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## Phase-Variance Optical Coherence Tomographic Angiography Imaging of Choroidal Perfusion Changes Associated With Acute Posterior Multifocal Placoid Pigment Epitheliopathy

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Acute posterior multifocal placoid pigment epitheliopathy (APMPPE) is characterized by bilateral multiple placoid white-gray lesions of the posterior pole at the level of the retinal pigment epithelium (RPE) and/or choriocapillaris, which self-resolve with recovery of vision.<sup>1,2</sup> The etiology is unknown, but APMPPE has been associated with viral prodromes and vasculitis. The placoid lesions are thought to represent focal RPE inflammation or choriocapillaris ischemia based on fluorescein angiogram findings of early hypofluorescence and late hyperfluorescence.

Phase-variance optical coherence tomographic angiography (pv-OCTA) allows noninvasive visualization of the choroidal blood flow in 3 dimensions in vivo.<sup>3,4</sup> A custom-built OCT system (125-kHz line rate, 855-nm central wavelength, and 4.5- $\mu$ m axial resolution) was used to image choroidal perfusion over a 1.5  $\times$  1.5-mm<sup>2</sup> area (equally spaced 350 A-scans in both x- and y-axes) in acute APMPPE.

### Report of a Case

A woman in her mid to late 20s with viral prodrome presented with blurry vision in the left eye for 1 week. Visual acuity was 20/20 OD and 20/400 OS. The right eye was normal, and the left eye had normal anterior segment and vitreous, but fundus showed multiple white-gray placoid lesions posteriorly with early hypofluorescence and late hyperfluorescence on fluorescein angiogram (Figure 1). Patchy hypoperfusion of the choriocapillaris and Sattler layer was noted on pv-OCTA corresponding to focal areas of photoreceptor and RPE irregularity on OCTB-scans (Figure 2A–C). Serology test results were negative for syphilis, sarcoidosis, and tuberculosis.

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After 2 weeks, similar placoid lesions were noted in the right eye. After 6 weeks, placoid lesions resolved bilaterally to develop pigment clumping (Figure 1D) and choroidal perfusion mostly normalized on pv-OCTA. Visual acuity and photoreceptor irregularity on OCT minimally improved (Figure 2D–F). By the 6-month follow-up, visual acuity was 20/20 OD and 20/30 OS, with almost normalized photoreceptor layer on OCT (Figure 2G).

This study conforms to the World Medical Association Declaration of Helsinki<sup>5</sup> and subsequent amendments. The research protocol was approved by the University of California, Davis Office of Human Research.

## Discussion

Acute posterior multifocal placoid pigment epitheliopathy was first described by Gass in 1968,<sup>1</sup> yet the pathophysiology remains unclear because there is no histopathology of the placoid lesions.<sup>2</sup> During the acute phase, indocyanine green angiography shows hypofluorescence of the lesions without late hyperfluorescence and large choroidal vessels have been seen coursing through hypofluorescent areas on fluorescein angiogram and indocyanine green angiography, findings suggestive of choriocapillaris ischemia.<sup>6</sup>

In this case of APMPE, serial pv-OCTA visualized the transient patchy hypoperfusion of the choriocapillaris and Sattler layer associated with acute APMPE. These changes can be compared with published normal pv-OCTA choroidal perfusion maps.<sup>3,4</sup> The resolution of the choroidal hypoperfusion on pv-OCTA preceded the resolution of photoreceptor abnormality on OCT and visual recovery. Thus, the pathophysiology of APMPE likely involves a primary hypoperfusion of the inner choroid with resultant RPE and photoreceptor abnormality and vision loss. Concurrent edema or infiltration of the RPE or choroid is possible.

Perfusion images were produced by en face projection of thin slices from pv-OCTA volumetric data at 8  $\mu\text{m}$  below the Bruch membrane for choriocapillaris visualization and 8 to 16  $\mu\text{m}$  below the Bruch membrane for Sattler layer visualization. By using pv-OCTA signals overlaid on OCT intensity B-scan images, the corresponding retinal structural changes can be studied relative to the perfusion changes (Figure 2A). These focal changes in choroidal flow signal on pv-OCTA do not correlate with shadowing artifacts from overlying RPE changes. The focal lobular areas of choroidal hypoperfusion at the level of the choriocapillaris extend into the deeper Sattler layer and appear to correspond to the placoid lesions.

In conclusion, this case report used pv-OCTA to demonstrate transient inner choroidal flow changes associated with APMPE. This and similar OCTA systems can provide new insights into the pathogenesis of this condition. Future larger studies may determine whether these choroidal perfusion changes are universally observed in APMPE and affect visual prognosis.

## Acknowledgments

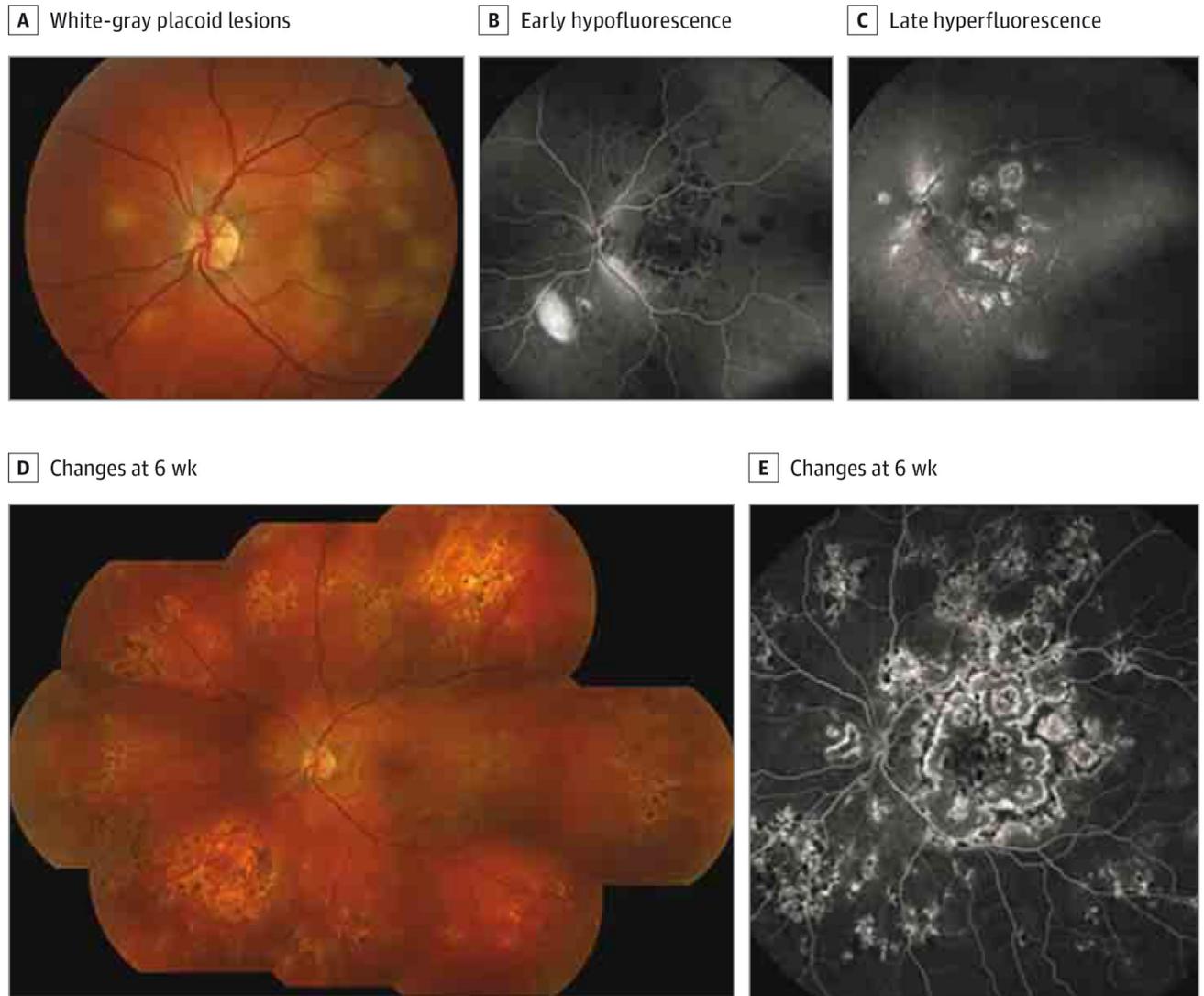
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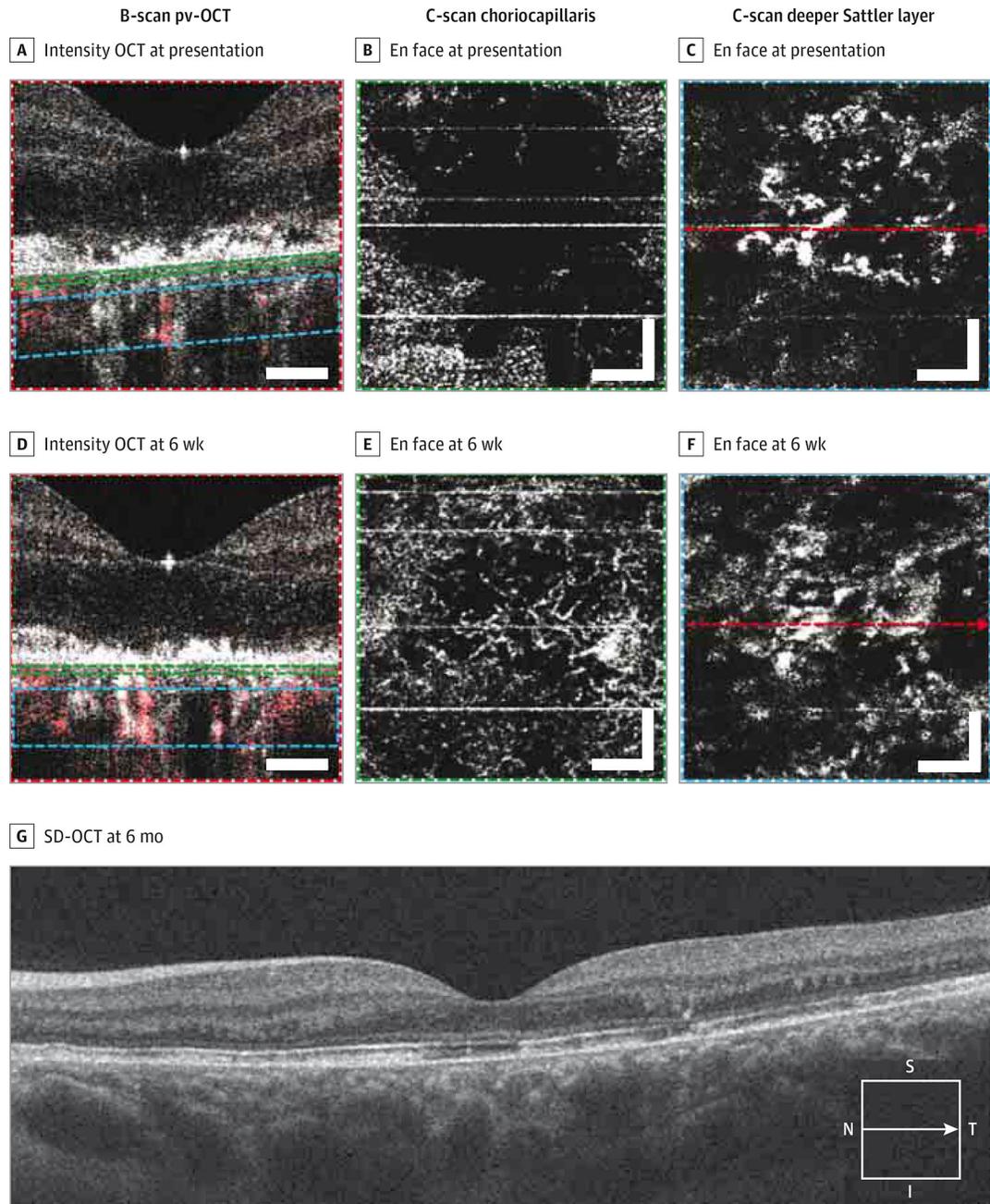
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**Figure 1. Fundus Photograph and Fluorescein Angiogram of the Left Eye**

At presentation, white-gray placoid lesions are seen (A), with early hypofluorescence (B) and late hyperfluorescence (C) on angiography; pigmentary changes developed by 6 weeks' follow-up (D and E).



**Figure 2. Left Macula Phase-Variance Optical Coherence Tomographic Angiography (pv-OCTA)** Choroidal perfusion (red, pv signal) superimposed on intensity OCT (A and D) and en face (B, C, E, and F). The images show patchy hypoperfusion of the choriocapillaris (B, green) and Sattler layer (C, blue) at presentation, resolving at 6 weeks' follow-up (D–F). Image sizes are 1.5 mm × 1.5 mm centered in the fovea; scale bar, 500 μm). G, Spectral-domain OCT (SD-OCT) at 6 months' follow-up. I indicates inferior; N, nasal; S, superior; and T, temporal.