

# Superior marginal pellucid degeneration

## *Degeneração marginal pelúcida superior*

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### ABSTRACT

*Pellucid marginal corneal degeneration (PMCD) is a rare bilateral noninflammatory ectatic peripheral corneal disorder, usually involving the inferior portion of the cornea, separated from limbus by an area of normal corneal thickness. There is protrusion of the cornea above the thinning area, resulting in high irregular astigmatism. Corneal Topography shows the characteristic, but not pathognomonic, image of a “butterfly wing”. A patient with superior PMCD - an atypical presentation of the disease - is presented, highlighting the importance of the inclusion of PMCD as a diagnostic hypothesis, in cases of non-inflammatory peripheral corneal thinning that does not compromise only the inferior corneal area.*

**Keywords:** Corneal diseases; Corneal topography; Corneal pachymetry; Case reports

### RESUMO

A degeneração marginal pelúcida (DMP) é uma ectasia corneana rara. Caracteriza-se por afinamento periférico, sem sinais inflamatórios, tipicamente inferior e bilateral, separado do limbo por área de espessura normal. Há protrusão da córnea acima da área de afinamento, resultando em elevado astigmatismo irregular. À topografia corneana, obtém-se a imagem característica, porém não patognomônica, em “asa de borboleta”. Relata-se, nesse caso, um paciente com DMP superior - apresentação atípica da doença - destacando a importância da inclusão da DMP como hipótese diagnóstica nos casos de afinamentos corneanos periféricos não inflamatórios, que não comprometam apenas a região inferior da córnea.

**Descritores:** Doenças da córnea; Topografia da córnea; Paquimetria corneana; Relatos de casos

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## INTRODUCTION

**P**ellucid marginal degeneration (PMD) is a rare idiopathic corneal ectasia. It is characterized by the peripheral, non-inflammatory and non-ulcerative thinning of the cornea;<sup>(1,2)</sup> as well as absence of opacity, neovascularization and deposit of corneal associated.<sup>(3)</sup> The thinning occurs, characteristically, on the lower area, within 4 to 8 hours, in a growing range of 1-2 mm, being separated from the corneoescleral limbus by area of normal thickness.<sup>(1,2)</sup> The corneal protrusion occurs above the thinning, in an area of normal thickness.

Usually, the involvement is bilateral and asymmetric. The incidence is higher in between the second and fifth decades of life, regardless of race or sex.<sup>(1,2)</sup> Up to now, there is no evidence of hereditary transmission.

The clinical status is characterized by the slow increase of low visual acuity due to the induction of irregular against-the-rule astigmatism.<sup>(4)</sup>

The topography shows an increase in the curvature in the inferior periphery, extending into the oblique horizontal hemi meridians, and the flattening of the vertical axis. That way, we can obtain the characteristic image, but not pathognomonic, of “butterfly wings”.<sup>(5)</sup>

The histological study demonstrates that the epithelium of the thinning region is presented intact, while the Bowman’s layer is missing or defective.<sup>(6)</sup> Despite the typical inferior presentation, there are reports of atypical cases with superior<sup>(7-9)</sup>, nasal<sup>(7,9)</sup>, temporal<sup>(9)</sup> thinning and all around the circle of the cornea.<sup>(10)</sup>

This work aims to report another atypical form of PMD, contributing for the description of such cases and for signaling the importance of the inclusion of the PMD as a diagnostic hypothesis in cases of peripheral inflammatory non corneal thinning that impairs not only the inferior region of the cornea.

## CASE REPORT

Male, 57 years, white, retired, was referred to our service of Keratoconus research, due to the significant astigmatism increase report in the previous year. He complained of low visual acuity for 5 years progressive, occasion in which initiated the use of glasses. Ophthalmologic and family background was denied and he presented systemic arterial hypertension comorbidity.

The best correction for visual acuity was 20/200 in the right eye (+0.50DE -5.50DC 80°) and 20/15 in the left eye (+0.75DE -5.50DC 95°).

In the previous biomicroscopy, it was possible to observe a clear cornea in both eyes, without evidence of inflammation, vascularization or epithelial changes, as well as the presence of superior thinning area in the right eye, in growing format, 2 mm from the limbus, extending from 11 to 2 hours (Figure 1); associated with protrusion without thinning on the inferior adjacent cornea. The left eye did not show detectable changes (Figure 2). The remainder of the previous segment, retina mapping and tonometry of applanation showed abnormalities in both eyes.

The corneal topography of both eyes showed irregular against-the-rule astigmatism, with “butterfly wings” pattern located in the superior side (Figure 3).

In the corneal tomography (Pentacam® - Oculus Optikgeräte, Wetzlar, Germany) of the right eye, the vertical Scheimpflug image

showed superior band thinning, regular corneal thickness below and above it, the protrusion under the thinning and the central corneal flattening (Figure 4). The left eye remained unchanged.

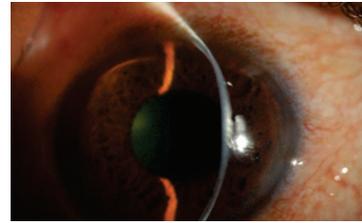


Figure 1: Peripheral thinning in the right eye.

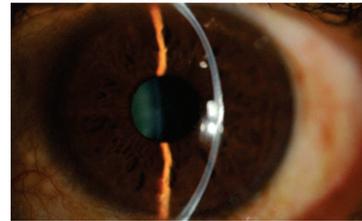


Figure 2: Normal corneal thickness in the left eye.

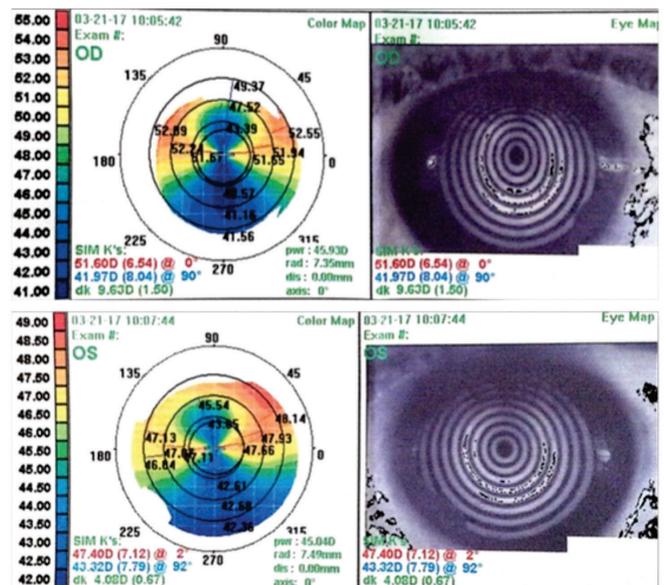


Figure 3: Topography with butterfly wing pattern in both eyes.

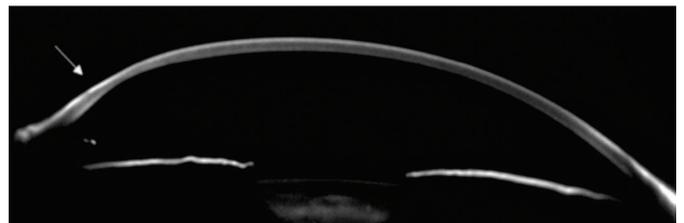
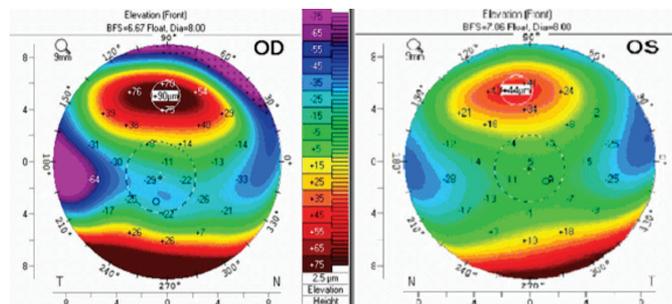


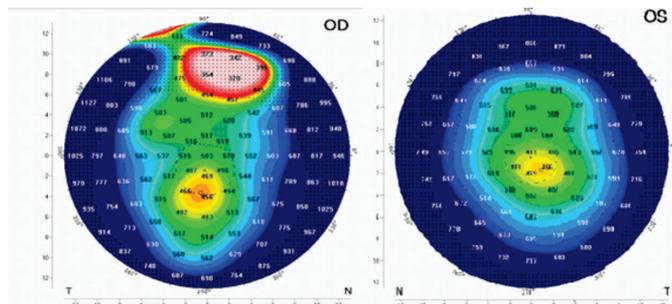
Figure 4: Image of vertical Scheimpflug (91-271o) with superior peripheral thinning (arrow signal).

In the previous elevation map compared to the best-fit sphere (BFS), we can observe if the peripheral superior elevation, corresponding to the ectasia, in both eyes. (Figure 5)



**Figure 5:** Previous elevation map compared to the best-fit sphere(8 mm).

In pachymetric map, a superior peripheral thinning area in the right eye was identified, which does not appear in the left eye. (Figures 6).



**Figure 6:** Pachymetric map (12 mm).

After the correlation of the physical examination with additional tests, it was established the diagnosis of PMD superior.

As an initial treatment, it was proposed the adaptation to rigid gas permeable contact lenses in his right eye. The best visual acuity reached 20/50, with important superior decentring of the contact lens and discomfort complaint.

Due to this failure, scleral contact lenses became an option, obtaining a satisfactory result as the standard adaptation to the following parameters: base curve 6.75 mm; -6.25 diopters; 16.50 diameter; 9.5 ZO and 4.98 SAG. The visual acuity reached 20/20, associated with the patient satisfaction concerning comfort.

## DISCUSSION

The differential diagnosis of corneal thinning in peripheral can prove challenging, especially in atypical cases, but is of fundamental importance for correct therapeutic approach. (11,12)

The initial difference between the inflammatory or non inflammatory causes is necessary. Among the inflammatory causes, we can emphasize keratitis (autoimmune, marginal staphylococcal, caused by rosacea and by exposure) and Mooren’s ulcer. In non inflammatory scenarios, the main differential diagnoses are Dellen, Keratoconus, keratoglobus, Terrien’s marginal degeneration and PMD. (13)

Keratoconus is the most common corneal ectasia(2) and the main differential diagnosis of PMD. Generally, it begins during puberty and proceeds until the third or fourth decades of life, (14) while the PMD begins later and has a slow progressive course.(8)

The main data to be analyzed for the distinction between the two diseases is the feature of corneal thinning; while in PMD it is peripheral and does not match the ectasia area, in Keratoconus, it appears as central or paracentral and coincides with the protrusion. (12,14) In moderate cases, such evaluation may be performed on previous biomicroscopy; however, in cases of incipient PMD, one must resort to additional assessment, as elevation maps and pachymetric.

Besides that, Keratoconus can present commemoratives, such as Fleischer’s ring, Vogt’s striae, Rizutti’s phenomenon and Munson’s sign,(14)missing in the PMD.

The keratoglobus constitutes another important differential diagnosis of PMD and it is distinguished by presenting its beginning at birth, diffuse thinning of the cornea, classic globoid protrusion and astigmatism without major irregularities. (1,15)

It is important to remember that, at the moment, it is not known whether the PMD, the Keratoconus and the keratoglobus are different pathologies or if they represent phenotypic variations of the same disorder. (1,16) Reports from the PMD association with Keratoconus(9) and keratoglobus(15,17) reinforce the idea that there may present different clinical aspects of the pathology.(16, 18,19)

Terrien’s marginal degeneration, on its turn, affects the age group similar to PMD and causes the increase of astigmatism. It differs by the presence of corneal opacity, axial lipid deposits to the thinning and presence of neovascularization. (1,14). However, attention must be paid to the fact that rare cases of hydrops on PMD are described and may generate scars and new vessels.(1,17,20)

So far, there are no specific and universally accepted topographic ratings for the diagnosis of PMD. (21) It is crucial to have in mind that the “butterfly wings” pattern in the topography does not define the pathology and it may appear in other corneal ectasias, including Keratoconus. This happens because the topographic patterns are artifacts of the representation method of the data and do not describe accurately the shape of the cornea. (5) So, for a correct diagnosis, it is essential that a detailed analysis on the slit lamp and pachymetry, as ways of highlighting the characteristic peripheral thinning.(22)

The vast majority of PMD cases can be properly handled only with clinical treatment. (12,23) In early stages, the use of glasses is an option. However, with the progression, the correction with glasses is usually not satisfactory due to high irregular astigmatism. (2)

Moderate cases commonly require gas permeable rigid contact lenses, which correct the vision adequately, but its adaption is difficult because of the unstable centration. An option for better stabilization and tolerance of the lens would be the choice of larger diameters.(17) Another option are bitoric lenses, indicated for oblated corneas. (21, 24) In more advanced cases, scleral lenses can be an option. (17)

Surgical options are considered in cases where there are no improvements with the clinical weaponry and they should be postponed as long as possible, once they present technical difficulties, unpredictability, irreversibility, long rehabilitation and higher rate of complications.(24-26) There are several surgical options available, but there is no consensus on which one would be the most effective.(4) Among them, there appears the penetrating keratoplasty,(1) the lamellar keratoplasty in ascending,(27) the

“tuck in” lamellar keratoplasty (“tuck in lamellar keratoplasty”-TILK),<sup>(25)</sup> the lamellar wedge resection,<sup>(27)</sup> intrastromal corneal ring<sup>(26-28)</sup> and the crosslinking.<sup>(27,29)</sup>

## REFERENCES

- Krachmer JH. Pellucid marginal corneal degeneration. *Arch Ophthalmol*. 1978; 96(4):1217–21.
- Krachmer JH, Feder, RS, Belin, MW. Keratoconus and related noninflammatory corneal thinning disorders. *Surv Ophthalmol*. 1984; 28(4):293-322.
- Schlaeppli V. La dystrophie marginale inferieure pellucide de la cornee. *Probl Actuels Ophthalmol*. 1957; 1: 672-7.
- Jinabhai A, Radhakrishnan H, O'Donnell C. Pellucid corneal marginal degeneration: A review. *Cont Lens Anterior Eye*. 2011; 34(2):56-63.
- Lee BW, Jurkunas UV, Harissi-Dagher M, Poothullil AM, Tobaigy FM, Azar DT. Ectatic disorders associated with a claw-shaped pattern on corneal topography. *Ophthalmology*. 2007; 144(1): 154-56.
- Parker D, McDonnell P, Barraquer J, Green W. Pellucid marginal corneal degeneration. *Cornea*. 1986; 5(2):115–23.
- Bower KS, Dhaliwal DK, Barnhorst Jr. DA, Warnicke J. Pellucid Marginal Degeneration with Superior Corneal Thinning. *Cornea*. 1997; 16(4): 483-85.
- Sridhar MS, Mahesh S, Bansal AK, Rao GN. Superior pellucid marginal degeneration. *Eye (Lond)*. 2004; 18(4): 393-99.
- Rao SK, Fogla R, Padmanabhan P, Prema M, Sitalakshmi G. Corneal topography in atypical pellucid marginal degeneration. *Cornea*. 1999; 18(3):265–72.
- Santos FX, Mocelin S, Machado R, Santos FHX, Sousa LB. Apresentação atípica de degeneração marginal pelúcida: relato de caso. *Arq Bras Oftalmol*. 2005; 68(6): 833-6.
- Kaushik S, Jain AK, Saini JS. Unilateral pellucid marginal degeneration. *Eye (Lond)*. 2003; 17(2):246–8.
- Tzelikis P, Cohen EJ, Rapuano C, Hammersmith K, Laibson PR. Management of pellucid marginal corneal degeneration. *Cornea*. 2005; 24(5):555–60.
- Garg P, Sangwan VS. Mooren's ulcer. In: Krachmer JH, Mannis MJ, Holland EJ, editors. *Cornea: fundamentals, diagnosis and management*. 3rd ed. St Louis: Mosby-Elsevier; 2011.
- Rabinowitz YS. Keratoconus. *Surv Ophthalmol*. 1998; 42(4):297–319.
- Karabatsas CH, Cook SD. Topographic analysis in pellucid marginal corneal degeneration and keratoglobus. *Eye (Lond)*. 1996; 10(4): 451–5.
- Sridhar M, Mahesh S, Bansal AK, Nutheti R, Rao G. Pellucid marginal corneal degeneration. *Ophthalmology*. 2004; 111(6): 1102–7.
- Biswas S, Brahma A, Tromans C, Ridgway A. Management of pellucid marginal corneal degeneration. *Eye (Lond)*. 2000; 14(Pt 4): 629-34.
- Kayazawa F, Nishimura K, Kodama Y, Tsuji T, Itoi M. Keratoconus with pellucid marginal corneal degeneration. *Arch Ophthalmol*. 1990; 109(4):486-7.
- Varley GA, Macsai MS, Krachmer JH. The results of penetrating keratoplasty for pellucid marginal corneal degeneration. *Am J Ophthalmol*. 1990; 110(2):149–52.
- Taglia DP, Sugar J. Superior pellucid marginal degeneration with hydrops. *Arch Ophthalmol*. 1997; 115(2):274-5.
- Tummanapalli SS, Maseedupally V, Mandathara P, Rathi VM, Sangwan VS. Evaluation of corneal elevation and thickness indices in pellucid marginal degeneration and keratoconus. *J Cataract Refract Surg*. 2013; 39(1):56-65.
- Belin MW, Asota IM, Ambrosio R, Khachikian SS. What's in a name: keratoconus, pellucid marginal degeneration, and related thinning disorders. *Ophthalmology*. 2011; 152(2):157-62.
- Kompella VB, Aasuri MK, Rao GN. Management of pellucid marginal corneal degeneration with rigid gas permeable contact lenses. *CLAOJ*. 2002; 28(3):140–5.
- Gruenauer-Kloevekorn C, Fischer U, Kloevekorn-Norgall K, Duncker GI, et al. Pellucid marginal corneal degeneration: evaluation of the corneal surface and contact lens fitting. *Br J Ophthalmol*. 2006; 90(3): 318–23.
- Kaushal S, Jhanji V, Sharma N, Tandon R, Titaly JS, Vajpayee R. “Tuck In” Lamellar Keratoplasty (TILK) for corneal ectasias involving corneal periphery. *Br J Ophthalmol*. 2008; 92(2):286–90.
- Mularoni A, Torreggiani A, di Biase A, Laffi GL, Tassinari G. Conservative treatment of early and moderate pellucid marginal degeneration: a new refractive approach with intracorneal rings. *Ophthalmology*. 2005; 112(4):660–6.
- Moshirfar M, Edmonds JN, Behunin NL, Christiansen SM. Current options in the management of pellucid marginal degeneration. *J Refract Surg*. 2014; 30(7):474-85.
- Jabbarvand M, Hashemian H, Khodaparast M, Bazvand F, Beheshtnejad. Outcome of complete intrastromal ring implantation using femtosecond laser in pellucid marginal degeneration. *Eye (Lond)*. 2015; 29(6):783-90.
- Hassan Z, Nemeth G, Modis L, Szalai E, Berta A. Collagen cross-linking in the treatment of pellucid marginal degeneration. *Indian J Ophthalmol*. 2014; 62(3):367-70.

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