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PELYUSİD MARGİNAL DEGENERASİYASI ZAMANI SKLERAL KONTAKT LİNZALARIN ÜSTÜNLÜYÜ (KLİNİK HAL)

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XÜLASƏ

Məqalədə pelyusid marginal degenerasiyası (PMD) üçün terapevtik yanaşma olaraq skleral kontakt linzalardan istifadənin üstünlükləri təqdim edilmişdir. PMD nadir rast gəlinən, iltihabı olmayan buynuz qişanın ektatik xəstəliyidir. Keratokonusla oxşar xüsusiyyətlərə malik olduğu üçün, PMD-nın doğru diaqnozunu və effektiv terapevtik yanaşmaların seçilməsi daha çətin ola bilər. Buynuz qişanın topoqrafiyasından istifadə edərək PMD-nın dəqiq diaqnozunu qoyduqdan sonra, həm skleral kontakt linzaların, həm də hibrid linzaların istifadəsindən sonra ən yüksək korreksiya olunmuş görmə itiliyində baş verən dəyişikliklər qiymətləndirilmişdir. Yanlış diaqnoz halları müalicə üsullarını gecikdirə bilər. Xüsusilə, iki xəstəlik arasında terapevtik yanaşmalar fərqli olduqda, bu, əlavə ağırlaşmalara səbəb ola bilər.

Açar sözlər: *pelyusid marginal degenerasiyası, skleral kontakt linza, astiqmatizm, keratokonus, hibrid kontakt linza, buynuz qişanın topoqrafiyası, görmənin reabilitasiyası*

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SUPERIORITY OF SCLERAL CONTACT LENS IN PELLUCID MARGINAL DEGENERATION (CLINICAL CASE)

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SUMMARY

This article aims to highlight the advantages of using scleral contact lenses as a therapeutic approach for Pellucid Marginal Degeneration (PMD). PMD is a rare non-inflammatory corneal ectatic disorder. Due to its similar features to Keratoconus, it is harder to correctly diagnose and use effective therapeutic approaches for PMD. After a definitive diagnosis of PMD using corneal topography, we have evaluated the change in best corrected visual acuity (BCVA) both after the use of scleral contact lens and in hybrid lens. It is vital to remember that various cases of misdiagnosis may not only delay the management and treatment modalities, but also these could further result in complications, especially when the therapeutic approach between two diseases may differ.

Keywords: *pellucid marginal degeneration, scleral contact lens, astigmatism, keratoconus, hybrid contact lens, corneal topography, visual rehabilitation*

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ПРЕИМУЩЕСТВО СКЛЕРАЛЬНЫХ КОНТАКТНЫХ ЛИНЗ ПРИ ПЕЛЛЮЦИДНОЙ МАРГИНАЛЬНОЙ ДЕГЕНЕРАЦИИ (КЛИНИЧЕСКИЙ СЛУЧАЙ)

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РЕЗЮМЕ

Целью этой статьи является оценка преимущества использования склеральных контактных линз в качестве терапевтического подхода при пеллюцидной маргинальной дегенерации (ПМД). ПМД — редкое невоспалительное эктатическое заболевание роговицы. Из-за схожести признаков с кератоконусом диагностика ПМД и выбор эффективных терапевтических подходов могут быть затруднены. После окончательной диагностики ПМД с использованием топографии роговицы были оценены изменения максимально корригированной остроты зрения как при использовании склеральных контактных, так и гибридных линз. Важно помнить, что ошибки в диагностике могут не только задержать начало лечения и ведения пациента, но и привести к осложнениям при различии терапевтических подходов к этим двум заболеваниям.

Ключевые слова: *пеллюцидная маргинальная дегенерация, склеральные контактные линзы, астигматизм, кератоконус, гибридные контактные линзы, топография роговицы, зрительная реабилитация*

Pellucid marginal degeneration (PMD) is a non-inflammatory progressive ectatic disorder of the cornea [1,2]. Pellucid corneas often cover a distorted geographic area which extends from the lower corneal edge to the center of cornea. This finding may lead to a misdiagnosis of Keratoconus. Although both diseases can cause severe visual deterioration, it is hard to differentiate by means of clinical exam. For distinction, it is advised to use corneal topography for a definitive diagnosis. In PMD, the ectatic zone, which is 1-2 mm from the limbus, lies above the point of the maximum corneal thinning [1,3,4]. It is typically characterized by a clear ectasia in the inferior and peripheral region of the cornea [1,4].

Some features of PMD include, inferior peripheral steepening extending into the mid-peripheral, inferior oblique corneal meridians, which results in a characteristic "crab-claw", "butterfly" or "kissing doves" appearance on the curvature/keratometric map on topography [1,3,4]. However, it is important to remember that these findings can also be observed in some cases of advanced keratoconus; hence emphasis must be laid on clinical and pachymetric correlation for a definitive diagnosis. The pachymetry of PMD patients shows a reversal from normal, thinner in the periphery (usually inferior) and thicker in the center [1,2].

In keratoconus, involvement of the central two-thirds of the cornea, ectasia, and thinning at a common location with the apex of cone shifting inferiorly along with the presence of signs like scissoring reflex, Vogt striae, Fleischer ring, and presence of asymmetric bow tie with a skewed radial axis on topography can be noted [1,3].

Treatment usually consists of vision correction with glasses in early cases, although a therapeutic approach of specially designed contact lens (ie. hybrid contact lenses, scleral contact lenses) are used in more pronounced cases. Hybrid Contact Lenses, have a central rigid zone with a circumferential peripheral soft skirt to utilize the visual performance of hard lenses and the comfort and stability of lenses. On the other hand, Scleral Contact Lenses (SCLs) are considered for the treatment of PMD, as they are more flexible gas-permeable contact lenses, designed to cover the entire corneal surface [1]. Most SCLs are 14 to 24 millimeters in diameter, vaulting over the cornea and resting on the sclera, as the fluid within keeps the eyes moist for longer. The main advantage of SCLs, apart from the provision of improved comfort and stable visual acuity, is that they delay keratoconus and PMD. The absence of corneal-lens interaction and the continued lubrication has been shown to improve comfort, dryness and quality of life in patients that have been intolerant to other contact lens modalities [3,5].

The aim of this case report is to assess improvement of visual acuity and alleviation of symptoms in our PMD patient who has been fitted with scleral contact lenses. It is also important to highlight that the patient had been previously misdiagnosed with Keratoconus, and other treatment modalities were found not to be beneficial.

Case Report

A 25-year-old female patient was consulted at Atakent Acıbadem Hospital, Ophthalmology outpatient clinic with the chief complaint of decreased vision in both eyes and itching sensation. There was a history of constant eye-rubbing but no other trauma to the eye was mentioned. Upon asking for any other medical history, she informed us that she had a prior diagnosis of keratoconus from another hospital. Her family history also revealed that her grandfather suffered from vision loss due to keratoconus.

In the Ophthalmological exam conducted, the visual acuity was initially noted as 0.8 on the right eye with -2.25 -1.25 AXIS 80, however the visual acuity was 0.5 with the -1.75 -2.25 AXIS 115 after correction. On the slit-lamp exam, central corneal thinning was observed, and the Fundus exam was unremarkable. In corneal topography 'kissing doves' appearance was noted bilaterally. Corneal thinnest region was 512 UM in the right eye and 487 UM in the left eye.

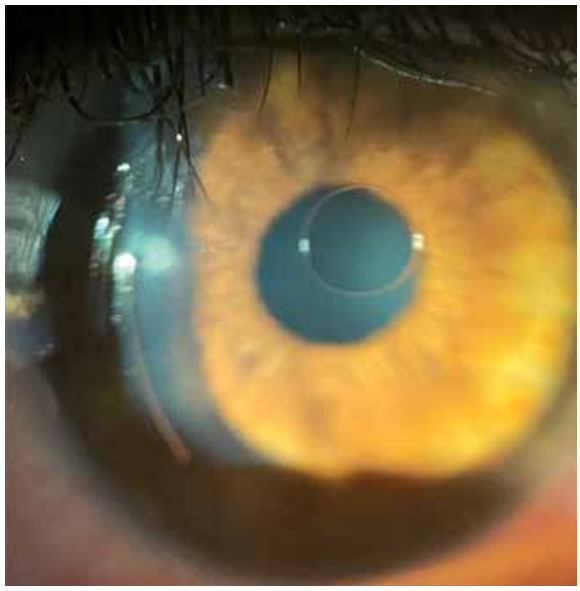
For the therapeutic approach, it was initially decided on the hybrid lens upon patient's wishes. After hybrid lenses were set, a visually disturbing bubble which was located on the central part of the visual axis was noted. In the follow-up examination, the patient was unsatisfied, as their vision didn't improve with the application of hybrid lenses. Hence afterwards Flexfit Scleral Contact Lens was applied to the patient.

The base curve (BC) parameters of the scleral contact lens were OD 8.45 and OS 8.25 and the powers of right and left eyes were +4 D and +2.50 D, respectively. Diameters of the scleral lenses were 16.3 mm. Bifocal Vaults were 3800 and 4000 on right and left eyes respectively. After the insertion, the patient was assessed by slit-lamp biomicroscopy to ensure the comfort of the fitting **(Figure 1)**.

Discussion

Herein we reported a PMD patient who had a prior misdiagnosis for Keratoconus. After the insertion of Scleral Contact Lenses (SCLs) to our patient, a significant increase in visual acuity was observed. Prior to the treatment, BCVA was recorded as 0.5 and

a)



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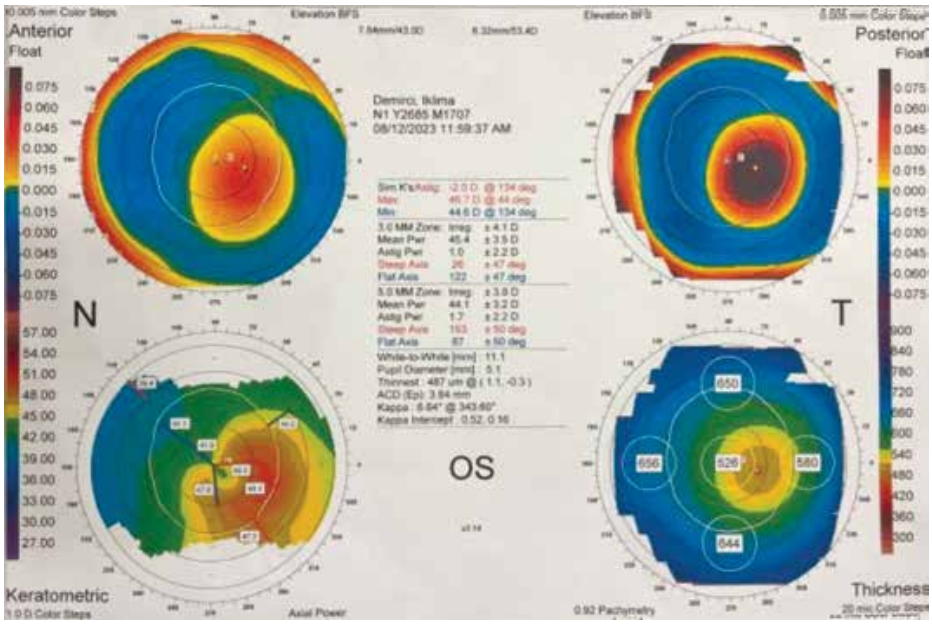

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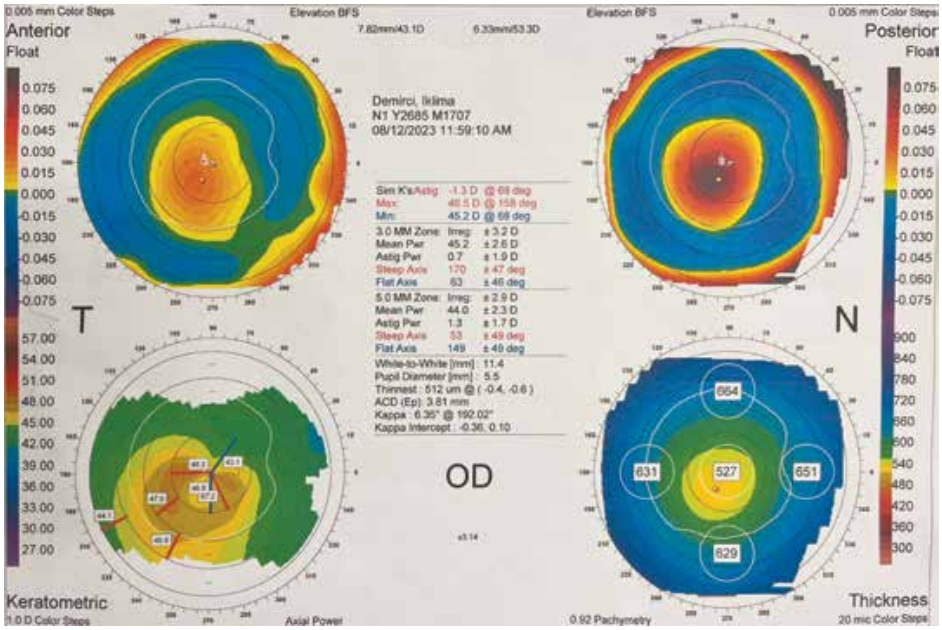

Figure 1. *a) Slit-lamp biomicroscopy after use with the scleral contact lens b) slit-lamp biomicroscopy after use with the hybrid contact lens c) OD - keratometric map on topography d) OS - keratometric map on topography.*

after SCL fitting, BCVA was recorded as 1.0. In addition to improved BVCA, the patient mentioned alleviation of the itching sensation, a resolution of dryness of the eye, as well as a notable improvement of blurry vision.

In our case report, we stated a female patient who had a bilateral asymmetrical presentation. Her visual complaint was primarily in the left eye, but after obtaining the corneal topography, the involvement of the right eye was noted. It is important to highlight that although PMD is regarded as a bilateral ectatic disorder, cases where asymmetrical or unilateral presentation can be of valuable contribution to research papers. As previously mentioned, atypical presentations of PMD are uncommon, but they have been previously reported in the Eye (Journal of Ophthalmology), where a 46-year-old man presented with chief complaint of progressive dimness of vision in the left eye. This unusual unilateral presentation of PMD is vital to differentiate this condition from other ectatic disorders [6].

Considering therapeutic approaches, our patient who was previously misdiagnosed with keratoconus, had minimal improvement regarding her symptoms when using prescription glasses. However, as the disease progressed, it was decided that using prescription glasses might not be a good therapeutic approach for the future. It was feared that it would potentially worsen the corrected visual acuity seeing that astigmatism might become irregular, with higher-order optical aberrations.

Conditions such as irregular astigmatism with high corneal toricity can render prescription glasses insufficient for correcting astigmatism. However, for correcting the visual acuity in various corneal ectatic disorders, other forms of initial treatment modalities like hybrid and scleral lens fitting were deemed to be an appropriate option. It was confirmed with various reports that the use of specially designed contact lenses was shown to be superior to prescription.

Specially designed contact lenses (CL) remain to be an effective and safe option for improving visual acuity in corneal ectatic diseases [7,8]. Current designs and materials have significantly expanded the application options for patients with various corneal ectatic disorders. According to the 2015 Global Keratoconus Consensus Report, RGP lenses have significantly improved the visual acuity

and three-dimensional depth perception than glasses in moderate to advanced keratoconus. Intolerance and complications secondary to RGP lenses have led to the development of safer and more comfortable designs, such as scleral and hybrid lenses [7,8,9]. In our case, the Airflex Hybrid and ICD- Flexfit scleral lenses were applied and specially with the use of scleral lenses, the visual acuity was increased.

A 2020 report on international contact lens prescribing trends showed that while the overall rate of gas GP lens prescribing has remained stable at %13, scleral lens prescribing which were negligible 15 to 20 years ago has increased to now account for about %3 of all GP lenses prescribed [10]. In this cohort, SL wearers had the steepest mean keratometry readings compared to patients prescribed other lens types and yet maintained a mean visual acuity of better than 20/30. This improvement in visual acuity is consistent with a larger percentage of patients in corneal GP, SL and hybrid lenses in the 2020 group compared to 2010. Corneal GP lenses continued to be the most frequent type of contact lens used, though SL use increased from zero in 2010 to nearly one in every five patients in 2020. This is comparable to a 2013 cross sectional study of 244 patients with keratoconus in which %68 was using corneal GP lenses. This trend is also consistent with a SCOPE study group survey of SL prescribers that reported over half of respondents fit their first patient with SLs between 2010 and 2015. The SCOPE study group also reported that for providers graduating after 2009, there was an increase in the number of new graduates trained in SL fitting and an increase in utilization of SLs outside of hospital and tertiary care settings [11].

The absence of corneal-lens interaction and the continued lubrication has been shown to improve comfort, dryness and quality of life in patients that have been intolerant to other contact lens modalities. Hybrid lenses can be made in keratoconus specific designs and have a high oxygen permeability (Dk) silicone hydrogel skirt to address prior issues with lens tightening and hypoxic-related corneal neovascularization that was previously seen with low Dk hydrogel skirts. Not only we have seen significant improvement in best correct visual acuity with SCL's, but patients were also significantly less likely to express lens-related discomfort with SCLs.

In our case, it was important to fit the patient with a treatment modality which would not further hinder our patients daily life, hence SCLs were preferred due to their gas-permeable properties, and them being suitable for a long-wear time.

The SCLs cover the cornea better due to its large diameter, better quality vision, comfortable use and cornea stabilization. On the other hand, hybrid lenses have in comparison low oxygen permeability and limited parameters, as well as limited movement of the diameters of the lens. Therefore they would not rank high in the advised treatment options. In addition, change in the basic curve doesn't make the expected change in the movement of the lens, hence, we are less inclined to use hybrid lenses as an option.

Conclusion

With the presentation of our case, we highlighted the advantages of SCLs like improving the comfort and the quality of vision in PMD patients who were misdiagnosed as keratoconus. Patients who have a low spectacle BCVA and a higher gain of visual acuity with scleral lenses are good candidates. The authors stated that PMD patients tolerate the scleral lens well. As a result, a scleral lens may be a good option for patients with irregular ocular surfaces.

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