Ophthalmology Practice

Macular function and morphology in acute retinal pigment epithelitis

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A 20-year-old man applied with vision loss in the left eye. Right eye examination was unremarkable. Best-corrected visual acuity (BCVA) in the left eye was 20/200. Fundus examination revealed a few yellow spots within a round-shaped macular lesion. Autofluorescence imaging showed hyperautofluorescence in the lesion. Central amplitudes in multifocal electroretinogram (mfERG) were depressed. The patient reported a rhinopharyngitis 7–10 days before the visual loss. The patient was diagnosed as acute retinal pigment epithelitis. BCVA improved gradually up to 20/20 in 4 weeks. mfERG amplitudes returned to normal. A slight pigmentary distortion was the only residual fundus finding.

Key words: Acute retinal pigment epithelitis, multifocal electroretinogram, optical coherence tomography

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Acute retinal pigment epithelitis (ARPE) causes acute and dramatic vision loss in young adults without any other health problems. As Krill and Deutmann first described this entity, it is known as Krill's disease.[1] Patients complain about vision loss with central scotoma and metamorphopsia. The main fundus finding is a macular pigment epithelial alteration. The spectrum of patient age ranges from 10 to 40. Males and females are equally affected. It is unilateral however bilateral cases have been reported.[2] It is a benign idiopathic inflammatory disease of retinal pigment epithelium that is self-limited and regress spontaneously.[3] Characteristic fundus findings are fine foveal pigment spots encircled by a lighter halo of yellow-white hypopigmentation. The lesions transmit fluorescence but don't cause leakage in fluorescein angiography (FA).[4]

In this report, we present a case of unilateral ARPE with retinal structural and electrophysiological findings.

Case Report

A 20-year-old man applied with acute vision loss in the left eye. Right eye examination was unremarkable. Best-corrected visual acuity in the left eye was 20/200. Fundus examination revealed a round-shaped hypopigmented macular lesion including a few yellow spots in the fovea [Fig. 1a]. At higher magnification [Fig. 1b], golden-colored lesions in a honeycomb pattern could be seen. FA showed a macular transmission hyperfluorescent ring in which patchy hyperfluorescent and normofluorescent areas existed [Fig. 1c]. No fluorescein leakage was observed. Fundus autofluorescence imaging showed scattered patchy hyperautofluorescent areas matching the hypopigmented fundus lesion [Fig. 1d]. Spectral-domain optical coherence tomography (SD-OCT, Spectralis OCT, Heidelberg Engineering, Heidelberg, Germany) demonstrated hyper reflective accumulations (humps) involving RPE inner band and photoreceptor outer segments (OSs). Outer nuclear layer and inner segment/OS (IS/OS) line was distorted because of the accumulations. RPE was thickened and irregular in the lesion [Fig. 1e]. Multifocal electroretinogram (mfERG, Roland-Consult RetiScan System, Wiesbaden, Germany)

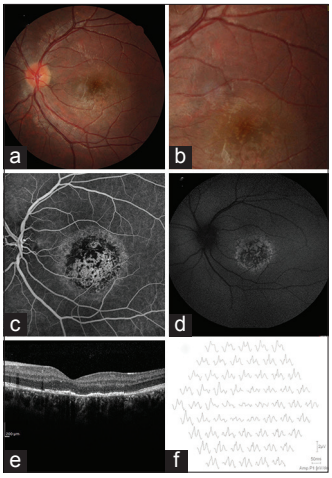

Figure 1: (a) Fundus photograph demonstrating round-shaped hypopigmented macular lesion including a few yellow spots in the fovea. (b) Higher magnification showing golden-colored lesions in a honeycomb pattern. (c) Fluorescein angiography showing a macular transmission hyperfluorescent ring in which pathcy hyperfluorecent and normofluorescent areas existed. (d) Fundus autofluorescence imaging showing scattered patchy hyperautofluorescent areas. (e) Spectral-domain optical coherence tomography image showing hyperreflective accumulations (humps) involving RPE inner band and photoreceptor outer segments. (f) Multifocal electroretinogram showing depressed central ring amplitude

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showed decreased amplitude in the central 2.3°.[5] The P1 amplitude in the central response was 50.3 nV/deg^2 [Fig. 1f]. Arden ratios in electrooculogram (EOG) were 2134 in the right eye and 1650 in the left eye. The patient reported blurred central area and metamorphopsia in Amsler Grid testing. Total error score in Farnsworth-Munsell-100 (FM-100) Hue test was 35 in the right eye and 157 in the left eye. The patient had no known systemic or ocular disease. The family history was unremarkable. The patient reported a viral (possibly) upper respiratory tract infection 7–10 days prior to the visual loss. A probable diagnosis of ARPE was suggested, and the patient was followed in next weeks.

Best-corrected visual acuity improved gradually up to 20/20 in the next 5 weeks with no treatment. A slight pigmentary distortion was the only residual fundus finding [Fig. 2a and b]. There was almost no change in FA and FOF imaging compared to the initial imaging [Fig. 2c and d]. SD-OCT showed almost entirely restored IS/OS line almost no distortion in the outer nuclear layer. Irregularities and humps decreased, but there were still some persisting hyper reflective spots in the RPE inner band and photoreceptor OSs [Fig. 2e]. The P1 amplitude of the central response increased to 114 nV/deg^2 in mfERG [Fig. 2f]. That was in the normative data range. Arden ratios were 2080 in the right eye and 2071 in the left eye [Fig. 3]. The patient reported significantly improved central vision in Amsler Grid testing when compared with the initial testing. Total error scores in the FM-100 Hue test was 42 in the right eye and 83 in the left eye.

Discussion

The diagnosis of ARPE depends on the presence of a characteristic fine pigment stippling in the macular area, at the level of the RPE, surrounded by yellow-white haloes of hypopigmentation.[1,2,4,6] No other ocular or systemic disease is associated. However, similar to our case, a preceding viral disease has often been reported.[4]

Spectral-domain optical coherence tomography findings suggest that the initial lesion in ARPE is possibly located at the level of the photoreceptor OS and RPE.[4,7] The disruption of photoreceptor IS/OS line with a wider disruption of the RPE inner band is almost typical in the reported cases.[4] The RPE inner band is considered to be related with the Verhoeff's membrane that is constituted by tight junctions of RPE cells or with basal infoldings and apical processes of RPE cells.[8] In our case, however, photoreceptor IS/OS line is discernible throughout the lesion. The main finding in our case was humps in the inner band of the RPE and photoreceptor OSs. Our findings support the speculation that ARPE is a postinflammatory response of the OS of photoreceptors and the RPE.

The exact pathogenesis of ARPE is still unclear. However, EOG findings support that it is a disease of RPE.[9] EOG explores the response of RPE to light and is a function of all RPE cells in the retina. However, the structural involvement is only seen in the macula in ARPE cases. That is probably related with the increased metabolic need of RPE cells in the macula with respect to RPE cells in the peripheral retina. RPE cells in the fovea ingest about 20–30 μm of OS/day.[7] The localized accumulation in SD-OCT images likely reflects undigested OSs of the photoreceptors. The diminished central amplitudes with progressively increasing amplitudes toward periphery

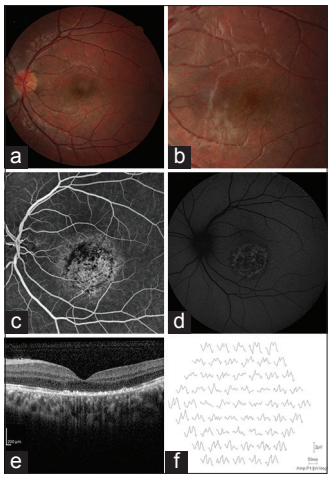

Figure 2: (a and b) Fundus photograps demonstrating residual slight pigmentary change. (c) Fluorescein angiography showing transmission hyperfluorescence in the lesion. (d) Fundus autofluorescence imaging showing hyperautofluorescent areas. (e) Spectral-domain optical coherence tomography image showing the decreased hyperreflective accumulations (f). multifocal electroretinogram showing normalized central ring amplitude

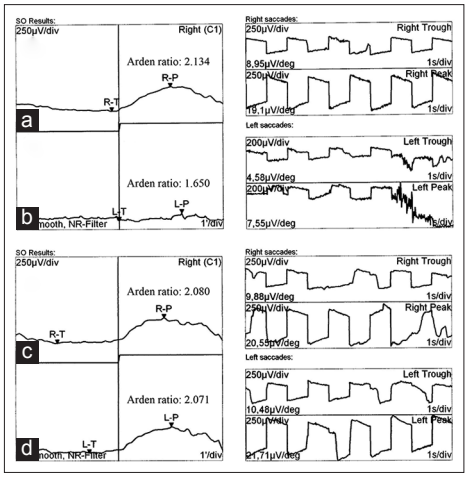

Figure 3: Electrooculogram recordings in the right (a) and left eye (b) during the initial examination and at the 5th week examination (c and d, respectively)

in mfERG confirm the functional deterioration of cone photoreceptors in the central retina. The electrophysiologic findings may mean that RPE cells in the peripheral retina can overcome the increased metabolic load although the RPE cells in the macula cannot due to the high concentration of photoreceptors. Our case showed that mfERG recording was more correlated to visual acuity than structural imaging modalities (FA, FOF, SD-OCT).

Conclusion

Spectral-domain optical coherence tomography and mfERG findings in our case support the hypothesis that RPE is possibly the initial site of involvement in ARPE. mfERG depression confirmed the functional involvement of cone photoreceptors in the disease process.

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