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 has 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 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 ongoing neovascular activity.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 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
pressure indicating worsening of IPAH, a right heart catheterization was performed and revealed that mean pulmonary artery pressure had increased by 27% compared with the previous measurement 9 months earlier (76 vs 60 mm Hg, respectively; reference range, 12-20 mm Hg). The right atrium and right ventricle pressures were also increased by 20% and 33%, respectively. The patient was diagnosed as having a severe exacerbation of IPAH and right heart failure, and she was admitted for aggressive treatment of fluid overload and IPAH.

Seven months later, her visual symptoms had resolved and visual acuity was 20/20 – 1 OD and 20/20 OS. She had improved episcleral vessels and fewer intraretinal hemorrhages. Spectral-domain optical coherence tomography showed complete resolution of the serous macular detachments (Figure 2B).

Comment. We report an unusual case of bilateral retinal microvascular abnormalities and serous macular detachment as the manifesting signs of exacerbation of IPAH. Idiopathic pulmonary arterial hypertension is rarely associated with ocular manifestations including central retinal vein occlusion, open-angle glaucoma, choroidal effusion, exudative retinal detachment, serous macular detachment, and diffuse macular edema. These manifestations are related to elevated episcleral, retinal, and choroidal venous pressure secondary to elevated systemic venous pressure.

Peripheral microaneurysms and mild capillary leakage with normal venous filling on fluorescein angiography have not been previously described in patients with IPAH. We believe that these abnormalities are precursors of central retinal vein occlusion and are secondary to increased retinal venous pressure causing chronic focal venous stasis, capillary damage, and focal breakdown of the blood-retina barrier.

To our knowledge, this is the first report of ocular findings as the initial manifestation of clinically significant exacerbation of IPAH warranting aggressive systemic treatment. Recognition of these subtle findings in our patient led to early intervention and treatment of severe IPAH before the development of more serious sequelae. Ocular manifestations should be recognized by clinicians, and systemic medical evaluation should be considered to prevent life- and vision-threatening complications.

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Figure 1. Clinical photograph, fundus photographs, and fluorescein angiogram. A, External photograph demonstrating bilateral dilated episcleral vessels. B, Montage fundus photograph of the right eye showing rare, subtle, scattered, temporal, peripheral, intraretinal hemorrhages (arrows). C, Montage fundus photograph of the left eye showing a temporal, peripheral, intraretinal hemorrhage (arrow). D, Fluorescein angiogram of the right eye showing mild vessel tortuosity, microaneurysms (arrows), and areas of capillary leakage (arrowhead) in the temporal periphery.
Ophthalmic Manifestations of Human Immunodeficiency Virus Infection in the Era of Highly Active Antiretroviral Therapy

Before the advent of highly active antiretroviral therapy (HAART), the rate of ophthalmic manifestations of human immunodeficiency virus (HIV) infection was very high. The lifetime risk of cytomegalovirus (CMV) retinitis, the most notorious of these complications, was once estimated at 30% in individuals with AIDS. In addition, before the widespread use of HAART in 1996, the 10-year cumulative incidence of CMV retinitis among HIV-positive patients at the San Francisco General Hospital, a large, urban, tertiary care medical center with a comprehensive HIV treatment center, was found to be 77.7%.2

In the current era of HAART, however, the incidence of CMV retinitis in individuals with AIDS has decreased by 80% nationwide, with an estimated incidence of 5.6/100 patient-years.1,3 Following the widespread introduction of HAART, the number of new cases of CMV retinitis in patients with AIDS decreased from 46 in 1994 to...