Acute Posterior Multifocal Placoid Pigment Epitheliopathy
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Abstract
- A 41yo white female presents with APMPPE. Treatment with oral corticosteroids is initiated and the multiple placoid lesions resolve. Disease presentation, treatment, and current research are presented here.

I. Case History
- Demographics: 41yo white female
- Chief Complaint: New onset floater OD x 1 day in the patient’s peripheral vision
- Ocular Hx: New onset floater with flashes OS x 3 days prior with a DFE by the referring optometrist revealing 4 white lesions in the posterior pole OS
- Medical Hx: Flu-like symptoms 1-2 weeks prior, sinus congestion, and recent blood work within normal limits including (-)ANA and (-)RF
- Medications: Sinus medication (unknown)

II. Pertinent Findings
- 1st Exam (Day 1)
  - BCVA: 20/25 OD, 20/25 OS
  - IOP with applanation tonometry: 12mmHg OD, 13mmHg OS
  - Anterior segment: Unremarkable, no A/C cells or flare
  - Posterior segment: 1 circular yellow-white placoid lesion OD superior to the macula, 4 circular yellow-white placoid lesions OS with 1 inferior to the macula and the remaining 3 in the superior arcade. No vitreous cells and no posterior vitreous detachments are present.
- 2nd exam (Day 4)
  - BCVA: 20/25 OD, 20/25 OS
  - IOP with tonopen: 18mmHg OD, 18mmHg OS
  - Anterior segment: Unremarkable, no A/C cells or flare
  - Posterior segment: Increase in placoid lesions OS, no vitreous cells present
  - See photos below
- 3rd Exam (Day 11)
  - BCVA: 20/25 OD, 20/40 OS
  - IOP with tonopen: 16mmHg OD, 16mmHg OS
  - Anterior segment: Unremarkable, no A/C cells or flare
  - Posterior segment: Increase in placoid lesions OU, no vitreous cells present
  - Tx: Oral prednisone 30mg PO qd
- 4th Exam (Day 18)
  - BCVA: 20/20 OD, 20/40 OS
  - IOP with tonopen: 17mmHg OD, 17mmHg OS
  - Anterior segment: Unremarkable, no A/C cells or flare
  - Posterior segment: Resolution of several placoid lesions OU with remaining lesions showing an increase in pigmentation, no vitreous cells present
  - Tx: Oral prednisone 30mg PO qd x 1 day, 20mg PO qd x 10 days, 10mg PO qd x 10 days
• Laboratory studies
  o Chest x-ray: within normal limits

• Imaging
  o Fluorescein Angiography: numerous placoid lesions that are hypofluorescent in early phase and hyperfluorescent in late phase OS

III. Differential Diagnoses
• Primary – Other white dot syndromes
• Other – Metastatic tumors, sarcoidosis, syphilitic retinitis, bacterial or viral retinitis, toxocariasis, toxoplasmosis, histoplasmosis, VKH

IV. Diagnosis & Discussion
• APMPPE is a rare, acute inflammatory condition found in healthy adults that is characterized by multiple yellow-white placoid lesions in the posterior pole at the layer of the RPE. It is a self limiting condition with favorable prognosis. It is classified as one of the white dot syndromes which are simply a group of inflammatory disorders that are characterized by multiple yellow-white lesions throughout the posterior pole.
• The pathophysiology of APMPPE is not well understood. The underlying mechanism is most likely an obstructive vasculitis that causes choroidal non-perfusion resulting in ischemic injury to the overlying RPE.
• The exact etiology of APMPPE is also not well known. It is believed to be inflammatory or autoimmune in nature and possibly a hypersensitivity induced vasculitis. APMPPE has been known to occur in conjunction with a broad variety of disorders including TB exposure, sarcoidosis, mumps, Lyme disease, toxoplasmosis, vaccinations, adenovirus 5 infection, use of penicillin or erythromycin, and use of oral contraceptives or hormone replacement.
• The overall incidence and prevalence of APMPPE is unknown. There is no racial or gender predilection. The disease typically affects young healthy individuals aged 16-40, with an age range of 8 to 66.
• Clinical symptoms include a sudden painless loss of vision in one or both eyes, a central or paracentral scotoma, photopsia, floaters, metamorphopsia, photophobia, viral prodrome, headaches, transient hearing loss. The viral prodrome has been reported in approximately 1/3 of patients with symptoms including fever, cough, swollen lymph glands, nausea, vomiting, malaise, and muscle or joint tenderness. Rarely patients experience neurological symptoms such as transient aphasia, numbness and weakness, or feelings of clumsiness.
V. Treatment & Management

- No treatment is necessary for APMPPE as it is a self-limiting condition. The use of corticosteroids is controversial as no evidence exists that proves they speed visual recovery or improve the outcome. They are however generally used in cases where the placoid lesions are extensive and bilateral or in cases where the placoid lesions involve the macula. The dosage for oral steroid treatment starts initially at 30-60 mg/day by mouth with the tapering schedule at the discretion of the prescribing physician. As with any medication, it is important to discuss the potential side effects with the patient. A neurology consult is important if the patient is experiencing any neurological symptoms to rule out a possible but rare association known as cerebral vasculitis.

- The prognosis is favorable with 80% of patients regaining vision of 20/40 or greater. Recurrences of this condition are rare. The visual prognosis decreases when there is foveal involvement, in patients > 60 yrs, in unilateral cases, when there is a long interval (>6mo) between eye involvement and when the disease is recurrent.

VI. Conclusion – Clinical pearls, takeaway points

- The visual acuity range during the disease course can range from 20/20 to 20/200 depending on the location of the placoid lesions. The placoid lesions usually resolve in 2 to 6 weeks but visual acuity may take up to 6 months for recovery. RPE mottling remains after resolution of these lesions. Choroidal neovascularization is an uncommon complication of APMPPE but has been reported.

- A careful review of systems is critical in patients with APMPPE due to its association with a wide variety of systemic disorders.

- Photodocumentation is an excellent way to learn disease progression.

VII. References (preliminary)