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OPHTHALMOLOGY
Case Report

Microperimetry, Spectral-Domain OCT and Fundus Flourescein Angiography in the follow up of a case of Acute Retinal Pigment Epithelitis (ARPE)

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Abstract

Microperimetry, Spectral-Domain OCT (SD-OCT), Fundus Flourescein Angiography (FFA) in the follow up of a case of Acute Retinal Pigment Epithelitis (ARPE).

Purpose: To observe the changes seen on SD-OCT and other objective tests in ARPE and correlate it with subjective findings on Microperimetry.

Methods: A patient presenting to our hospital suspected clinically to have acute retinal pigment epithelitis underwent FFA, SD-OCT (Spectralis, Heidelberg, Germany) and microperimetry (MAIA), full field and multifocal electroretinography and contrast sensitivity. SD-OCT and MAIA were repeated on follow up to correlate subjective and anatomic improvement.

Results: Baseline OCT showed Retinal Pigement Epithelium (RPE) thickening and discontinuity of the macular inner and outer segment (IS-OS), abnormal increased reflectivity involving the outer nuclear layer and photoreceptors and minimal involvement of the RPE. Microperimetry revealed reduced average threshold (23 db) and abnormal macular integrity. Full field ERG showed no changes. Contrast sensitivity was reduced as compared to the other eye. Treatment resulted in resolution of the RPE lesions on SD-OCT and restoration of macular IS-OS continuity. Microperimetry showed improvement in average macular threshold (28.7 db) with better macular integrity.

Conclusion: SD-OCT shows distinctive changes that help in the diagnosis of ARPE. Restoration of macular IS-OS and resolution of RPE lesions correlate well with subjective visual improvement.

Keywords: *Microperimetry; SD-OCT; ARPE; Retinal Pigment Epithelitis; ERG*

Summary statement

This case report establishes the correlation between the subjective findings of microperimetry and the clinical investigations like SD-OCT in a case of ARPE.

Introduction

Acute retinal pigment epithelitis (ARPE), also known as Krill's disease, was first described by Krill and Deutman in 1972 as a bilateral, benign, self limited disease [1]. There are many recent reports [2-5]. That describes the Spectral Domain Optical Coherence Tomography (SD-OCT) findings in ARPE. Lesions of the RPE, inner and outer segments of photoreceptors and external limiting membrane and subretinal deposits have all been implicated in the pathology of ARPE. Poor visual acuity at baseline and involvement of the ELM or ONL on SD-OCT at baseline were associated with the incomplete recovery of visual acuity [2]. In a report of 3 cases, in the acute stage, SD-OCT demonstrated abnormal hyper reflectivity involving the photoreceptor outer segment layer and hypo reflectivity involving the associated RPE layer in all cases. In the chronic stage, SD-OCT showed decreased abnormal reflectivity. Resolution of hyper-reflective and disrupted

lesions has been correlated with an improvement in Snellen's visual acuity. Contrastingly increase in hyper-reflectivity also has been correlated with improvement in visual acuity [6]. Transmission hyperflourescence has been noted on angiography [7].

Microperimetry or fundus perimetry enables us to precisely localize the functional map on the retina enabling us to precisely correlate the anatomical and functional findings. Most previous reports have used Snellen's visual acuity to correlate anatomic findings and to our best knowledge this is the first report to correlate microperimetry and pathological changes in ARPE. One report shows correlation of anatomic and functional improvement seen on OCT and visual fields respectively [5]. We have used microperimetry in our study that gives us not only the thresholds at various points but also a macular integrity assessment that is a probability of anatomic damage. We hence aim to correlate a more accurate functional assessment with microperimetry with precise anatomic correlation as seen on SD-OCT.

Case Report

A 19 year old female presented with diminution of vision in the right eye (OD) since 1 week. The best corrected visual acuity on Snellen's chart was 6/24 in OD and 6/6 in the left eye (OS) at presentation. On examination of OD, the anterior segment was within normal limits and the intraocular pressure by applanation tonometry was 18 mm Hg. Fundus examination by indirect ophthalmoscopy & slit lamp biomicroscopy revealed fine pigment stippling & blunting of the foveal reflex. OS examination revealed no abnormalities. On enquiry, the patient gave a history of "a flu like illness" a week back that resolved with non steroidal anti-inflammatory drugs.

OD fundus fluorescein angiography showed pinpoint hypo fluorescent spots in the macular region. The SDOCT (Spectralis, Heidelberg Engineering, Heidelberg, Germany) (Figure: 1b) 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. Macular Integrity Assessment Test (MAIA-Center Vue) (Figure: 1a) showed abnormal macular integrity and average threshold but normal fixation stability. Full field ERG revealed no abnormalities. Contrast sensitivity was reduced as compared to OS. Systemic blood investigations namely haemoglobin, total and differential leukocyte count, Erythrocyte sedimentation rate and random blood sugar were unremarkable. Urine analysis showed trace protein.

A probable diagnosis of OD Acute Retinal Pigment Epithelitis (ARPE) was made. After 2 weeks she was symptomatically much better the vision in OD had improved to 6/9, N6. MAIA (Figure: 1c) showed improvement in the macular integrity. SDOCT (Figure: 1d) examination showed improvement in the RPE morphology with reduction in the thickening and the RPE irregularity. The SD OCT also showed reappearance of the IS/OS junction and the ELM.

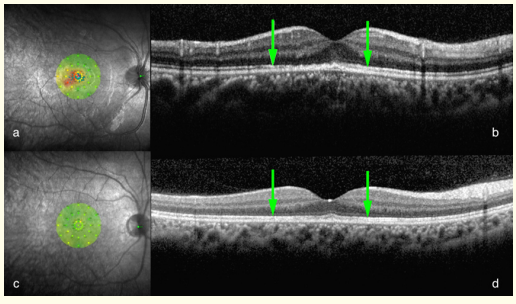

Figure 1: *a. MAIA of OD at baseline showing decreased sensitivity (red indicating decreased sensitivity; green indicating normal sensitivity).*
b. SD-OCT at baseline showing RPE hyper-reflectivity and disruption of the IS/OS junction and COST layer.
c. MAIA after 2 weeks showing improvement in thresholds.
d. SD-OCT after 2 weeks showing restoration of the IS/OS, COST layer and RPE.

Discussion

The white dot syndromes are a group of inflammatory chorio-retinopathies of unknown etiology with overlapping clinical features which have in common a unique and characteristic appearance of multiple yellow-white lesions affecting multiple layers of the retina, retinal pigment epithelium (RPE), choriocapillaris, and the choroid [8].

A viral prodrome, acute painless central visual loss in otherwise healthy young adults and discrete white spots in the posterior pole should lead us to the differential diagnosis of multiple evanescent white dot syndromes (MEWDS), acute posterior multifocal placoid pigment epitheliopathy (APMPPE) and ARPE. The anterior chamber is generally quiet, but mild vitritis and electroretinography findings can be present in MEWDS and APMPPE; a feature absent in ARPE.

SD-OCT shows characteristic changes in all 3 diseases. MEWDS mainly shows changes in the photoreceptors [9]. Disruption of IS/OS junction and intraretinal edema can be seen in APMPPE [10]. Lesions of the RPE, inner and outer segments of photoreceptors and external limiting membrane and sub-retinal deposits have all been implicated in the pathology of ARPE.

Fundus lesions in ARPE mostly heal without scarring whereas in MEWDS and APMPPE scarring may be present.

MEWDS, APMPPE and ARPE are self limiting disease mainly requiring observation. Occasionally CNVM may complicate MEWDS and APMPPE directing further treatment. APMPPE has a risk on CNS vasculitis and hence a systemic review is essential.

The self resolving nature of ARPE can be seen on both the SD-OCT and the microperimetry of our patient. Improvement of macular sensitivity thresholds to normal with simultaneous anatomic improvement on SD-OCT establishes a strong clinicopathological correlation in ARPE.

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