

Surgical Treatment of a Superiorly Located Pellucid Marginal Degeneration Associated with Cataract

Üst Kadran Yerleşimli ve Kataraktın Eşlik Ettiği Bir Pellucid Marjinal Dejenerasyon Olgusunun Cerrahi Tedavisi

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ABSTRACT Pellucid marginal degeneration (PMD) is a rare disorder of the peripheral cornea causing corneal thinning, generally at the inferior quadrants of the cornea. Here, we present a 75 year old patient with blurred vision in his right eye. On ophthalmic and corneal topographic examinations, PMD located at the superior quadrant of the cornea and grade 3 cataract were observed. Corneal wedge resection and primary suturation were performed in an attempt to cause scar formation and flattening in the superior quadrant of the cornea. This enabled us to obtain keratometric measurements and perform cataract surgery with reliable biometric measurements. This is the first reported case with a superiorly located PMD treated with uncomplicated lamellar crescent wedge resection and consequent successful biometry acquisition and cataract surgery.

Keywords: Corneal diseases; cataract; pellucid marginal degeneration; wedge resection

ÖZET Pellucid Marjinal Dejenerasyon (PMD) genellikle korneanın alt kadranlarında yerleşimli olup kornea periferinin incilmesiyle seyreden ve nadir görülen bir korneal bozukluktur. Yetmiş beş yaşındaki erkek hasta kliniğimize sağ gözde görmede azalma şikayeti ile başvurdu. Yapılan oftalmolojik muayene ve kornea topografik incelemesinde kornea üst kadranda yer alan PMD ve eş zamanlı evre 3 katarakt saptandı. Hastamıza korneal lameller kama rezeksiyonu ile birlikte birincil dikiş tekniği uygulanarak skar oluşumu ve buna bağlı kornea üst kadranda düzleşme sağlandı. Cerrahi girişim sonrası iyileşme dönemi tamamlandıktan sonra, biyometrik ve keratometrik ölçümler yapılabildi ve komplikasyonsuz katarakt cerrahisi uygulandı. Üst kadran yerleşimli PMD hastamız, perforasyonsuz lameller kama rezeksiyon sonrasında başarılı biyometrik ölçümlerin alınabildiği ve bu sayede uygun göz içi lens seçimi ile katarakt cerrahisi uygulanan ilk PMD olgusudur.

Anahtar Kelimeler: Kornea hastalıkları; katarakt; pellucid marjinal dejenerasyon; kama rezeksiyon

Pellucid marginal degeneration (PMD) is a rare disorder of the peripheral cornea causing corneal thinning, generally located at the inferior quadrants of the cornea. However the involvement can rarely include superior, temporal and nasal sides of the cornea.¹ It can be associated with atopic dermatitis and keratoconjunctivitis.² The disease generally occurs in the fourth or fifth decade and can cause irregular against-the rule astigmatism with crescent like thinning of the affected corneal zone.³ There are some methods described in the treatment of PMD such as contact lenses, which can be sometimes difficult to fit, or surgical treatments like lamellar keratoplasty, central penetrating keratoplasty and wedge corneal resections with limbal relaxing incisions.^{4,5}

In this case, we report a patient with superiorly located PMD who underwent lamellar corneal wedge resection and primary suturation before a consecutive cataract surgery.

CASE REPORT

A 75 year old male patient presented to our clinic with decreased vision in both eyes. His corrected visual acuity was hand motion in the right eye (with pinhole) and 5/100 in the left eye (with +3.50 +6.0 axis 175). Ocular examination showed presence of papillary reaction and severe allergic conjunctivitis in both eyes and bilateral superior cone formation due to eye rubbing. Intraocular pressure measurements were 10mmHg in the right eye and 14 mmHg in the left eye. He had grade 3 nuclear sclerosis in the right eye determined with scheimpflug photography. His fundus examination was normal. However, his refraction, keratometry and biometry could not be measured in that eye using autorefractokeratometry (Topcon kr8900) and biometry (Zeiss IOLMaster 500) due to superior corneal irregularity. Corneal topography (Wavelight Oculyzer2 Pentacam) analyses showed superior peripheral thinning and crab claw pattern on the superior cornea (Figure 1) resembling superior pellucid corneal marginal degeneration. We planned lamellar crescent wedge resection on the superior cornea with primary tight suturation in order to develop a scar tissue and flatten the superior cornea. We planned to repeat keratometric and biometric measurements after the healing period and informed consent was taken before surgery.

A 120 degree crescent incision on the superior cornea with a 19 gauge stiletto knife was made and a corneal tissue including $\frac{3}{4}$ depth of the cornea with 1mm width and 3mm length was removed. Then, the cornea was sutured with 10-0 nylon sutures and the sutures were buried (Figure 2).

After 6 weeks of follow-up, we removed the corneal sutures and noticed that there was a scar formation along the resection site. The patient's visual acuity in the right eye increased to 20/100.

We were able to obtain biometry, refraction and keratometry measurements before cataract surgery.

With the biometry obtained we were able to calculate the intraocular lens power and then, we performed phacoemulsification and intraocular lens implantation without any complications. Patient's final visual acuity two weeks after cataract surgery was 20/50 with a steeper superior cornea formation.

DISCUSSION

Pellucid marginal degeneration is a rare corneal ectatic disorder of the peripheral thinning cornea without inflammation.⁶ PMD generally occurs at the four o'clock quadrant in the inferior cornea and rarely involves other quadrants including superior corneal site.³ It can be associated with atopic dermatitis and keratoconjunctivitis like in our patient and PMD formation may be explained by abnormal rubbing hypothesis causing keratectasia due to severe allergic conjunctivitis.^{2,7}

PMD and keratoconus are both corneal ectatic disorders and are often misdiagnosed. In keratoconus generally a central cone exists, however in PMD peripheral corneal thinning occurs which is accompanied by steepening just superior to the thinned zone and a 1.0 to 2.0 mm margin of normal cornea that lies between the thinning and the limbus. A crab claw pattern in corneal topography is characteristic for PMD.⁸ Our patient had a peripheral corneal thinning and a characteristic crab claw corneal topographic pattern on the superior corneal quadrants (Figure 1A). Other differential diagnoses for his patient may be Terrien and Furrow corneal degenerations. Terrien marginal degeneration generally presents with white-yellow, punctate, stromal opacifications associated with lipid deposition and vascularization. Also, a pseudopterygium formation may be observed. However, our patient had a clear peripheral cornea and none of the above mentioned findings were observed. Furrow degeneration generally presents between arcus senilis and the limbus with subtle changes in the corneal contour. It cannot be precisely evaluated by keratometry or keratoscopy. Topographic findings and the absence of arcus senilis in our patient helped us to exclude Furrow degeneration.^{9,10}

Spectacle correction and contact lenses should be considered in early and moderate cases. Al-

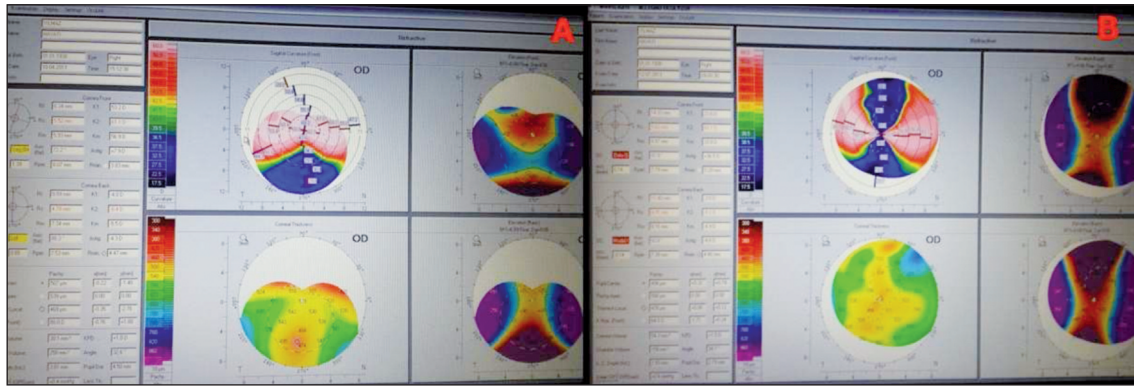


FIGURE 1: A: Pentacam analyses before wedge resection. Image quality is low due to photophobia and blepharospasm due to severe allergic conjunctivitis. Steepening in the superior quadrant with increased anterior and posterior elevation is observed in the right eye; **B:** Pentacam analyses after wedge resection. Flattening in the superior quadrant is observed, when compared to the preoperative evaluation.

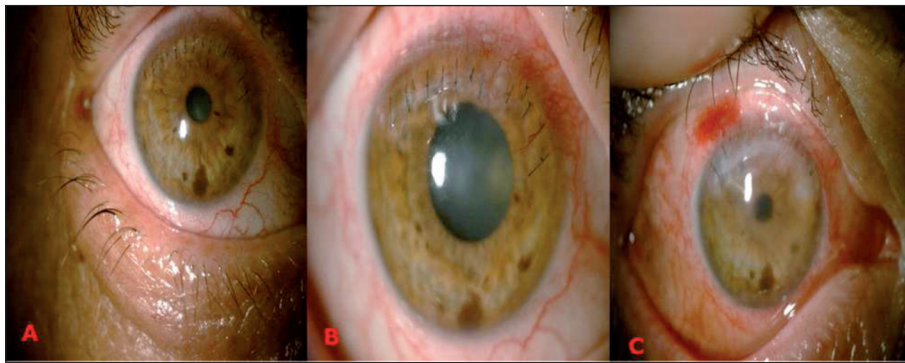


FIGURE 2: A: Post operative 1. day after wedge resection; **B:** Post operative 1. week after wedge resection. Scar formation and flattening are observed in the superior quadrant; **C:** After cataract surgery with tightened superior cornea due to scar formation.

though, PMD patients are typically poor candidates for penetrating keratoplasty because of peripheral thinning, surgery may be considered for patients whose vision is not adequately corrected by contact lenses when there is sufficient peripheral corneal thickness beyond the area of thinning.¹¹ A number of surgical procedures for the management of PMD have been described. These include crescentic wedge resection, crescentic lamellar keratoplasty, central penetrating keratoplasty (PK), oversized central PK, inferiorly decentered PK, epikeratophakia, and thermokeratoplasty.^{4,12}

In addition to PMD, our patient also had a grade 3 nuclear cataract which reduced the visual acuity. The main problem in this case was to obtain a reliable keratometry to calculate the intraocular lens power. Because of the irregular

astigmatism and cataract, spectacle correction would be ineffective and the other option may be rigid gas permeable contact lenses, however our 75 year old contact lens intolerant patient, the severity of the disease and our inability to obtain biometric measurements before cataract surgery made us to choose a surgical option.⁴ Therefore, we first planned crescent like wedge resection in order to treat the PMD zone, then we obtained a reliable keratometry to calculate intraocular lens power and performed phacoemulsification.

PMD can be managed with soft or rigid contact lenses. However, in advanced cases it is difficult to fit contact lenses. Therefore, surgical treatment may be required such as wedge resection, lamellar crescent resection, intracorneal rings or grafting techniques like lamellar or penetrating

keratoplasty.^{1,13} We performed lamellar crescent wedge resection and primary suturation to avoid the risks of penetrating keratoplasty.

Corneal crescent wedge resection technique allows us to treat peripheral corneal thinning by removing the abnormal ectatic tissue and retaining the host cornea without the need to introduce allograft tissue.¹⁴ This technique successfully improved corneal shape and astigmatism, with a resultant improvement and allowed us to obtain biometric measurements before cataract surgery. To the best of our knowledge, this is the first case in the literature having a superiorly located PMD accompanied with cataract, treated with lamellar crescent wedge resection.

In conclusion, lamellar crescent resection and primary suturation seems to be an effective way of treating pellucid marginal degeneration.

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Conflict of Interest

No conflicts of interest between the authors and / or family members of the scientific and medical committee members or members of the potential conflicts of interest, counseling, expertise, working conditions, share holding and similar situations in any firm.

Authorship Contributions

Onur Gokmen: Contributed to design of the work, data collection, data analysis and interpretation and drafting the article; Dilek Dursun: Altinors contributed to critical revision of the article and the final approval of the version to be published.

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