Acute Posterior Multifocal Placoid Pigment Epitheliopathy Associated with Gastrointestinal Stromal Tumor and Hurthle Cell Tumor

Daniel D. Kim
Ghassan Ghorayeb

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A 50-year-old Caucasian male presented with 5 days of significant central vision changes in both eyes. About 4 days
prior to visual symptoms, he started a viral like illness with severe headaches, fevers, chills, and joint pain. He was given Tamiflu by an urgent care clinic after being diagnosed with the flu.

His vision was found to have a best corrected visual acuity of (BCVA) 20/25 OD and count fingers OS. Intraocular pressures: 14mmHg OD 15mmHg OS. Brisk pupil reactions were found with no afferent pupillary defect in both eyes. Extraocular movements were full. Anterior chamber examination showed normal cornea, iris, and lens with deep chambers and no cell/flare in both eyes. Posterior segment examination showed a clear media with no vitritis as well as normal disc and vessels. There were, in the posterior pole of both eyes, multiple yellow-white chorioretinal placoid lesions more significant on the left eye (Figure 1(a)).

Spectral domain optical coherence tomography showed the placoid lesions with disruption of the RPE, external limiting membrane, and ellipsoid zone as well as small focal points of hyperreflective material at the level of the ellipsoid zone (Figure 1(c)). Fundus autofluorescence (FAF) showed the placoid lesions to have hyperautofluorescence center with hypoautofluorescence edges (Figure 1(b)). Fluorescein angiogram showed the placoid lesions had the characteristic early blocking with late hyperfluorescent staining of edges (Figure 1(d)). Based on imaging and clinical exam, the patient was diagnosed APMPEE.

Due to the concern for cerebral vasculitis, the patient was admitted for imaging and treatment. A lab work-up showed an elevated ESR and CRP, positive IgG toxocara, and toxoplasma. IgM toxocara and toxoplasma were negative and the rest of the lab workup was negative. MRI brain, CTA head/neck, and lumbar puncture performed were found to be normal. After ruling out infectious causes, the patient was started on intravenous high dose steroids with transition to PO steroids of 1mg/kg and a planned slow taper.

After a couple of weeks of starting steroids, the patient had an incidence of bright red blood per rectum and underwent a rapid steroid taper as well as a colonoscopy. A biopsy was performed during the colonoscopy which showed a gastrointestinal stromal tumor. The lesion later excised showed on pathologic analysis a high grade gastrointestinal stromal tumor. The patient was advised that he may need adjuvant chemotherapy.

Also during work-up of the GIST, a thyroid nodule was found. Subsequent fine needle biopsy of the lesion showed atypical Hurthle cells. Genetic testing of the atypical cells showed benign characteristics with low malignant potential. The patient currently is pending excision of the thyroid lesion.

On the 3-month follow-up visit on no systemic steroids, the BCVA stabilized at 20/25 with the patient’s paracentral scotomas persisting. Imaging showed maturing of the lesion with stable size on FAF, more apparent late staining on FA, and mild improvement of the ellipsoid zone on the edges of the lesions on OCT (Figures 2(b), 2(c), and 2(d)). On the 5-month follow-up, the BCVA was 20/20 OU.

3. Discussion

To our knowledge this is first case of APMPEE diagnosed along two different tumors: a gastrointestinal stromal tumor and a Hurthle cell tumor. Further investigation will be needed over time to see if the two tumors are associated with APMPEE.

There are a couple of known associations with APMPEE: cerebral vasculitis and viral illness. Both associations

**Figure 1:** Images taken 3 days after onset of symptoms. Fundus photo, yellowish white subretinal placoid lesions (a). FAF, placoid lesions with central hypoautofluorescence with hyperautofluorescent edges (b). OCT Fovea, top: OD and bottom: OS. RPE and ellipsoid zone attenuation of placoid lesions. Subretinal hyperreflective material (c). FA, left: early phase OS. Central and right: late phase OD and OS. Early blocking with late staining (d).
have been well described in the literature. Case et al. note a review of literature of 23 case reports with ischemic and/or hemorrhagic stroke secondary to cerebral vasculitis [5, 6]. Viral illness also has been described [7]. More rarely Wegener's granulomatosis [9] and other infectious causes such as borreliosis [11] have been linked to APM-PEE.

There is also a case report of a renal cell carcinoma associated with APMPEE. It was speculated that circulating immune complexes from the renal cell carcinoma may have been the cause of APMPEE [10]. For our patient, both solid tumors are not known to secrete circulating factors or proteins. The tumors do have specific genetic components that affect treatments such as imatinib for GIST [12]. We certainly acknowledge that this association may be coincidental, yet the near simultaneous presentation raises the suspicion as to if and how these tumors are related to APMPEE.

This case, much like other APMPEE diagnoses, has a history of viral prodrome, but both tumors were present and certainly affected the patient’s immunological state. This unique case gives incentive to consider other causes or associations including neoplasm for APMPEE.

Consent

Consent has been obtained.

Conflicts of Interest

Daniel D. Kim and Ghassan Ghorayeb declare that there are no conflicts of interest regarding the publication of this paper.

References


