Photoreceptor Changes in Acute and Resolved Acute Posterior Multifocal Placoid Pigment Epitheliopathy Documented by Spectral-Domain Optical Coherence Tomography

Acute posterior multifocal placoid pigment epitheliopathy (APMPPE) is generally believed to be a self-limiting inflammatory disorder affecting the retinal pigment epithelium (RPE). Vision usually recovers within weeks. The exact cause and site of the pathologic abnormality are not confirmed. We describe spectral-domain (SD) optical coherence tomography (OCT) changes during acute and resolved phases, which support initial inflammation and swelling followed by progressive degenerative changes affecting the photoreceptor layers and outer retina.

Report of a Case. A 21-year-old man had headache and blurred vision (visual acuity was counting fingers OD, 20/30 OS) for 3 days. Anterior and vitreous chambers were quiet. Funduscopy showed multifocal yellow placoid lesions in the posterior pole bilaterally (Figure 1A). These were hypofluorescent early and stained late on fluorescein angiography, and they were hypofluorescent throughout on indocyanine green angiography (Figure 1B). Magnetic resonance imaging of the brain showed no abnormalities. Three pulses of intravenous methylprednisolone sodium succinate, 500 mg each, were given. New lesions were observed in the left eye as initial lesions began to resolve in the right eye. At 5 months, visual acuity was 20/20 OU. Funduscopy showed that the cream-
colored lesions had resolved, leaving a diffuse area of pigmentary change (Figure 1C).

Spectral-domain OCT (Spectralis HRA; Heidelberg Engineering, Heidelberg, Germany) at the initial visit showed increased signal throughout the outer nuclear layer. The represented RPE was intact, but the 2 hyperreflective bands above (believed to represent the photoreceptor inner and outer segment junction and the Verhoeff membrane) were disrupted. Figure 2A shows a cut through the fovea (in the right eye) with corresponding 3-dimensional reconstruction showing the area of cream-colored lesions. Intervening areas of normal retina showed preservation of normal tomographic architecture on SD-OCT.

The SD-OCT imaging was performed during the resolved phase (3-dimensional OCT; Topcon Corp, Tokyo, Japan), which showed outer nuclear layer thinning. The lines representing the inner and outer segment junction and the Verhoeff membrane remained ill defined (Figure 2B and C).

Comment. The cause of APMPPE is believed to be either ischemic or inflammatory. The typical findings on fluorescein angiography have been attributed to masking by the overlying cream-colored patches at the RPE and outer retina level. Some investigators believe that the changes on indocyanine green angiography are also due to masking, although others have proposed choriocapillaris occlusion as the cause.1-3

Time-domain OCT studies have shown nonspecific hyperreflectivity in the outer retinal layers.1-3 Clouding of the RPE cell cytoplasm and/or an increase in cellularity due to inflammation have been proposed as the underlying cause. Anterior displacement of the neuroretina and outer reflective band have been observed and are proposed to be due to chorioiditis.1 However, details of the actual layers within the outer retina are limited by the resolution of time-domain OCT (10 µm). One study using a research tool, ultra–high-resolution OCT (3-µm resolution), described outer nuclear layer backscattering during the acute phase followed by photoreceptor atrophy.2 We report here changes in the acute and resolved phases of APMPPE studied with commercially available SD-OCT (5-µm resolution), clearly showing photoreceptor layer disruption during the acute phase.

While APMPPE often has a relatively benign prognosis, a recent series found incomplete visual recovery in a large proportion of patients.6 In our patient, despite visual acuity returning to 20/20, SD-OCT clearly showed photoreceptor atrophy and lack of reconstitution of the 2 hyperreflective bands representing the inner and outer segment junction and the Verhoeff membrane. While the border between affected and unaffected areas appeared well demarcated during the acute phase, pigmentary and atrophic changes seen in the resolved phases were much more diffuse on both funduscopy and SD-OCT. In particular, disruption of the hyperreflective bands representing the inner and outer segment junction and the Verhoeff membrane was seen in areas that initially appeared normal tomographically during the acute phase (Figure 2B and C). We hypothesize that in addition to the inflammatory component, secondary progressive degenerative changes of the photoreceptor and RPE may play a role in tissue destruction, resulting in the commonly recognized pigmentary changes in resolved APMPPE.

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Anterior Chamber Angle Assessment

Gonioscopy remains the gold-standard method for evaluation of the anterior chamber angle. Different imaging devices have been developed to study angle anatomy, including ultrasound biomicroscopy, slitlamp-adapted optical coherence tomography (SL-OCT), and Scheimpflug photography. In eyes with angle closure, indentation gonioscopy is necessary to determine the underlying pathophysiology and to differentiate between appositional closure and peripheral anterior synchiae. A technique to perform indentation gonioscopy with ultrasound biomicroscopy using a small eyecup has been previously reported. Although SL-OCT has been used to evaluate eyes with angle closure, its clinical applicability is usually limited to measurements of angle parameters. We investigated the use of an indentation SL-OCT technique to assess the angle configuration and underlying mechanisms in eyes with gonioscopically determined angle closure. The study was approved by the institutional review board of the New York Eye and Ear Infirmary.

Report of Cases. Case 1. A 48-year-old Asian man with normal intraocular pressures and no contributory history was referred for evaluation of a narrow anterior chamber angle. Indentation gonioscopy revealed appositional angle closure without peripheral anterior synchiae. Imaging by SL-OCT (1310-nm diode laser; Heidelberg Engineering, Heidelberg, Germany) under dark room conditions demonstrated a convex iris profile and iridotrabecular contact (Figure 1A). During gentle indentation of the central cornea with a scleral depressor under topical anesthesia, the iris assumed a concave profile and the angle recess opened (Figure 1B), consistent with plateau-iris configuration. Case 2. A 64-year-old white man with chronic angle-closure glaucoma, patent laser iridotomies, and intraocular pressures in the teens who was receiving 2 glaucoma medications was noted to have peripheral anterior synchiae during indentation gonioscopy. Examination by SL-OCT showed a convex iris profile along the anterior lens capsule associated with iridotrabeular contact (Figure 1C). Although the iris moved posteriorly and assumed a concave profile during indentation, the iridotrabeular contact persisted (Figure 1D), consistent with the presence of peripheral anterior synchiae.

Comment. Slitlamp-adapted optical coherence tomography is a rapid, noncontact test that images the entire anterior segment in less than 1 second. Performed in the sitting position, it is particularly suitable for in vivo imaging of anterior segment structures, which can be distinguished based on their varying optical characteristics (optical axial resolution <25 µm; lateral resolution 20-100 µm). These features distinguish it from ultrasound biomicroscopy technology. Because the indentation process affects the quality of the central im-