Case Report

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Spectral-domain optical coherence tomography finding of acute retinal pigment epitheliitis

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Abstract:

Acute retinal pigment epitheliitis (ARPE) is a rare, transient macular disorder affecting healthy young adults. We describe the morphologic appearance of the retina by spectral-domain optical coherence tomography (SD-OCT) and to evaluate both the anatomic changes and the functional visual acuity changes over time in the course of disease. A 35-year-old healthy male presented with 1-week history of sudden-onset bilateral central scotoma with blurry vision. He denied trauma, excessive sun exposure, or drug abuse history or alkyl nitrites before. The medical and ocular examinations were unremarkable. The best-corrected visual acuity (BCVA) was 20/200 (OU) at the initial visit. Slit-lamp examination result was normal. Fundus examination revealed subfoveal yellowish lesions with a halo-like pigment in both eyes. The SD-OCT imaging showed subtle disruption of the retinal pigment epithelium (RPE) and abnormal hyperreflectivity throughout the full thickness of the foveola in both eyes. Six weeks later, the BCVA improved to 20/30 (OU) without any treatment. Six months later, the BCVA observed deteriorated to 20/50 (OU). SD-OCT demonstrated ellipsoid zone and cone outer segment tips line defects at the fovea with sharply defined borders. One year later from the initial visit, the BCVA improved to 20/20 (OU), but persisted macular microhole presents on the SD-OCT. The patient was followed for 1 year without any treatment. Thereafter, we noted that in the case of poor initial visual acuity, external limiting membrane, or outer nuclear layer involvement, as determined by SD-OCT, at the baseline might need longer time for visual acuity. The natural course of ARPE may involve the demonstration of a minor outer retinal defect that is similar to a macular microhole. In ARPE, like SD-OCT findings, the location of the initial lesion is the photoreceptor outer segments. It is not just limited to the RPE.

Keywords:

Acute retinal pigment epitheliitis, inflammation, optical coherence tomography, photoreceptors, retinal pigment epithelium

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Introduction

In 1972, Krill identified acute retinal pigment epitheliitis (ARPE),[1] an uncommon, transient macular disorder that has been demonstrated to affect healthy young adults.[2-4] Typical presentations by patients are blurred vision that is painless or central scotoma.[2,4,5] Findings that are derived from the fundus examinations include fine pigment stippling in small areas that are encompassed by hypopigmented haloes (yellow-white) in the macular region.[1-5] Despite the report of bilateral cases,[5] the effects induced on most patients are unilateral. Treatment is obviated in most cases because of the self-limiting characteristic of the disease, with acceptable visual acuity being restored in 6–12 weeks.

We recorded the morphologic appearance of the retina by spectral-domain optical coherence tomography (SD-OCT) and we evaluated anatomic data as well as functional data over the disease course.

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Case Report

A 35-year-old male in a healthy state presented with a 1-week history of sudden onset bilateral central scotoma with blurred vision. He denied trauma, excessive sun exposure, or drug abuse history or alkyl nitrites inhalation history before. The medical and ocular history data were unremarkable. The best-corrected visual acuity (BCVA) obtained for him in both eyes was 20/200 at the initial visit. Slit-lamp examination showed normal anterior segment of both eyes. Fundus examination revealed subfoveal yellowish lesions with a halo-like pigment in both eyes [Figure 1a and b]. SD-OCT images showed hyperreflective lesions that stretched from the retinal pigment epithelium (RPE) layer toward the outer nuclear layer (ONL) without exudation in both eyes (OU) [Figure 1c and d]. No specific treatment was given for this patient. Ten days later, the noted ONL hyperreflectivity was gradually resolved and external limiting membrane (ELM) was

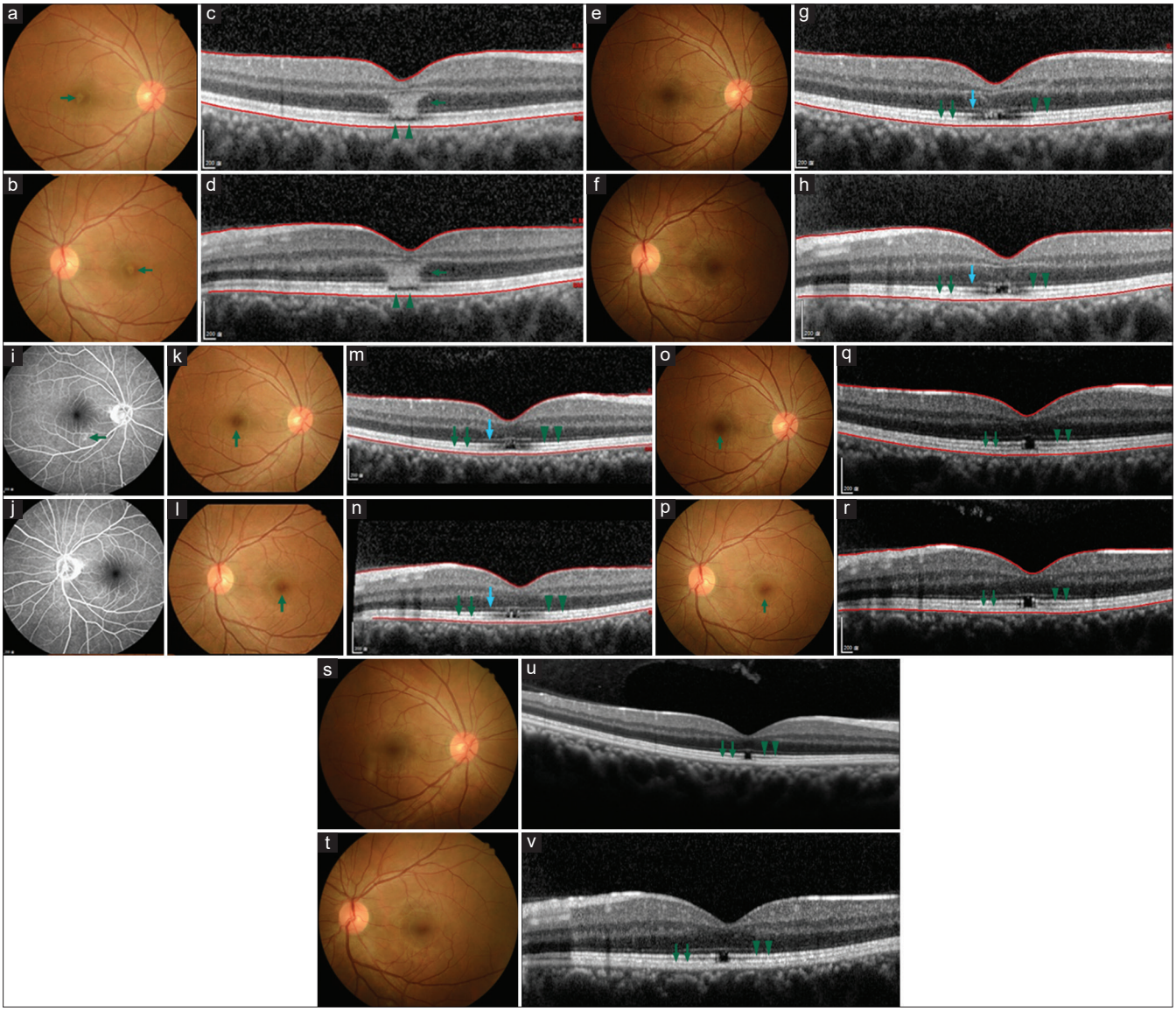

Figure 1: Color fundus at the initial visit. (a and b) The subfoveal yellowish lesion with halo-like pigment (arrow) at the foveola in both eyes. (c and d) The spectral-domain optical coherence tomography showed that the abnormal boots shape hyperreflectivity lesion extended from the inner layer of the retinal pigment epithelium (arrowheads) to the outer nuclear layer (arrow) layer without exudation in both eyes. Ten days later, the visual acuity were 20/60 (OD) and 20/200 (OS). (e and f) The hypopigmented areas slightly disappeared. (g and h) Spectral-domain optical coherence tomography showed the reconstruction of external limiting membrane (blue arrow) but still disorganized ellipsoid zone (arrowheads) and cone outer segment tips (green arrows). Six weeks later, the visual acuity improved to 20/30 (OU). (i and j) The fluorescein angiography showed mild window defect (arrow) on the macular without leakage in the right eye and unremarkable in the left eye. (k and l) Color fundus showed subtle pigmentary changes (arrow) at the fovea in both eyes. (m and n) The external limiting membrane (blue arrow) was intact, and the ellipsoid zone (arrowheads) was gradually recovering, but the cone outer segment tips (green arrows) was still disrupted. Six months later, the visual acuity deteriorated to 20/100 (OD) and 20/50 (OS). (o and p) The color fundus showed persistent subtle pigmentary changes (arrow) at the fovea in both eyes. (q and r) Spectral-domain optical coherence tomography showed the ellipsoid zone (arrowheads) and cone outer segment tips (arrows) line defects at the fovea with sharply defined borders. One year later from the initial visit, the best-corrected visual acuity improved to 20/20 (OU). (s and t) Color fundus showed slight pigmentary changes at the fovea in both eyes. (u and v) Spectral-domain optical coherence tomography showed the disruption of the ellipsoid zone (arrowheads) and cone outer segment tips (arrows) line gradually smaller than 6 months ago

reconstructed, as depicted by SD-OCT images [Figure 1g and h]. The hypopigmented areas slightly disappeared on the color fundus [Figure 1e and f]. Six weeks later, the BCVA improved to 20/30 (OU). We observed from color fundus examination minute pigmentary variations that were persistent and occurred at the fovea in both eyes [Figure 1k and l]. SD-OCT showed the ELM to be intact and the ellipsoid zone (EZ) to gradually recover; nevertheless, the cone outer segment tips line (COST) was still disrupted 6 weeks later [Figure 1m and n]. Fluorescein angiography was performed to him 6 weeks later, noting mild window defects on the macula without leakage in the right eye and noting unremarkable findings in the left eye [Figure 1i and j]. Six months later, the BCVA observed in the case mild deteriorated to 20/50 (OU). We also noted from color fundus examination minute pigmentary variations that were persistent and occurred at the fovea in both eyes [Figure 1o and p]. SD-OCT demonstrated EZ and COST line defects at the fovea with sharply defined borders [Figure 1q and r]. One year later from the initial visit, the BCVA improved to 20/20 (OU). Color fundus showed slight pigmentary changes at the fovea in both eyes [Figure 1s and t]. However, disruption persisted in the EZ and COST line which appeared on the SD-OCT but smaller than 6 months ago [Figure 1u and v].

Discussion

The pathophysiology of ARPE has yet to be elucidated; descriptions of associated clinical findings are limited to only a few case reports. Previously, electrophysiological and ophthalmic findings indicate ARPE to be an inflammatory RPE disorder.[1,6] At present, from the literature review, treatment is observed in most cases because of the self-limiting characteristic of the disease. Therefore, we decided not prescribing treatment for this patient.[4,5] Hsu *et al*. revealed time-domain OCT findings associated with ARPE, suggesting that the primary site affected may be the foveal cone photoreceptors, because of the disrupted inner-outer segment band and the relatively intact RPE–choriocapillaris band.[7] Baillif *et al*. used SD-OCT in four individuals, suggesting that the area initially involved in ARPE may be the intersection connecting the photoreceptor outer segments and the apical side of the RPE cells.[8] In 2017, Iu *et al*. speculated that in the ARPE acute phase, the ELM constituted the site for the localization of major inflammation, and that the hyperreflective lesion could be an indication of inflammatory debris accumulation or photoreceptor edema, engendering volume expansion, and ELM displacement.[9] In addition, the RPE was proposed to play a role only as a secondary response in ARPE.[10] In our case, we noted that the mechanism of the resolution of the hyperreflective lesion observed in ARPE entailed height reduction. This resolution may indicate that the greatest inflammation was localized at the outer edge of the neurosensory retina. These features suggest that the first site of inflammation may be the outer retina, as opposed to the RPE.

In general, the visual acuity was restored in 6–12 weeks. However, our SD-OCT findings of incomplete visual recovery after 6 months may be attributed to considerable baseline involvement of the retina, including the RPE inner layer, COST, EZ, ELM, and ONL. The cone photoreceptor cell bodies are located at a level similar to that of the ELM, external to the rod photoreceptors, as revealed by histological analysis.[11] Accordingly, the observed ELM as well as ONL involvement could be related to cone photoreceptor nucleus damage, a phenomenon that could be an irreversible destruction or need longer time for restoration. We might predict that it needs to be taken more time for visual recovery for patients who have exhibited poor initial visual acuity and baseline ELM or ONL involvement, as revealed by SD-OCT.

Six months after the initial consultation in our case, we noted a tiny defect in the outer retina (typically designated a "macular microhole"), as depicted by the SD-OCT images. Zambarakji *et al*. described that macular microholes are lesions in the outer retina, involving the photoreceptor segment and possibly the RPE.[12] From the literature review, eyes with macular microholes, the occurrence of macular structural abnormalities has been observed in the outer retina including EZ, COST, or/and retinal RPE disruption.[13,14] This larger cone disruption area may interpret a poorer visual acuity at that time. However, after 1-year follow-up, the macular microhole was gradually smaller than 6 months ago. It was believed that photoreceptor cell body and the inner segment remain intact in the beginning, and the cell can regenerate its outer segment and visual acuity improved after 1-year follow-up. Based on our observation, the natural course of ARPE may be in the form of a macular microhole and may ultimately be caused by damage to the outer segments of photoreceptors.

Based on the electrophysiological findings, researchers have previously hypothesized that ARPE may represent an inflammatory RPE disorder. However, with the advancement of OCT technology, we know that this disease may originate from the outer retinal areas, not just limit to RPE. Although full visual function recovery usually takes for 6–12 weeks in most cases, poor initial visual acuity and ELM or ONL involvement at the baseline were possibly to be take longer time for visual acuity recovery. ARPE is a rare macular disease; the details of its physiopathology are not known, but they warrant further research.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

The authors declare that there are no conflicts of interests of this paper.

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