Research Article / *Araştırma Makalesi*

Ege Journal of Medicine / *Ege Tıp Dergisi* 2020; 59 (3): 192-195

Anterior segment evaluation in unilateral oculodermal melanocytosis

Unilateral okülodermal melanositozda ön segment değerlendirilmesi

Melis Palamar

Ege University, Faculty of Medicine, Department of Ophthalmology, Izmir, Turkey

Abstract

Aim: To compare the anterior segment parameters of effected and normal eyes of unilateral oculodermal melanocytosis (ODM) patients.

Materials and Methods: A retrospective chart and Scheimpflug camera database review for unilateral ODM patients were conducted. Anterior chamber depth (ACD), anterior chamber volume (ACV), anterior chamber angle (ACA) width, central corneal thickness (CCT), keratometry, pupilla diameter parameters of the eye with ODM (Group 1) was compared to the normal (Group 2) eye.

Results: Sixteen patients with a mean age of 32.50±12.20 (range 12 and 48) were enrolled. No difference between groups were detected in terms of ACD, ACV, CCT, keratometry, pupilla diameter (p=0.655, p=0.812, p=0.131, p=0.434, p=0.325, p=0.509, respectively). Although ACA was lower in Group 1 the difference was not statistically significant (p=0.057).

Conclusion: Although not statistically significant, there seems to be a tendency for shallow ACA readings in unilateral ODM eyes when compared to normal contralateral eyes. However, more studies with larger number of patients that evaluate this issue are needed.

Keywords: Anterior segment, cornea, eye, oculodermal melanocytosis, topography.

Öz

Amaç: *Tek taraflı okülodermal melanositoz (ODM) hastalarının etkilenmiş gözleri ve karşı taraf normal gözlerini ön segment parametreleri açısından karşılaştırmak.*

Gereç ve Yöntem: *Tek taraflı ODM olan hastaların dosyaları ve Scheimpflug kamera ile alınmış ölçümleri retrospektif olarak incelendi. ODM'li gözler (Grup 1) ve karşı normal gözler (Grup 2) ön kamara derinliği (ÖKD), ön kamara hacmi (ÖKH), ön kamara açısı (ÖKA) genişliği, santral kornea kalınlığı (SKK), keratometri, pupilla çapı parametreleri açısından karşılaştırıldı.*

Bulgular: *Ortalama yaşları 32,50±12,20 (12 ve 48 arası) olan 16 hasta çalışmaya dahil edildi. Gruplar arasında ÖKD, ÖKH, SKK, keratometri, pupilla çapı değerleri açısından farklılık saptanmadı (sırasıyla, p=0,655, p=0,812, p=0,131, p=0,434, p=0,325, p=0,509). ÖKA değerleri Grup 1 gözlerde Grup 2 gözlerden daha düşük olmasına rağmen bu fark istatistiksel olarak anlamlı bulunmadı (p=0,057).*

Sonuç: *İstatistiksel olarak anlamlı olmamakla beraber ÖKA değerleri tek taraflı ODM hastalarının etkilenmiş gözlerinde karşı taraf normal gözlerden daha dar olma eğilimindedir. Bu konuda daha çok sayıda hasta ile yapılacak yeni çalışmalara ihtiyaç vardır.*

Anahtar Sözcükler: *Göz, kornea, okülodermal melanositoz, ön segment, topografi.*

Corresponding author: Melis Palamar
Ege University, Faculty of Medicine, Department of Ophthalmology, Izmir, Turkey
E-mail: *melispalamar@hotmail.com*
Application date: 24.07.2019 Accepted: 16.03.2020

Introduction

Ocular/oculodermal melanocytosis is a congenital periocular pigmentary condition that can lead to development of uveal melanoma, estimated at 1 in 400 affected patients (1). It was first described by Ota (2) as a periocular pigmentary condition. This condition is known with several names including *melanosis oculi*, *nevus of Ota*, and *oculocutaneous melanosis*. This congenital pigmentary abnormality consists of excess melanocytes within the periocular skin, sclera, uvea, orbit, meninges, palate, or tympanic membrane. Although it is generally unilateral, rare bilateral cases might also be seen. The melanocytic pigmentation can involve the skin (dermal) or eye (ocular) alone, or can be seen in both tissues (oculo-dermal). The main concern with this problem is the risk for development of melanoma, predominantly in the uvea (3, 4). It is also known that glaucoma and anterior uveitis are more commonly seen in these eyes (5).

The evaluation of the anterior segment parameters enables important information that improves our knowledge and understanding of aqueous humour dynamics, primary open-angle glaucoma, pigmentary glaucoma, and ocular pharmacokinetics. Pentacam rotating Scheimpflug camera (Oculus Optikgeräte GmbH, Wetzlar, Germany) enables quantitative information and qualitative imaging of the anterior and posterior surfaces of the cornea, anterior chamber, iris, and lens (6, 7).

In this study, it is aimed to investigate the effect of oculodermal melanocytosis on anterior chamber depth (ACD), anterior chamber volume (ACV), anterior chamber angle (ACA) width, central corneal thickness (CCT), keratometry, and pupilla diameter. To the best of our knowledge, the comparison of these parameters with Pentacam in unilateral oculodermal melanocytosis patients have not previously been reported.

Materials and Methods

A total of 16 unilateral oculodermal melanocytosis patients (4 males, 12 females) with the mean age of 32.50 ± 12.20 (range 12 and 48) were enrolled. All patients were examined, and all measurements were performed by the same ophthalmologist. The demographic data (age and gender) and clinical evaluation including best-corrected visual acuity (BCVA), intraocular pressure (IOP), oculodermal melanocytosis site were noted. The affected (Group 1) and contralateral normal eyes (Group 2) were compared. None of the individuals had any corneal or posterior segment pathology, glaucoma, history of previous eye surgery or trauma, history of using topical/systemic medications or systemic diseases that could interfere the eye. None of the individuals demonstrated corneal abnormality related to ocular/oculodermal melanocytosis and none had the history of contact lens use.

All Pentacam measurements were obtained under standart dim light as previously described (7-9). The Pentacam CES system is based on a 180° rotating Scheimpflug camera which can take 12–50 single images to reconstruct the anterior chamber. Herein, anterior segment reconstructions were produced with 25 single captures. After completing a scan, Pentacam software constructs the 3-dimensional image of the anterior segment and calculates the anterior chamber parameters. This imaging provides measurements of ACD, ACV, ACA width, CCT, pupil size and keratometry.

The study and data collection were compliant with the principles of the Declaration of Helsinki. The study was approved by the Ethical Board of Ege University Faculty of Medicine (70198063-050.06.04). Statistical analysis was performed with SPSS for Windows Version 12.0 (SPPS Inc., Chicago, IL, USA). All data were reported as averages ± standard deviations (SD). T test was used for parametric and Mann-Whitney U test was used for nonparametric data. A value of $p< 0.05$ was considered statistically significant.

All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards. Informed consent was obtained from all individual participants included in the study.

Results

All the eyes in Group 1 had both dermal and ocular components and had dark fundus. Mean BCVA of Group 1 and Group 2 were 0.94 ± 0.22 (range, 0.1 and 1) and 1.00 ± 0.00 (range, 1 and 1), respectively (p=0.509). Mean IOP of Group 1 and Group 2 eyes were 14.50 ± 1.82 (range, 10 and 18) mmHg and 14.37 ± 1.89 (range, 10 and 18) mmHg, respectively (p=0.780).

Table-1. The demographics and the Scheimpflug camera measurements of the effected (Group 1) and normal (Group 2) eyes.

	Affected eye (Group 1) (Mean, SD, range)	**Normal eye (Group 2)** (Mean, SD, range)	**p value**
BCVA	0.94 ± 0.22 (0.1-1)	1.00 ± 0.00 (1-1)	0.509
IOP (mmHg)	14.50 ± 1.82 (10-18)	14.37 ± 1.89 (10-18)	0.780
ACD (mm)	2.94 ± 0.32 (2.25-3.47)	2.93 ± 0.33 (2.30-3.50)	0.655
ACV (mm)	163.12 ± 35.91 (96-216)	159.00 ± 35.19 (108-217)	0.812
ACA width (mm)	35.03 ± 6.26 (22.10-44.70)	32.42 ± 5.90 (22.90-48.40)	0.131
Pupil size (mm)	3.90 ± 1.03 (2.86-6.39)	4.09 ± 1.12 (2.56-6.33)	0.509
CCT (micrometer)	565.43 ± 42.55 (516-658)	572 ± 42.58 (497-639)	0.150

SD: standard deviation; BCVA: best corrected visual acuity; IOP: intraocular pressure; ACD: anterior chamber depth; ACV: anterior chamber volume; ACA: anterior chamber width; CCT: central corneal thickness.

Mean ACD measurements of Group 1 and Group 2 eyes were 2.94 ± 0.32 (range, 2.25 and 3.47) mm and 2.93 ± 0.33 (range, 2.30 and 3.50) mm, respectively (p=0.655) (Table-1). Mean ACV of Group 1 and Group 2 were 163.12 ± 35.91 (range, 96 and 216) mm and 159.00 ± 35.19 (range, 108 and 217) mm, respectively (p=0.812).

Mean ACA width measurements of Group 1 and Group 2 eyes were 35.03 ± 6.26 (range, 22.10 and 44.70) degree and 32.42 ± 5.90 (range, 22.90 and 48.40) degree, respectively (p=0.131).

Mean pupil size measurements of Group 1 and Group 2 eyes were 3.90 ± 1.03 (range, 2.86 and 6.39) mm and 4.09 ± 1.12 (range, 2.56 and 6.33) mm, respectively (p=0.509).

Mean CCT measurements of Group 1 and Group 2 eyes were 565.43 ± 42.55 (range, 516 and 658) micrometer and 572 ± 42.58 (range, 497 and 639) micrometer, respectively (p=0.057).

Mean K1 values of Group 1 and Group 2 eyes were 42.53 ± 1.46 (range, 40.60 and 44.70) diopters and 42.23 ± 1.38 (range, 40.20 and 45.00) diopters, respectively (p=0.434). Mean K2 of Group 1 and Group 2 eyes were 43.31 ± 1.62 (range, 40.90 and 45.60) diopters and 43.10 ± 1.32 diopters (range, 41.50 and 45.50), respectively (p=0.325).

Discussion

Ocular/oculodermal melanocytosis is a well-known predisposing factor for uveal melanoma (1, 10, 11). The largest study in the literature on ocular/oculodermal melanocytosis is by Teekhasaenee et al. (5) which evaluated 194 Thai patients. Elevated IOP developed in the ipsilateral eyes of 15 patients at initial examination and in five others during follow-up, for a total of 10.3% of the patients. Three patients had congenital or late congenital glaucoma, 14 had ocular hypertension or open-angle glaucoma, and three had acute angle-closure glaucoma. Five of the patients with open angles had acute pressure rises accompanied by anterior uveitis. They concluded that glaucoma is common in eyes with oculodermal melanocytosis and may develop at any age. For this reason, they recommended regular examination of these patients for any glaucomatous or uveitic signs. No signs of glaucoma, elevated IOP or uveal reaction were detected in this case series.

Oculodermal melanocytosis eyes were demonstrated to have thicker ciliary body with higher ultrasound reflectivity on ultrasound biomicroscopy compared with the unaffected eye and with the noninvolved sectors in the same eye (12). Although oculodermal melanocytosis is known to be confined to uvea the increased volume of uveal tissues might be suspected to indirectly cause anterior segment changes. Moreover, it was shown that cornea may present melanocytosis of the epithelial, stromal, and endothelial layers, and in some cases, the pupil may respond poorly to pharmacological dilation (5). This data brings about the idea of effected anterior segment parameters such as ACD, ACA

width, etc. in these eyes. However, this study showed us that no statistically significant difference in terms of ACD, ACV, CCT, keratometry, pupilla diameter is evident between the normal and effected eyes. Although ACA was lower in oculodermal melanocytosis positive eyes the difference did not reach statistical significance. It is possible that these patients with narrow angles might develop glaucoma in time. For this reason, as was suggested earlier, these eyes should be followed-up for early IOP rise detection.

The small sample size of 16 cases in this study might be questioned. However, it is a significant number of patients considering the fact that only patients with unilateral involvement are included which enabled us to have a control for each case (contralateral normal eye).

Conclusion

Although, there seems to be a tendency for shallow ACA readings, oculodermal melanocytosis do not affect the anterior segment parameters as measured with Scheimpflug camera. Studies with larger number of patients comparing choroidal thickness and anterior segment changes are still needed.

Conflict of interest: The authors have not declared any conflict of interest in this study.

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