Letters

RESEARCH LETTER

Acute Posterior Multifocal Placoid Pigment Epitheliopathy as a Choroidopathy: What We Learned From Adaptive Optics Imaging

Acute posterior multifocal placoid pigment epitheliopathy (APMPPE) is a posterior uveitis included in the spectrum of white dot syndromes. It usually affects young healthy individuals who develop photopsias, paracentral scotomas, and decreased vision. Clinically, APMPPE presents with multiple, bilateral, gray-white, placoid lesions that evolve over several weeks, leaving foci of hypopigmentation and pigment clumping. We describe an analysis of multimodal imaging of APMPPE using both ultra-wide-field imaging (200Tx; Optos) and adaptive optics (AO) imaging (rtx1; Imagine Eyes).

Report of a Case | A 25-year-old white man with bilateral blurred vision for several weeks was diagnosed as having APMPPE in both eyes based on characteristic funduscopic findings. Best-corrected visual acuity was 20/400 OU on presentation, improving to 20/25 OU over 16 weeks of follow-up. Multimodal imaging at baseline and subsequent visits (**Figure 1** and **Figure 2**) showed 3 types of lesions:

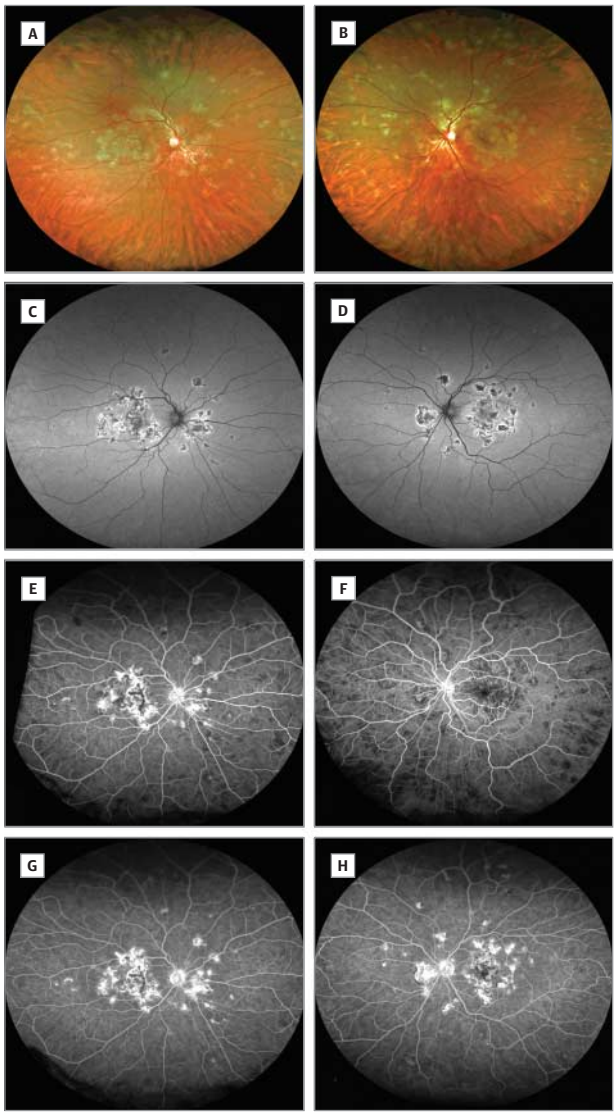
- Type 1: white on color photographs, hypofluorescent early and isofluorescent late on fluorescein angiography (FA), and isoautofluorescent on fundus autofluorescence (FAF). These lesions were mostly located anterior to the equator at baseline and disappeared over time.
- Type 2: white on color photographs, hypofluorescent early and staining late on FA, and hypoautofluorescent on FAF. These lesions became atrophic.
- Type 3: pigmented on color photographs, hypofluorescent early and late on FA, and hyperautofluorescent on FAF. These pigmented lesions were located at the margins of the placoid lesions in the left eye and occurred more centrally in the right eye. Over time, they appeared to migrate from the periphery into the center of the placoid lesions.

Numerous discrete dark spots were detected on AO imaging and analyzed through multimodal imaging. They were brownish on color photographs. Some were hyperautofluorescent on FAF. They were distributed along the choroidal vasculature on AO imaging (Figure 2).

Discussion | The primary chorioretinal layer involved in APMPPE has yet to be elucidated. Although Gass[1] originally described the placoid lesions occurring at the level of the retinal pigment epithelium (RPE), Deutman et al[2] hypothesized that acute inflammation of the choriocapillaris might be the initial insult and the RPE changes might be a subsequent manifestation. Integrated data from FAF imaging and other imaging modalities have demonstrated that the choroidal lesions on FA and indocyanine green angiography are more numerous and widespread than the RPE lesions on FAF imaging.[3] As a consequence, the RPE appears to be affected secondarily to the choroidal lesions.

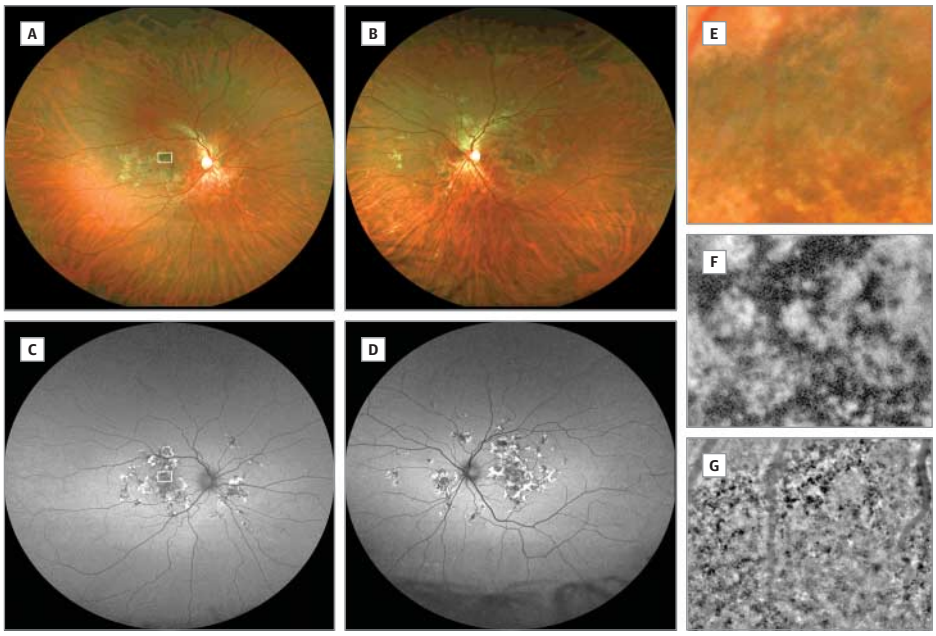
The first *P* of APMPPE stands for posterior, but our case shows numerous placoid lesions anterior to the equator (type

Figure 1. Multimodal Imaging at Presentation



Multimodal imaging at presentation, using the 200Tx ultra–wide-field retinal imaging system (Optos). Color photographs reveal numerous white-gray lesions throughout the periphery, both anterior and posterior to the equator (A and B). The lesions anterior to the equator are silent on fundus autofluorescence imaging (C and D) and hypofluorescent early (E and F) and isofluorescent late (G and H) on fluorescein angiography. The lesions that stain late on FA show mostly hypoautofluorescence on FAF. The pigmented lesions are hypofluorescent early and late on FA and show hyperautofluorescence on FAF.

Figure 2. Multimodal Imaging 4 Weeks After Presentation



Multimodal imaging 4 weeks after presentation, including adaptive optics imaging. As compared with Figure 1, note the disappearance of the peripheral lesions on color photographs (A and B). The persistent lesions appear more pigmented in both eyes (B and C). The pigmented lesions show hyperautofluorescence and the nonpigmented lesions show hypoautofluorescence on fundus autofluorescence imaging (C and D). The area in the white squares (A and C) has been analyzed with adaptive optics imaging. Color photographs show pigmented spots (E), some of which are hyperautofluorescent on fundus autofluorescence imaging (F). These spots correspond to the numerous dark spots detected on adaptive optics imaging, which are distributed along the choroidal vessels (G).

1). If these lesions were at the level of the RPE, one would expect them to be visualized on FAF imaging. However, they were completely silent on FAF imaging, suggesting that they may be located deeper than the RPE, at the level of the choriocapillaris. We distinguished 3 types of lesions based on their multimodal imaging characteristics in APMPPE. Type 1 lesions seem to be located at the level of the choriocapillaris and are transient. Type 2 and 3 lesions are at the level of the RPE and are permanent.

The pigmented lesions were distributed along the choroidal vasculature on AO imaging (Figure 2). They may correspond to pigment-laden macrophages or accumulation of pigment granules and may indicate a secondary reaction at the level of the RPE induced by an inflammatory process at the level of the choroidal vasculature. Their size and shape were not consistent with RPE cells.

Acute posterior multifocal placoid pigment epitheliopathy is characterized by prominent RPE changes, but the permanent RPE damage (type 2 and 3 lesions) may be secondary to an acute transient choroidal inflammatory process (type 1 lesions).

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Study concept and design: Mrejen, Gallego-Pinazo, Freund.
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Analysis and interpretation of data: Mrejen, Gallego-Pinazo, Freund.
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Critical revision of the manuscript for important intellectual content: All authors.
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