Long-term Follow-up of Outer Retinal Tubulation Documented by Eye-Tracked and En Face Spectral-Domain Optical Coherence Tomography

Outer retinal tubulation (ORT) is a rearrangement of the photoreceptor layer in response to retinal injury.1,2 Seen clinically with spectral-domain optical coherence tomography (SD-OCT) and confirmed on histopathologic sections, these rosette-like structures occur in a variety of retinal disorders characterized by photoreceptor disruption.3,4 On SD-OCT, ORT appears as round or ovoid hyporeflective structures with hyporeflective margins. The margins are believed to represent the inner segment–outer segment junction of the photoreceptor cells or the ellipsoid portion of the photoreceptor inner segment.5 These tubules often contain hyperreflective material thought to represent deranged photoreceptor outer segments.1,2 A branching pattern of the tubes may be detected with curved en face SD-OCT. Owing to a cystic-like appearance on OCT B-scans, ORT may be confused with cystoid macular edema related to leakage from choroidal neovascularization or other retinal diseases. We describe a patient who was treated with anti–vascular endothelial growth factor therapy for neovascular age-related macular degeneration (AMD) during a 6-year follow-up period.

Report of a Case. A 76-year-old woman visited for routine follow-up of neovascular AMD. She had previously received 1 treatment of combined verteporfin photodynamic therapy and intravitreal triamcinolone acetonide (4.0 mg/0.1 mL) in September 2005 and then received 35 intravitreal injections of ranibizumab (0.5 mg/0.05 mL) in her left eye approximately every 7 weeks from October 2005 through February 2012. Visual acuity was 20/400 at the initial visit, improved to 20/70 after starting...

Figure. Long-term follow-up of outer retinal tubulation (ORT). A, Color photograph and curved en face spectral-domain optical coherence tomographic image obtained in January 2009 showing the branching ORT structures in the left eye of a patient receiving intravitreal ranibizumab for neovascular age-related macular degeneration. B-E, Eye-tracked spectral-domain optical coherence tomographic B-scan images obtained in January 2009 showing ORT structures in various cuts through the central macula overlying quiescent choroidal neovascularization. F, Color photograph and curved en face spectral-domain optical coherence tomographic image obtained in February 2012 showing a small decrease in the size of the ORT structures. G-J, Spectral-domain optical coherence tomographic B-scan images obtained in February 2012, eye-tracked to the same scan lines as in B-E, showing a decrease in the size of the ORT structures.
ranibizumab therapy, and remained stable during the entire follow-up period. Outer retinal tubulation was first documented with eye-tracked and curved en face SD-OCT in January 2009 (Figure, A-E). The patient then underwent SD-OCT evaluations at each visit, most recently in February 2012 (Figure, F-J).

Comment. With the widespread adoption of SD-OCT in diagnosing and monitoring retinal disease, ORT has become a more commonly recognized occurrence in eyes with focal disruptions of the outer retina related to multiple diagnoses.1 These structures appear to represent rearrangement of the photoreceptor layer in response to injury, in which surviving photoreceptors form new lateral connections with neighboring cells.1,4 Most commonly, ORT is observed in eyes with choroidal neovascularization due to diagnoses such as neovascular AMD, pseudoxanthoma elasticum, multifocal choroiditis, and central serous chorioretinopathy, but it has also been described in nonneovascular disorders such as AMD with geographic atrophy, retinal detachment, Bietti crystalline retinopathy, and retinitis pigmentosa.1,5 In eyes undergoing treatment with intravitreal anti–vascular endothelial growth factor, ORT is typically found in areas in which, prior to treatment, there had been substantial intraretinal fluid that presumably damaged the outer retinal architecture.1

This case illustrates the relative stability of ORT during a multiyear follow-up period. Eye-tracked and curved en face SD-OCT was helpful in documenting this stability. We have observed similar stability in many eyes with ORT, most commonly in eyes receiving long-term intravitreal anti–vascular endothelial growth factor therapy for neovascular AMD. As described previously, the volume of presumed fluid within the ORT structures may transiently fluctuate in response to intravitreal anti–vascular endothelial growth factor, but the number and distribution of the structures typically remain constant.1 This particular case illustrates a gradual but slow decrease in the size of the ORT structures, presumably due to progressive photoreceptor atrophy. The stability of ORT during years of follow-up further supports the concept that these structures themselves are not a sign of ongoing neovascular activity. Awareness of ORT is important so that its presence is not mistaken for a sign of leakage, potentially leading to unnecessary treatment.1,2

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Ophthalmologic Diagnosis of Exacerbation of Idiopathic Pulmonary Arterial Hypertension

Idiopathic pulmonary arterial hypertension (IPAH; formerly known as primary pulmonary hypertension) is a rare condition usually affecting young to middle-aged women in whom idiopathic obliteration of pulmonary arteries results in increased pulmonary artery pressure and pulmonary vascular resistance, leading to right heart failure and subsequent elevation in systemic venous pressure.1 Ocular abnormalities have rarely been reported with IPAH.2,3 Herein, we illustrate a case in which ocular findings were the initial manifestation of clinically significant deterioration in a patient with IPAH.

Report of a Case. A 28-year-old woman had blurred vision and metamorphopsia for 5 days, affecting the right eye more than the left. Her ocular history was unremarkable. Her medical history was negative for diabetes and hypertension and was significant for IPAH diagnosed 3 years prior and treated with continuous intravenous infusion of epoprostenol via an indwelling catheter and a portable infusion pump.

On examination, best-corrected visual acuity was 20/32 ± 2 OD and 20/20 – 2 OS. On anterior segment examination, bilateral dilated episcleral vessels were noted and intraocular pressures were normal (Figure 1A). Fundus examination revealed peripapillary myelinated nerve fibers in the right eye and rare, small intraretinal hemorrhages in the temporal periphery bilaterally (Figure 1B and C). Fluorescein angiography showed normal choroidal and retinal perfusion, scattered microaneurysms, and areas of mild capillary leakage in the temporal periphery in both eyes (Figure 1D). Spectral-domain optical coherence tomography (Heidelberg Engineering) revealed serous macular detachment in the right eye greater than in the left (Figure 2A). B-scan ultrasonography did not show choroidal thickening.

Given the recent onset of ocular symptoms and findings that were consistent with elevated systemic venous