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Unilateral Pellucid Marginal Degeneration: A Case Report

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Resume

Introduction

Pellucid marginal corneal degeneration (PMCD) is a progressive, non-inflammatory disease of the lower cornea. It is often confused with keratoconus because of clinical, histological and topographical similarities.
The aim of our work is to study the clinical and topographic characteristics of the disease, its differential diagnosis and therapeutic strategies through a unilateral case and a review of the literature.

Methods

We report the case of a 37-year-old female patient, without any particular history, who presented a progressive decrease in visual acuity of the right eye without any notion of redness or associated pain.

Results

The clinical examination found a Best corrected visual acuity of 4/20 in the right eye (OD) and 20/20 in the left eye (OS). In biomicroscopy, we observed a band of cornea thinned inferiorly, separated from the limbus by a crescent of normal cornea in the right eye. The cornea of the adelphic eye is normal.
Topography of the right eye showed corneal thinning that predominated inferiorly on the pachymetric map.The elevation maps objectified a corneal protrusion above the thinning area. On the axial map, we noted flattening of the upper vertical hemimeridians and camber of the oblique and lower vertical hemimeridians. Anterior astigmatism of -8.5 D at 107° was noted on the OD, and -0.4 D at 142.5° on the OG. In contrast, the left eye had a normal topographic appearance. As a treatment, scleral lens placement in the right eye was proposed.

Discussion

The diagnosis of the disease is initially clinical, where functional signs include decreased visual acuity or worsening of significant irregular astigmatism, and biomicroscopy shows a thinned inferior corneal band. Topography confirms the diagnosis, as well as distinguishes MPD from other non-inflammatory ectatic diseases of the horn such as keratoconus, posterior keratoconus by classically showing a very peripheral inferior ectasia with a "butterfly wing" appearance. The treatment is based, in early stages, on glasses, lenses or intracorneal rings. In advanced stages, more invasive treatments are necessary, such as corneal resections and transplants.

Conclusion

Pellucid marginal corneal degeneration is a rare idiopathic non-inflammatory ectatic corneal disease characterized by a narrow band of corneal. Typically a bilateral condition, but iit can be rarely unilateral. The corneal topography is a key fadiagnosis

Keywords :*pellucid marginal degeneresence , corneal topography, pachymetry*

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I. Introduction

Pellucid marginal corneal degeneration (PMCD) is a progressive ectasizing non-inflammatory disease affecting the lower part of the cornea. It is often confused with keratoconus for clinical, histological and topographical similarities.

The aim of our work is to study the clinical and topographical characteristics of the disease, its differential diagnosis and its therapeutic strategies through an atypical unilateral observation and a review of the literature.

II. Observations:

We report the case of a 37-year-old patient with no particular history who presented to ophthalmology for a progressive decrease in visual acuity in the right eye without any notion of redness or associated pain. Clinical examination found Bestvisual acuity corrected (BCVA) to 4/20 With -1.00 (-12.5 to 107 °) in the right eye (OD) and 20/20 with -0.50 (-0.5D to 142 °) in the left eye (OS). In biomicroscopy, the left eye had shown a normal appearance and regularity, without protruding stigmata. But on the right eye, an irregular outline with a

thin clear strip below the cornea, separated from the limbus by a normal thinning corneal edge, without obvious fisherman's ring, lipid disposition, or vascularization.

The corneal topography (TOPCON CA-800 corneal analyzer) of the right eye showed a bowtie-shaped with astigmatism measuring -8.5 D at 107 °, with corneal thinning in the low position on the map pachymetric. The elevation maps showed a corneal protrusion above the thinning zone. On the axial map, we noted a flattening of the upper vertical hemimeridians and a camber of the oblique and lower vertical hemimeridians. In contrast, the left eye presented a normal topographic appearance, with an astigmatism measuring -0.4 D at 142.5 °. This aspect discloses a PMCD of the right eye.

The intraocular pressure measured by the pneumotometer was 12 mm Hg in the right eye and 14 mm Hg in the left eye. And examination of the dilated fundus revealed no abnormalities in either eye

As a treatment, our patient was equipped with scleral glass, with visual acuity received 16/20, the patient refused any surgery procedures

III. Discussion

Pellucid marginal corneal degeneration is a bilateral, non-inflammatory, non-ulcerative, progressive ectacic corneal disease (1) it progresses very slowly to become symptomatic between the 3rd and 5th decade, with extreme cases diagnosed after the age of 60 years (2-3). the etiology of this rare condition remains unknown, but some studies postulate its association with progressive connective tissue disease (4,5).

The characteristic sign of PMD is the inferior peripheral thinning band, following stromal tissue loss of up to 80% in severe forms (6). Respecting an area of 1.0 to 2.0 mm of the lower limbus, without scar, or lipid deposition or vascularization(6-7)

Classically, there is a flattening of the vertical meridian above the band of thinning, This condition generates on topographic analysis as in our case characteristic bow-tie appearance of marked "against-the-rule" astigmatism Without a peripheral steeping (8), with the 'crab-clow' pattern or 'butterfly 'at the anterior curvature map, which may be in common with the subtype of keratoconus denominated in some series as "pellucid-like keratoconus" (PLK) (9)

Biomicroscopy assessment alone generally allows the distinction between PMD and corneal disorders of inflammatory origin such as mooren ulcer (10) advanced stage Terrien marginal degeneration (11).hawovere, the differentiation between PMD and keratoconus is extremely complicated especially with the pellucid -like-keratoconus (PLK) which can be easily mis-diagnosed as a true PMD in the clinical practice (9).Hence the interest and the usefulness of the new scheimpflug imaging-based devices of patchymetric map for an appropriate diagnosis of PMD.

Although PMCD has classically been described as bilateral. As in our case, some authors have described unilateral forms which could be an atypical asymmetry in time and probably on the degree , requiring prolonged regular control(12), Atypical locations have also been reported, including 3 cases of superior PMDCD and one On nasal location (13), as well as associations with dermatological conditions particularly scleroderma (5), and the ictyosis vulgaris (14)

Several surgeries procedures could be used to improve visual acuity when glasses and contact lenses do not provide adequate visual correction.

IV. Conclusion

Pellucid marginal corneal degeneration is a rare idiopathic non-inflammatory ,non ulcerative corneal disease characterized by a narrow band of corneal. It is a rare corneal disorder that shares many clinical characteristics with other corneal ectasias, such as keratoconus.

Typically a bilateral condition .it can be associated with other pathologies, as it can be rarely isolated, and unilateral. The corneal topography has a major interest in its diagnosis ans its follow-up.

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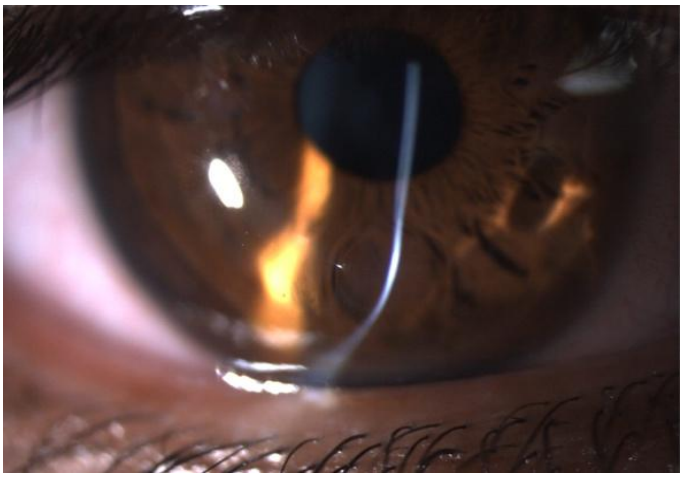

Figure 1: Slit view of the right eye of the patient showing the the pellucid marginal degeneration.$

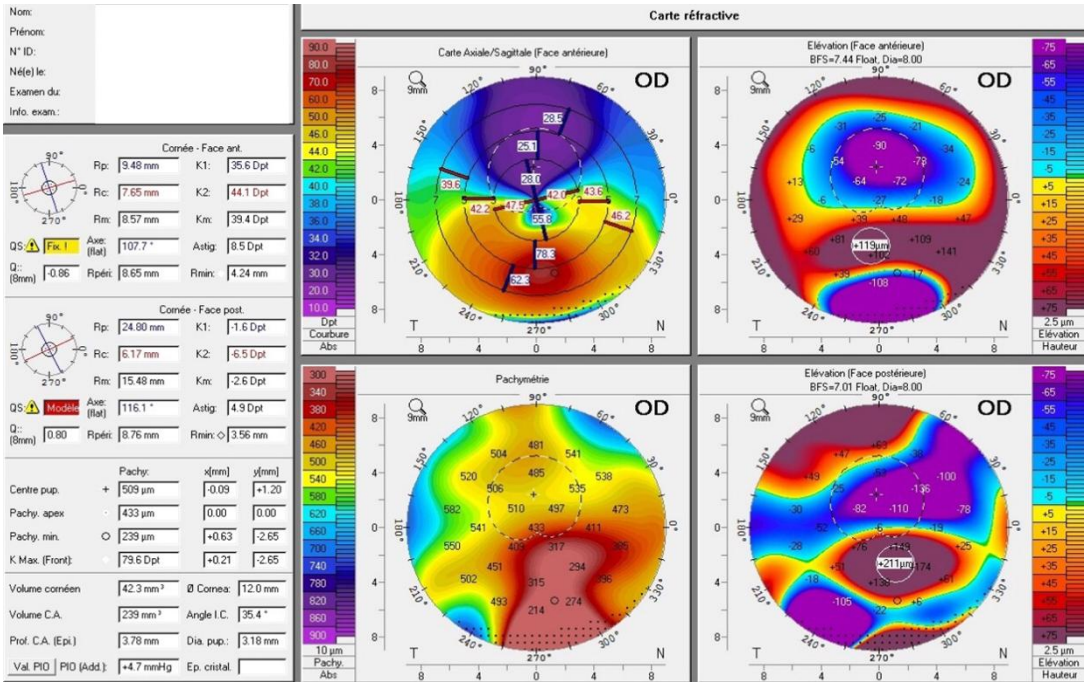

Fig2: corneal topography of the right reveling an aspect of pellicud marginal degeneration

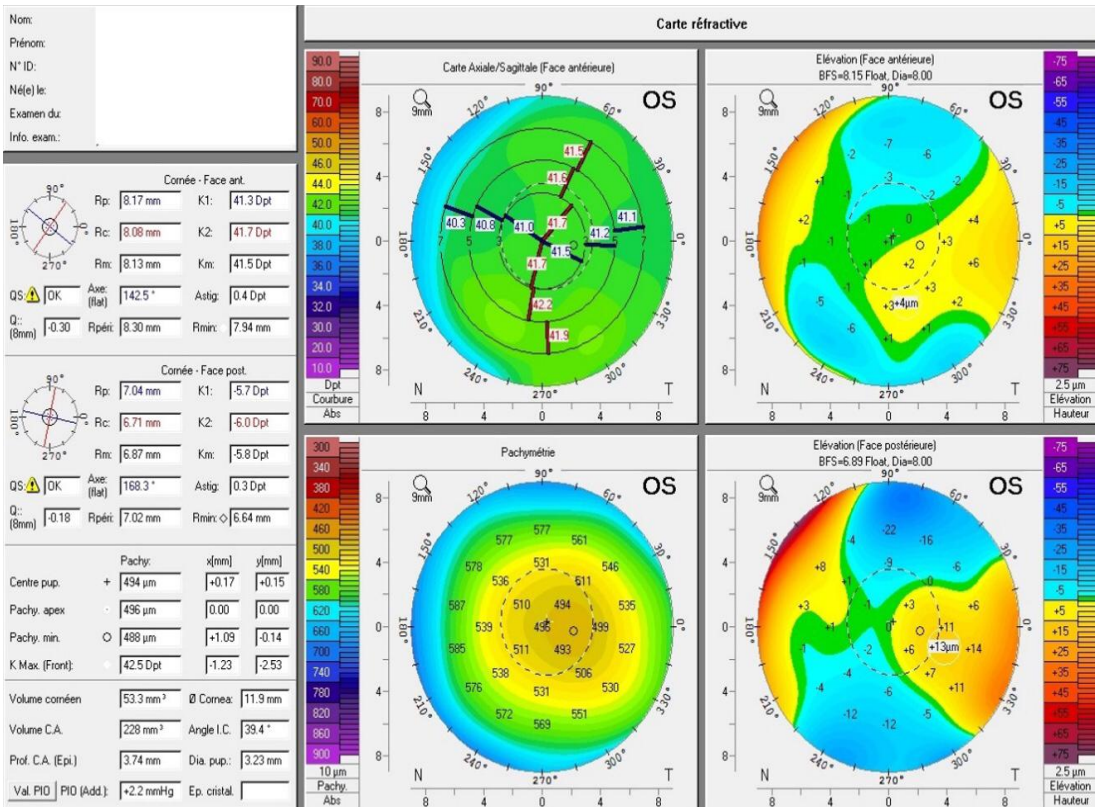

Fig3: normal corneal topography apprearence of the left eye

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