Retinal Pigment Epithelial Detachment With Disgorgement in Age-Related Macular Degeneration Observed With OCT

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ABSTRACT: This report describes a rare appearance of retinal pigment epithelial changes in a 71-year-old woman with known long-standing, non-exudative age-related macular degeneration. She presented with visual distortion in her right eye and was found to have a retinal pigment epithelial detachment (RPED) on optical coherence tomography (OCT). Over the following 8 years, sequential OCT imaging revealed an appearance and progression of a break in the existing RPED, disgorgement of material from within the RPED, and appearance of hyper-reflective spots within the inner retinal layers, suggesting pigment epithelial cell migration. Visual acuity remained stable over this period. The RPED resolved spontaneously without treatment. The patient later developed new intraretinal hemorrhage, which was treated with intravitreal bevacizumab.


INTRODUCTION

Age-related macular degeneration (AMD) is among the leading causes of vision loss in the elderly, affecting an estimated 20 to 25 million people worldwide. The non-exudative form of AMD is characterized by the formation of drusen between the retinal pigment epithelium (RPE) and Bruch’s membrane and is associated with atrophic changes in the RPE layer. Retinal pigment epithelial detachment (RPED) is a common finding in both the non-exudative and exudative forms of AMD.

The RPE is a dynamic and reactive epithelial cell layer that responds to injurious stimuli with atrophic changes, proliferation, hypertrophy, pigmentary changes and/or migration into the inner retinal layers. Optical coherence tomography (OCT) is increasingly used to assess AMD progression. Recent advances in OCT technology allow for high-resolution assessment of the retinal layers, including the RPE.

The present case report describes a novel phenomenon of an apparent intra-RPED material release into the inner retinal layers depicted by an appearance of hyper-reflective spots within the inner retina, as observed over several years using OCT in a patient with longstanding, non-exudative AMD.

CASE REPORT

A 71-year-old woman with a 20-year history of non-exudative AMD presented to the New England Eye Center, Tufts Medical Center, Boston, in February 2004, for further management of visual distortion in her right eye. On initial presentation, her best corrected visual acuity was 20/40 in her right eye and counting fingers at 2 feet in her left eye. Dilated fundus examination and fundus photography of the right eye showed drusen and RPE changes with areas of fibrosis in the macula (Figure 1A), and disciform macular scar in the left eye (Figure 1B). OCT imaging of the right eye using the Stratus OCT (Carl Zeiss Meditec, Dublin, CA) showed a mild fibrotic pigment epithelium detachment in the right eye (Figure 1C). Neither fluorescein angiography (Figure 1D) nor indocyanine green angiography of the right eye showed evidence of choroidal neovascularization.

Over the ensuing 8 years, the patient was observed through annual follow-up. She maintained a stable visual acuity of 20/40 during this time.
Sequential OCT images of the right eye, however, obtained on Stratus OCT and then Cirrus spectral-domain OCT (Carl Zeiss Meditec, Dublin, CA) showed an appearance of a break in the existing RPED, disgorgement of material from the within the RPED, and appearance of hyper-reflective spots within the inner retinal layers (Figure 2).

At a follow-up visit in 2012, 8 years after her initial presentation, she had new distortion of vision in her right eye. Her best corrected visual acuity was 20/70. Dilated fundus examination, fundus photography, and OCT imaging now revealed new intraretinal fluid and parfoveal hemorrhage with a resolution of the previous RPED (Figure 3A-B). Fluorescein angiography showed evidence of temporal choroidal neovascularization with evidence of late-phase leakage. In the setting of decreased visual acuity and these new findings, she was treated with three injections of intravitreal bevacizumab over the course of 3 months, with stabilization of her vision at 20/70. Although her vision did not improve, at the most recent follow-up visit in 2013, the hemorrhage had resolved, and OCT imaging showed a reduction in intraretinal fluid with no evidence of a pigment epithelial detachment (Figure 3C-D).

**DISCUSSION**

Here we report a novel appearance of a break in an existing RPED and an apparent intra-RPED material release with appearance of hyper-reflective spots within the inner retina, as observed on OCT, in a patient with long-standing, non-exudative AMD. These RPE changes may represent or share a relationship with the process of RPE migration in the setting of AMD. A recent investigation by Ho et al using an ultra-high-resolution spectral-domain OCT system demonstrated the migration of RPE cells into the inner retinal layers as well as co-localization of RPE cell migration with pigment clumping in patients with dry AMD. This process of migration has been found to occur in as many as 40% to 60% of patients with AMD.
The RPE changes observed in this patient were not associated with the presence of underlying drusen on OCT or evidence of RPE clumping on fundus examination. Of note, the RPED resolved spontaneously without treatment, a finding that could represent progressive atrophic changes attributed to the natural progression of AMD.

The relationship between RPE migration and visual outcome in AMD patients has not yet been clearly defined. In healthy individuals, the RPE cell layer is composed of cuboidal cells whose basement membrane forms the inner layer of Bruch’s membrane.\(^5\) As a response to an injurious process such as AMD, the pigment layer may migrate via the passage of free desquamated cells or cell clusters found within the neurosensory retina. It has been theorized that the RPE migration occurs by a process of chemotaxis mediated by inflammatory mediators such as TNF alpha, IL-1, and MCP-1.\(^3\)

In summary, this case illustrates novel RPE changes in the setting of non-exudative AMD. Such changes require further investigation to determine their natural history and their effect on visual outcome in patients with AMD.

**REFERENCES**