An Atypical Case of Recurrent Acute Posterior Multifocal Placoid Pigment Epitheliopathy
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Abstract:
A 14-year-old Hispanic male presents with unilateral decreased vision. White retinal lesions, more numerous in the left eye, are discovered and the patient is subsequently diagnosed with acute posterior multifocal placoid pigment epitheliopathy (APMPPE).

I. Case History:
   o Patient demographics: 14-year-old Hispanic male
   o Chief complaint: Blurry central vision, OS only
   o Past ocular history: No prior injuries, surgeries, or ocular conditions
   o Past medical history: No recent illnesses or surgeries
   o Family medical history: Hypertension and heart disease
   o Medications: No oral or topical medications
   o Allergies: No known drug or environmental allergies
   o Social history: Denied smoking or alcohol use, looking forward to starting football

II. Pertinent Findings:
   o BCVA at initial visit: 20/20 OD, 20/50 OS
   o BCVA when condition recurred (week 7): 20/80-2 OD, 20/30-1
   o BCVA at most recent exam (week 13): 20/25-2 OD, 20/20-2 OS
   o Pupils: Equal and reactive to light with (-) RAPD
   o EOM: Full range of motion
   o CVF: Full to finger counting OD, OS
   o Amsler Grid: Within normal limits OD, central abnormality OS
   o IOP: 16 mmHg OD, 19 mmHg OS
   o SLE: Unremarkable, anterior chamber had no cells or flare OU
   o DFE at initial exam:
      ▪ OD: Single area of hypo-pigmentation in macula
      ▪ OS: Yellow lesions in varying stages, RPE changes and pigment mottling within the macula
      ▪ OU: No vitreous cells
   o DFE at most recent exam:
      ▪ OD: Chronic, uniform grey lesions from posterior pole into mid-periphery
      ▪ OS: Chronic, uniform grey lesions from posterior pole into periphery
      ▪ OU: No vitreous cells
Special Testing:
- Fluorescein angiography: Early hypo fluorescence followed by late hyper fluorescence of the retinal lesions.
- OCT: Within normal limits with no breaks in Bruch’s membrane or fibrosis
- Fundus photos were taken throughout the course of treatment for documentation.

III. Differential Diagnosis
- Primary: Acute posterior multifocal placoid pigment epitheliopathy, OU
- Secondary: Ampigenous choroiditis OU, ruled out via fluorescein angiography

IV. Discussion
- Acute posterior multifocal placoid pigment (APMPPE) is a rare posterior uveitis that affects the outer retina, the retinal pigmented epithelium and choriocapillaris.
- Patients complain of decreased vision that often occurs unilaterally at first with the second eye being affected later.
- APMPPE has no gender or racial predilection, but patients ranging from 20 to 50 years of age are at the highest risk.
- APMPPE is usually a self-limiting condition without recurrences, but when the fovea is affected, systemic corticosteroids may be used to hasten resolution and limit scarring.
- This case is unique in that the patient had recurrence of symptoms six weeks after starting oral prednisone, near the end of his taper.
- Other unique features of this case are the patient’s young age and lack of viral flu-like symptoms preceding the condition.

V. Treatment and Management
- Referred patient for complete blood count (CBC) and purified protein derivative (PPD) test
  - CBC within normal limits and PPD negative
- Oral prednisone treatment schedule (each bullet represents an office visit):
  - 60mg daily for one week
  - 60mg daily for two days, then reduce to 40gm daily for two weeks
  - 20mg daily for one week, 10mg daily for one week, 5mg daily for one week, 2.5mg daily for one week
  - Patient presented with decrease vision now in OD. Treatment increased to 60 mg daily for 3 days
  - 60mg daily for one week
  - 40mg daily for 4 days, 20mg for one week
  - 2.5mg every other day for one week and then stop

Bibliography:


Vianna, R. et al. The white dot syndromes. Arq Bras Oftalmol. 2007

VI. Conclusion/Clinical Pearls:

- Be aware of, and educate your patient on, possible side-effects of medications prescribed. The patient and his mother appreciated being told of possible irritability and jitteriness before treatment was initiated.

- APMPPE is one of several “white dot syndromes” including ampigenous choroiditis. It is important to be aware of the features that differentiate your diagnosis from these similar-appearing diseases.

- Diseases do not always respond to treatment the same with every patient and can have unexpected recurrences, as in this case.