Title: Acute Posterior Multifocal Placoid Pigment Epitheliopathy: A case of marked asymmetric retinal lesions and rapid resolution

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Abstract: This is a case report of a 29 year old female with APMPPE OD. It is unusual due to marked asymmetric presentation, unilateral symptomatology and rapid resolution.

I. Case History

Twenty nine year old black female presents to Woodhull Medical and Mental Health Center complaining of significant pain OD, non-specific decrease in vision, and an associated headache over the preceding week. The patient's ocular history is significant for severe dry eye OU, for which the patient was treated at New York Eye and Ear Infirmary. The patient reports having obtained a thorough rheumatology work up at the time, though no underlying inflammatory etiology was found. Medical health is significant for anemia and current medications include ferrous sulfate and Loestrin.

II. Pertinent Findings

Best corrected distance visual acuities were 20/200 OD, PH NI, and 20/25- OS. Pupils were observed to be equal, round and reactive to light and examination of the anterior segment structures were unremarkable OU. Intraocular pressures of 10 mm Hg OD and 11 mm Hg OS were measured. A 50% red desaturation was noted in the right eye. Fundus examination OD revealed diffuse edema of the retinal nerve fiber layer and multiple acute yellow-white placoid subretinal lesions of the posterior pole, the retinal periphery was flat and intact 360
degrees. No change in clinical appearance of the posterior pole and retinal periphery was noted OS. The results of an Optical Coherence Tomography examination revealed large subretinal and intraretinal cystic spaces at the fovea OD (total foveal thickness: 640 microns; total macular volume: 13.07 mm$^3$), no intraretinal or subretinal elevations were noted at the fovea OS (total foveal thickness: 146 microns; total macular volume: 6.45 mm$^3$).

Fluorescein angiography performed the same day indicated early hypofluorescense of the lesions followed by late stage hyperfluorescence OD and pinpoint areas of late hyperfluorescence despite no retinal placoid lesions found OS. Subsequent visual field testing was suggestive of a central depression OD and full OS. Obtained blood work was unremarkable.

III. Differential diagnosis

A leading diagnosis of Acute Posterior Multifocal Placoid Pigment Epitheliopathy (APMPPE) OD was made upon clinical presentation and confirmed with fluorescein angiography. Other differential diagnosis included: Multiple Evanescent White Dot Syndrome, Multifocal Choroiditis, Punctate Inner Choroidopathy, Serpiginous Choroiditis, Birdshot Chorioretinopathy and Lymphoma.

IV. Diagnosis and discussion

Acute Posterior Multifocal Placoid Pigment Epitheliopathy was first described by Gass in 1968 as an acquired, self-limiting inflammatory retinal disorder primarily affecting the retinal pigment epithelium of otherwise healthy young adults. Classified as a White Dot Syndrome, it involves: (1) a rapid loss of central vision secondary to multiple sub-retinal placoid lesions within the posterior pole; (2) rapid resolution of these lesions with permanent alteration in the retinal pigment epithelium and minimal damage to the adjacent retina and choroid; (3) significant visual recovery which continues for several weeks or months after clinical resolution of the acute placoid lesions.

APMPPE is often bilateral but may be asymmetric, has no sexual predilection, mostly found to affect whites and tends to occur in the third
decade of life. Patients often present complaining of acute decrease in visual acuity, headache and central or paracentral scotoma. Prodromal viral symptoms may or may not be present prior to acute onset. Though mainly ocular in nature and rarely found to have associated systemic manifestations, cases of APMPPE and central nervous system involvement such as hemiparesis, ataxia and cerebral vasculitis have been reported.

Though the exact pathogenesis of APMPPE remains undetermined, it has been suggested to be a result of an obstructive vasculitis at the level of the choroid leading to hypoperfusion and ischemia of the choroidal lobules and the overlying retinal pigment epithelium and photoreceptors. Resolution of the inflamed choroid and retina later results in retinal pigment epithelial atrophy and focal areas of hyperpigmentation. Diagnosis is made based on clinical presentation and fundus examination; however, fluorescein and indocyanine green angiography as well as spectral-domain optical coherence tomography can aid in the diagnosis. Fluorescein angiography is characterized by early hypofluorescence followed by late hyperfluorescence and indocyanine green angiography indicates early and late hypofluorescence corresponding to the multifocal placoid lesions. SD-OCT in resolved cases of APMPPE is indicative of a photoreceptor atrophy and disruption of the inner and outer segment junction. Electroretinograms in patients with APMPPE are unremarkable.

The disorder is self-limiting and while visual acuity can be poor as counting fingers on or following presentation, visual recovery is often 20/40 or better and generally regained within 3-4 weeks of acute onset.

V. Treatment, management

The patient was started on Pred Forte 1% ophthalmic solution QID OD at time of diagnosis. Although fluorescein findings were classic for APMPPE, an ERG was ordered given unilateral clinical presentation. The patient was instructed to return in one week for an Avastin/Decadron combination intravitreal injection OD. Despite a progressive worsening in best corrected visual acuity at one week followup: 20/400 OD, stable OS, planned treatment was withheld due to
a marked improvement in clinical appearance OD. Repeated Optical Coherence Tomography examination at one week indicated a significant decrease in subretinal and intraretinal cystic spaces at the fovea in the right eye (total foveal thickness: 205 microns; total macular volume: 6.61 mm$^3$), stable OS. Early hypofluorescence and late hyperfluorescence were noted on repeated fluorescein angiography OU; however, markedly improved from one week prior. The patient was instructed to continue Pred Forte 1% QID. At last followup best corrected visual acuities were 20/30 OD, PH NI, and 20/30 OS. At the time of abstract submission, the patient is scheduled for followup in 1 week and ERG results are still pending.

Multiple routes of corticosteroids administration have been used in the treatment of APMPPE and any associated systemic manifestations; however, there is no evidence that the visual outcome is affected. Additional studies are required at this time so to evaluate the dosage, duration and benefit of steroid treatment. No therapy is indicated in most cases and routine monitoring on a one to two week basis is the current recommended mode of treatment.

VI. Conclusion

This case is unusual in that evidence of retinal lesions and APMPPE symptomalogy were found on clinical presentation in the right eye only, despite nearly all reported cases of APMPPE indicating a bilateral onset or involvement of the second eye within a few days of the first. However true to its self-limiting nature, resolution of retinal lesions occurred within 2 weeks of onset with improvement in visual acuity occurring shortly thereafter. The patient continues to be followed for signs of presentation in the other eye.

References:

Cheung CM, Yeo IY, Koh A. Photoreceptor changes in acute and resolved acute posterior multifocal placoid pigment epitheliopathy


