Spectral-Domain Optical Coherence Tomography of White Dot Fovea

White dot fovea is thought to be a benign condition and was originally recognized in 1997 by Yokotsuka and associates. It is characterized by the appearance of multiple tiny, white dots on the surface of the foveola that typically are arranged in a ringlike pattern at the foveal margin; the appearance can simulate a macular hole. In that early report, nearly all (28 of 30) cases described were bilateral, and all patients were Japanese. Fekrat and Humayun also identified the same condition in an African American patient with an asymptomatic, single, ringlike, white macular lesion in the right eye.

To our knowledge, white dot fovea has not been described using optical coherence tomography (OCT). Herein, we present 3 patients with asymptomatic findings in both maculae identical to those presented by Yokotsuka and associates and Fekrat and Humayun and show spectral-domain OCT (SD-OCT) images through the foveal abnormalities.

Report of Cases. Case 1. A 31-year-old Hispanic man presented with progressive vision loss in the right eye. He had no ocular history or other medical problems and did not take any medications. He was found to have a macula-off rhegmatogenous retinal detachment, which was repaired with a combination scleral buckle and vitrectomy surgery. One month after surgery, the patient was noted to have a macular hole in the same eye. A moderate posterior subcapsular cataract was present and visual acuity was 20/200 in that eye. Fundus examination of the right eye revealed granular, whitish, crystalline opacities in the perifoveal region (Figure 1A). In the asymptomatic left eye, visual acuity was 20/20, and similar granular opacities were present (Figure 1B). Spectral-domain OCT revealed a small full-thickness macular hole in the right eye. The left eye had a posterior vitreous detachment (PVD) and a normal foveal contour. In both eyes, there was a ring of perifoveal hyporeflective opacities located within the inner retina (Figure 2).

Case 2. A 43-year-old African American woman was referred to our clinic by her optometrist for bilateral macular lesions found during routine eye examination. She was asymptomatic and had a best-corrected visual acuity of 20/20 OU. She had no medical or ocular history and was taking no medications. On fundus examination, there were bilateral white, granular opacities located in the fovea (Figure 1C and D). Spectral-domain OCT revealed hyperreflective opacities located in the inner aspect of the fovea, with an otherwise normal retinal appearance and contour in both eyes. A partial PVD with vitreofoveal attachment was evident on OCT of the left eye (Figure 3).

Case 3. A 71-year-old African American woman was referred to our

Figure 1. Color fundus photographs of the maculae of case 1 and case 2. A, Close-up color fundus photograph of the right (OD) macula of case 1. A posterior subcapsular cataract is causing the media to be hazy. B, Close-up color fundus photograph of the left (OS) macula of case 1. A ring of white perifoveal granular material is visible. C, Close-up color fundus photograph of the right (OD) macula of case 2. A ring of white perifoveal granular material is evident. D, Close-up color fundus photographs of the right and left macula of case 2. In both eyes, fine whitish granules are evident throughout the fovea, with a higher-density “ring” formed by the granules at the peripheral edge of the fovea.
clinic with a diagnosis of early bilateral age-related macular degeneration. She described a few weeks’ history of mild blurred vision in the left eye. She had a history of chronic obstructive pulmonary disease and hypertension and was taking ramipril, simvastatin, inhaled ipratropium bromide, and inhaled fluticasone propionate/salmeterol xinafoate. On examination, her visual acuity was 20/25 OD and 20/40 OS. She had moderate nuclear sclerotic cataracts in both eyes, greater in the left. Fundus examination revealed small, white, foveal, granular opacities in both eyes (not shown). There were no signs of age-related macular degeneration such as drusen or retinal pigment epithelial abnormalities in either eye. Spectral-domain OCT revealed hyperreflective opacities located in the inner retina of the fovea, with an otherwise normal retinal appearance and contour. A partial PVD with vitreofoveal attachment was present in both eyes (Figure 4).

Comment. White dot fovea is thought to be a benign condition that was an incidental bilateral finding in 3 of our patients, 2 during routine eye screening and 1 who developed a unilateral retinal detachment. All 3 patients in our study, as well as the other patients described in the literature, had darkly pigmented fundi, which may accentuate the visualization of this condition.1,2 It is possible that the prevalence of white dot fovea is higher than the limited reports in the literature would suggest, as it is asymptomatic and would only be detected with detailed fundus biomicroscopy.

It is unclear what the white “dots” or granules represent histopathologically. Yokotsuka and colleagues1 were able to visualize the tiny granules with scanning electron microscopy in 5 autopsy specimens from 5 eyes with white dot fovea. The granules were 5 to 20 μm in diameter and were thought to be present either on the surface or in the superficial layer of the fovea. The configuration of the granules suggested the appearance of glial cells. Using SD-OCT, we detected hyperreflective granular lesions within the superficial foveal layer, suggesting the granules are intraretinal rather than on the retinal surface.

Yokotsuka and associates1 described variability in the appearance of the white granules. In most patients in their series, as in case 1 of our series, a ring of granules around the foveal perimeter was evident. In other patients, the granules...
ules were present over the entire fovea, as in cases 2 and 3 in our series. In addition, on SD-OCT images from case 1, the lesions highly scatter and block OCT signal from penetrating to deeper retinal layers. In cases 2 and 3, conversely, the lesions are finer, are located along a thin portion of the inner fovea, and span the entire foveola.

In cases 2 and 3, there is some evidence of partial PVD with vitreofveal attachment on OCT. Although this OCT appearance occurs as a normal stage of PVD progression, it is possible that the white granules may form secondary to mild, persistent vitreofveal traction. This is speculation, however, and it remains unclear why the granular opacities form.

In summary, we describe SD-OCT findings in 3 patients with white dot fovea. In this condition, hyperreflective granular material is visualized in the inner retinal layers of the fovea both clinically as well as on OCT. Darkly pigmented fundi seem to enhance visualization of the white foveal granules. It is unknown what the granules are composed of or what structure of the retina they represent, and further studies are needed to elucidate the pathogenesis, prevalence, and potential risk associations of white dot fovea.

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Thrombophilia in Patients With Retinoblastoma Receiving Ophthalmic Artery Chemosurgery

We have previously reported on our experience of ophthalmic artery chemosurgery (OAC) for the treatment of retinoblastoma, during which heparinization is intended to reduce the risk of thromboemboli forming at the catheter contact site. After femoral artery puncture but prior to catheterization, intravenous heparin is given to reach a target activated coagulation time (ACT) of 200 to 300 seconds (or 2-3 times the baseline), usually achieved with a single bolus of 70 IU/kg. Furthermore, the femoral arterial sheath is slowly flushed with a heparinized saline solution (500 IU of heparin in 500 mL of saline). Since the procedure usually lasts less than 1 hour, no further heparin is given. Herein, we report on 3 patients with heritable thrombophilic conditions: 1 that was known and prophylactically managed prior to catheterization and 2 that were identified following adverse events related to chemosurgery.

Report of Cases. Case 1. An 11-month-old girl with unilateral retinoblastoma, Reese-Ellsworth group 5B, International Classification D, received 5 cycles of OAC. Given a history of Factor V Leiden in the maternal grandmother, the patient underwent a coagulation workup and was found to have a heterozygous prothrombin mutation. At the beginning of the procedure, it was discovered the usual dose of heparin was insufficient, and 125 IU/kg of heparin were required to reach the target ACT. Chemosurgery was performed via the orbital branch of the middle meningeal artery in 2 cycles and was balloon assisted for 3 cycles. All OAC sessions showed good ophthalmic artery filling and choroidal blush, and her treatment course was uneventful. Following adjuvant laser, cryotherapy, and plaque brachytherapy, tumor control was achieved.

Case 2. As previously reported, a 27-month-old boy with unilateral retinoblastoma, Reese-Ellsworth...