Acute Retinal Pigment Epitheliitis: A Case Report

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Introduction

Acute retinal pigment epithelitis (ARPE), also known as Krill’s disease, has been described by Krill and Deutman in 1972. It is a rare, self-limited macular disorder affecting healthy young adults characterized by sudden impairment of central vision which may be associated with metamorphopsia due to alteration of the macular pigment epithelium. The disease equally affects males and females, it is unilateral in 75% cases, and usually resolves spontaneously without treatment within a few months. The diagnosis of ARPE is made based on clinical suspicion, as well as fundus findings in the form of delicate darkish macular spots surrounded by a lighter halo-like zone and absence of intraocular inflammation.

Case Report

49 years old male presented to us with complaints of sudden onset blurring of vision in the right eye since 3 days. He was a diabetic on Insulin since 15 years. On examination, his vision was 6/60 with -10 D sphere-1.50 D cyl 35 º in the right eye & 6/9 with -6 D sphere -1 D cyl 160ºin the left eye, and near vision with +1.75D improved to N36 in the right eye & N8 in the left eye.

Anterior segment examination was within normal limits and the intraocular pressure recorded by applanation tonometry was 18mm Hg in both the eyes. Fundus examination by indirect biomicroscopy & slit lamp microscopy revealed a myopic disc with small greyish white lesion superotemporal to the fovea along with blunting of the foveal reflex in the right eye. Amsler chart showed a metamorphopsia in the central area in the right eye whereas left eye was normal.

He underwent fundus fluorescein angiography which was unremarkable. Optical coherence tomography (OCT) examination with spectral domain (SD OCT) (Spectralis, Heidelberg Engineering, Heidelberg, Germany) revealed thickening and undulations of the retinal pigment epithelium(RPE) with disruption of the inner segment / outer segment (IS/OS) junction and the external limiting membrane (ELM) in the subfoveal region and the area corresponding to the greyish white discoloration. Other layers of the retina appeared normal. A probable diagnosis of Acute Retinal Pigment Epithelitis (ARPE) in the right eye was made.

Though Retinal pigment epithelitis is a self limiting condition, we put the patient on a short course of systemic steroids after advising strict control of diabetes mellitus. He was reviewed after 2 weeks. On examination, his vision had improved to 6/12 N10 in the right eye. SD OCT examination revealed slight decrease in the RPE undulations, but the disruption of the IS/OS junction and the ELM still persisted.

On his review after 2 months, he was symptomatically much better. Visual acuity testing revealed improvement to 6/9 N6 in the right eye. SD OCT examination showed return of the RPE to normal morphology with reduction in the thickening & loss of the undulations and the reappearance of the IS/OS junction and the ELM.

Discussion

ARPE is an acute, rare usually idiopathic self limiting inflammatory condition affecting adults in the third to fifth decade.

White dot syndromes, especially multiple evanescent white dot syndromes (MEWDS), should be considered in the differential diagnosis of ARPE. White dot syndromes are characterized by multifocal white lesions and are accompanied by mild vitritis. Our patient was relatively young and had no signs of intraocular inflammation. The main lesions responsible for MEWDS cause damage to the photoreceptor outer segment but not the RPE, whereas ARPE is a condition primarily affecting the RPE.

In acute posterior multifocal placoid pigment epitheliopathy (APMPPE), multiple, large placoid lesions start in the posterior pole and extend to the post-equatorial fundus. ARPE lesions usually resolved without scarring, whereas the lesions in patients with APMPPE are replaced by RPE changes upon resolution.

Chittum and Kalina attempted to define ARPE by describing its angiographic characteristics. Hsu et al. first described the optical coherence tomography (OCT) findings in three cases of ARPE, performed a few days after the onset of disease. He reported abnormal increased reflectivity involving the ONL and the RPE with absence of intra-retinal, sub-retinal, or sub-RPE fluid suggesting that the initial lesion could involve the foveal photoreceptors.

Nathalie et al. reported a case of ARPE in which a definite disruption in the IS/OS junction, as well as an undulation of the RPE were found on SD-OCT which showed a decrease in both the length of the IS/OS disruption and the RPE irregularity on recovery.

Kim et al. reported a similar case of a young man with blurred vision in his left eye and a gray-white lesion in the macula on ophthalmoscopy. Spectral domain optical
coherence tomography (OCT) demonstrated a disruption in the photoreceptor inner and outer segment (IS/OS) junction and undulation of the retinal pigment epithelium (RPE) with backscattering. After three months without any treatment, visual acuity and visual field gradually normalized, with the recovery of continuity in the photoreceptor IS/OS junction, as well as decreased RPE irregularity with minimal backscattering on SD OCT.

**Conclusion**

In ARPE, visual acuity recovery is associated with the recovery of continuity in the photoreceptor IS/OS junction, as well as decreased RPE irregularity on SDOCT. On the basis of these findings, we suggest that the morphologic changes in inflammation related to ARPE seem to be transient.

**References**