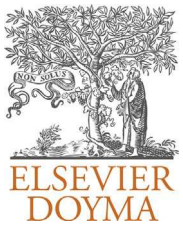
Journal of Optometry (2014) 7, 241–245



Journal of Optometry

www.journalofoptometry.org

CASE REPORT

Macular pigment optical density spatial distribution measured in a subject with oculocutaneous albinism

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Received 10 February 2014; accepted 7 March 2014

KEYWORDS
Macular pigment optical density;
Oculocutaneous albinism;
Spatial distribution

Abstract
Purpose: Previous studies of macular pigment optical density (MPOD) distribution in individuals with oculocutaneous albinism (OCA) have primarily used objective measurement techniques including fundus reflectometry and autofluorescence. We report here on a subject with OCA and their corresponding MPOD distribution assessed through heterochromatic flicker photometry (HFP).
Methods: A subject with a history of OCA presented with an ocular history including strabismus surgery of the LE with persistent amblyopia and mild, latent nystagmus. Best corrected visual acuity was 20/25- RE and 20/40- LE. Spectral domain optical coherence tomography (SD-OCT) and fundus photography were also obtained. Evaluation of MPOD spatial distribution up to 8 degrees eccentricity from the fovea was performed using HFP.
Results: SD-OCT indicated a persistence of multiple inner retinal layers within the foveal region in the RE and LE including symmetric foveal thickening consistent with foveal hypoplasia. Fundus photography showed mild retinal pigmented epithelial (RPE) hypopigmentation and a poorly demarcated macula. OriginPro 9 was used to plot MPOD spatial distribution of the subject and a 33-subject sample. The OCA subject demonstrated a foveal MPOD of 0.10 with undetectable levels at 6 degrees eccentricity. The study sample showed a mean foveal MPOD of 0.34 and mean 6 degree eccentricity values of 0.03.
Conclusions: Consistent with previous macular pigment (MP) studies of OCA, overall MPOD is reduced in our subject. Mild phenotypic expression of OCA with high functional visual acuity may represent a Henle fiber layer amenable to additional MP deposition. Further study of MP supplementation in OCA patients is warranted.
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http://dx.doi.org/10.1016/j.optom.2014.03.001
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PALABRAS CLAVE
densidad óptica del pigmento macular;
albinismo oculocutáneo;
distribución espacial

Distribución espacial de la densidad óptica del pigmento macular medida en un paciente con albinismo oculocutáneo

Resumen
Objetivo: Los estudios previos sobre distribución de la densidad óptica del pigmento macular (DOPM) en pacientes con albinismo oculocutáneo (AOC) han utilizado principalmente técnicas de medición objetivas que incluyen la reflectometría y la autofluorescencia del fondo de ojo. Reportamos aquí el caso de un paciente con AOC y su correspondiente distribución de la DOPM, evaluada mediante fotometría intermitente heterocromática.
Métodos: Un paciente con diagnóstico de AOC se presentó con antecedentes oculares que incluían cirugía de estrabismo del OI, ambliopía persistente y un leve nistagmo latente. La agudeza visual mejor corregida era de 20/25- OD y 20/40- OI. También se realizaron tomografía de coherencia óptica de dominio espectral (SD-OCT) y fotografía del fondo de ojo. La evaluación de la distribución espacial de la DOPM de hasta 8 grados de excentricidad desde la fóvea se realizó mediante fotometría intermitente heterocromática.
Resultados: La SD-OCT indicó la persistencia de múltiples capas internas de la retina dentro de la región foveal en el OD y OI, incluyendo engrosamiento foveal simétrico, consistente con hipoplasia foveal. La fotografía del fondo de ojo mostró una leve hipopigmentación epitelial de la retina, y una mácula débilmente demarcada. Se utilizó OriginPro 9 para trazar la distribución espacial de la DOPM del paciente, y una muestra de 33 sujetos normales. El paciente de AOC mostró una DOPM foveal de 0,10 con niveles indetectables a excentricidades de 6 grados. La muestra control del estudio mostró una DOPM foveal de 0,34 y unos valores de excentricidad a 6 grados de 0,03.
Conclusiones: En consonancia con estudios anteriores sobre el pigmento macular en AOC, la DOPM total se redujo en nuestro paciente. La expresión fenotípica leve del AOC con alta agudeza visual funcional puede ser compatible con la presencia de la capa de fibras de Henle, susceptible de deposición adicional de pigmento macular. Son necesarios futuros estudios sobre el suplemento de pigmento macular en pacientes con AOC.
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Introduction

Research has identified the constituents of human macular pigment (MP) as lutein, zeaxanthin, and a lutein-metabolite, *meso*-zeaxanthin.[1] MP is a membrane-bound compound found primarily within the photoreceptor axons (Henle fiber layer) and the inner plexiform layer and to a lesser extent the retinal pigmented epithelium[2] and photoreceptor outer segments.[3] Macular pigment optical density (MPOD) is highest in the central retina peaking at the fovea and falling to undetectable levels outside of 7° of eccentricity from the fovea.[4]

Oculocutaneous albinism (OCA) has been associated with photophobia, nystagmus, reduced retinal pigment and foveal hypoplasia. Previous MP studies of OCA showed that subjects with low levels of ocular melanin had low levels of retinal carotenoids and photophobia symptoms that were partially explained by reduced MPOD.[5,6] This study measured MPOD distribution in a subject with OCA.

Methods

This study utilized customized heterochromatic flicker photometry (cHFP) to measure the spatial distribution of MP across the central 16° of the retina and compared results against measures of intraocular scatter. Only right eye (RE) measurements were taken including both the foveal peak density of MP and the integrated value across the central 16° of retina in relation to glare disability and intraocular scatter. The use of human subjects was approved by the UMSL Institutional Review Board and adhered to the Declaration of Helsinki.

A subject with a history of oculocutaneous albinism (OCA) participated in the same testing as a study cohort consisting of 33 subjects with unremarkable ocular history and minimum best corrected visual acuity of 20/25. The subject's medical history included a diagnosis of OCA at 1 month of age with no associated albinism syndromes. Ocular history included strabismus surgery of the LE at 1 year of age, persistent amblyopia of the LE, and latent nystagmus. Best corrected visual acuity was 20/25- RE and 20/40- LE. Anterior segment examination showed mild, diffuse iris trans-illumination of equal caliber in both eyes. Posterior segment evaluation recorded a hypopigmented fundus with a poorly demarcated macula and mild transparency suggestive of reduced MP in both eyes. MPOD measurement was performed by the subject with translucent patching of the LE to minimize nystagmus. A Cirrus optical coherence tomographer (Zeiss Meditech) was used

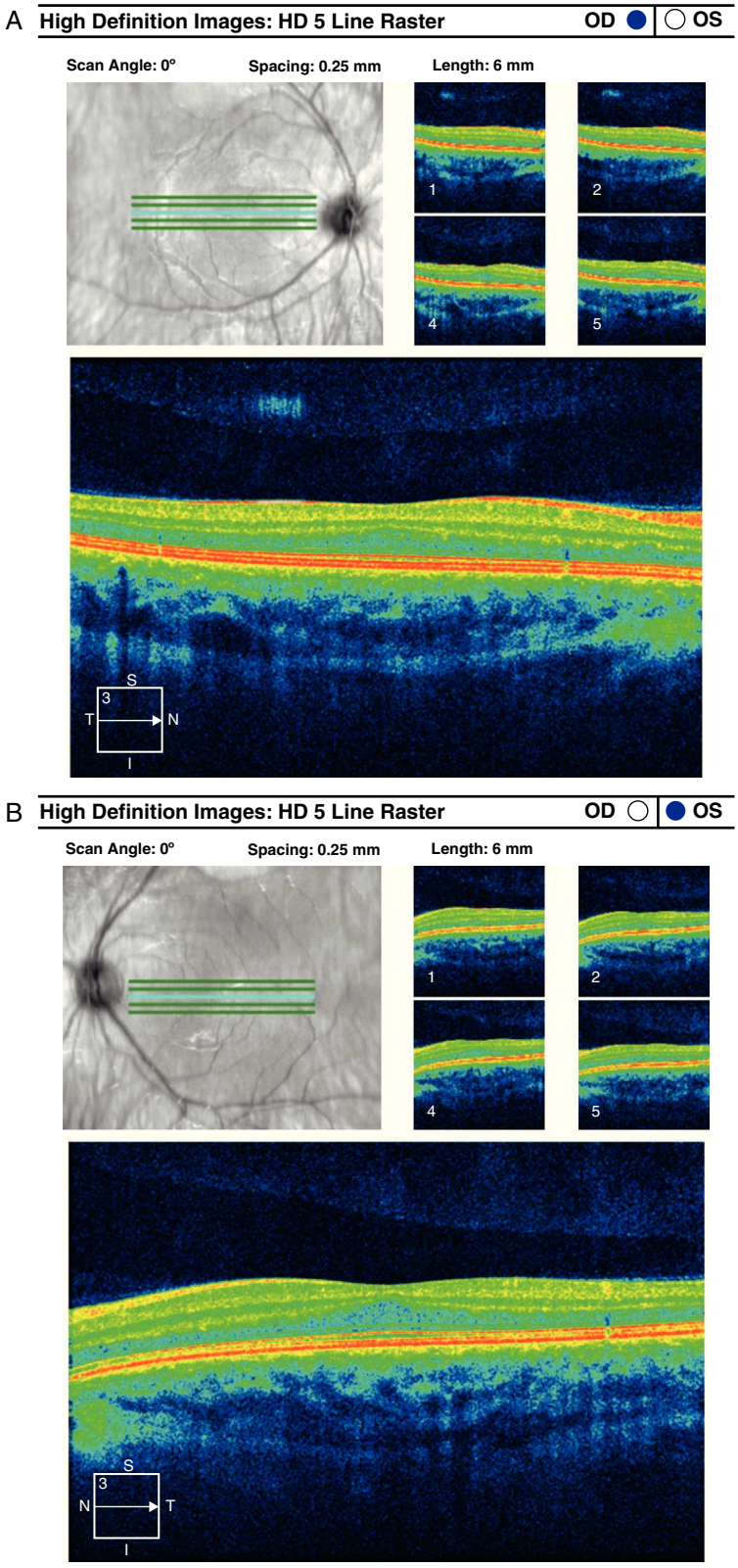

Figure 1 (A) SD-OCT HD 5-Line Raster imaging of the subject's RE. (B) SD-OCT HD 5-Line Raster imaging of the subject's LE.

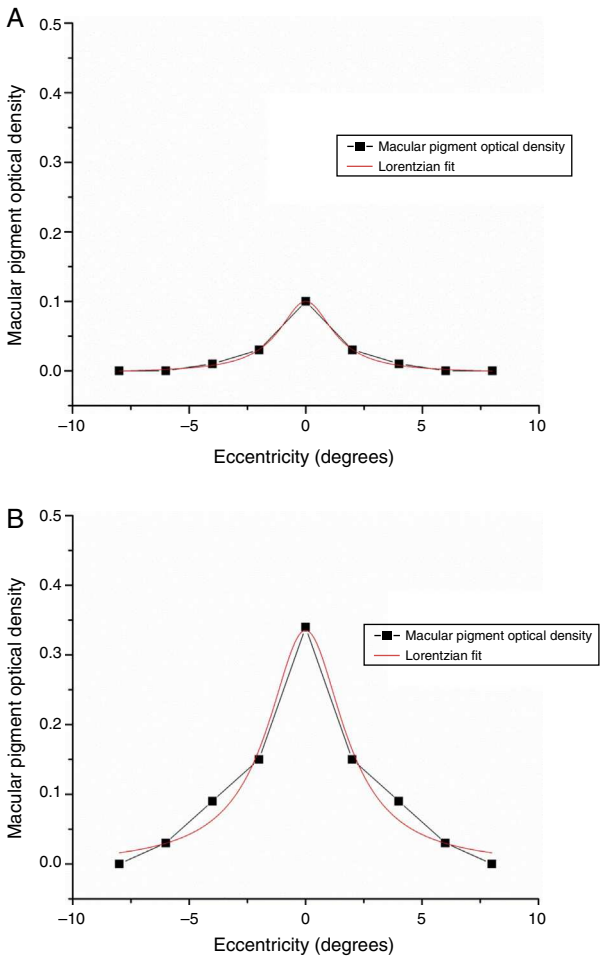

Figure 2 (A) MPOD spatial distribution of the subject's RE fit with a Lorentzian function. (B) Overall mean MPOD spatial distribution of a 33-subject sample fit with a Lorentzian function.

to perform a high-definition 5-line raster scan. A C-Quant device (Oculus, USA) was used to determine an intraocular scatter value for the RE of the subject and each study subject by calculating the mean of five consecutive trials that met validity and repeatability measures.

Results

SD-OCT results revealed the presence of multiple inner retinal layers within the foveal region and symmetric thickening consistent with foveal hypoplasia of both eyes (Fig. 1A and B). The subject demonstrated MPOD levels of 0.10 at the fovea, 0.03 at 2°, 0.01 at 4° and undetectable at 6° of eccentricity in the RE. MPOD results for the study sample included mean values of 0.34 at the fovea, 0.15 at 2°, 0.09 at 4° and 0.03 at 6° of eccentricity in the RE. OriginPro9 software was used to fit the MPOD spatial distribution pattern to a Lorentzian function for both the subject and the study sample. Differences in the area under the curve and spatial distribution can be seen in the MPOD profile of the subject versus study sample (Fig. 2A and B). The subject demonstrated a mean intraocular scatter value of 1.28 while intraocular scatter values from study sample showed a mean value of 0.78 with a $\sigma = 0.13$.

Discussion

OCA affects the eyes, skin, and hair and may be tyrosinase-negative or tyrosinase-positive, and follows autosomal recessive inheritance patterns. Cardinal signs of OCA from a SD-OCT evaluation include absence of a central foveal depression, presence of the nerve fiber layer, ganglion cell layer, and other inner retinal layers across the fovea, and increased reflectivity of the choroid due to reduced levels of melanin.[7]

To our knowledge, this is the first assessment of MPOD using cHFP in an OCA subject. According to previous MP studies of OCA,[5,6] the foveal MPOD values found for our subject are relatively high. The foveal MPOD value of 0.10 log unit may be a result of mild phenotypic expression of OCA in our subject. This may also account for the mild latent nystagmus accompanied by the high functional acuity resulting from the near normal foveal morphology identified by SD-OCT imaging. Near normal morphology of the photoreceptor axon layer and the inner plexiform layer in foveal hypoplasia may allow macular carotenoid deposition through oral supplementation.[8] Increased MPOD may reduce glare and photophobia-related symptoms experienced in OCA and increase the protection of the underlying photoreceptor layer through anti-inflammatory, antioxidant and short wavelength light absorption properties. The amenability of macular carotenoids may hold promise in other central retinopathies with decreased levels of MPOD.[9,10] Further study of MPOD response to oral supplementation in subjects with OCA is warranted.

Conflicts of interest

The authors have no conflicts of interest to declare.

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