune process.
  - Although the causes are unclear, each reported case has an underlying characteristic: bilateral serous retinal detachment and subretinal accumulation of hyperautofluorescent yellowish material in the posterior pole. This case is atypical because it is unilateral and does not have prominent yellowish deposits in the posterior pole.
  - With the help of the OCT, previous cases have linked the yellow deposits to shallow neurosensory retinal detachments and others to RPE detachments.
  - Treatment options include observation or steroids. Oral or intravitreal injections of steroids have been used in the past, but with, mixed results.

V. Treatment, management

- **Management**:
  - Referral to retina specialist
- **Treatment**:
  - In the literature, AEPVM is usually self-limiting with full recovery that can span from 12 to even 24 months. Oral and intravitreal injections of steroids have been used in the past with little success. More reported cases have led to normal macular thickness with no residual effect on visual acuities with observation alone.
  - No sequelae has occurred in the 20 cases reported
  - Further testing to correlate potential causes may include: metastatic cutaneous melanoma, inflammation, viral infection, infections of the upper respiratory system, sinus infections, and paraneoplastic syndromes.
- **Prognosis**: Unknown
- **Results**: Patient has been seen for six times to date with follow-ups spaced between 5-9 weeks apart:
  - BCVA OS fell from 20/40 at initial visit to 20/70+2 at the most recent visit
- Fundus appearance and macular OCTs have remained stable
- Pt not symptomatic enough to want treatment and wishes to observe

**Bibliography**


**VI. Conclusion**

- **Clinical pearls**
  - As research continues, diagnostic and treatment modalities will also be improved. Continuing education is vital.
  - When a diagnosis is unclear, the use of relevant ancillary testing and literature review can aid in the diagnosis and management of difficult cases.

**Comments to Reviewer:**
Clinical images available for presentation: Fundus photos, FA photos, serial OCT images

Primary Topic: Posterior Segment

Keywords: Acute Exudative Polymorphous Vitelliform Maculopathy, Polymorphous Maculopathy Syndrome