Progression of Pellucid Marginal Degeneration

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Background

- Pellucid marginal degeneration (PMD) is a rare disease that results in progressive peripheral corneal thinning and ectasia.[1]
- PMD is similar to keratoconus (KC), although tends to occur more peripherally and may be underestimated in prevalence.[2]
- This case highlights challenges of PMD diagnosis, prognosis, and management in an aging patient.

Case Presentation

The patient is an African American female, currently 54-years old. She initially presented in September 2017 and has been followed for the past 6 years. The following is a summary of her clinical findings.

Table 1. Pertinent clinical findings.

Year	2017	2020	2023
BCVA	OD: 20/25 OS: 20/25	OD: 20/30 OS: 20/50	OD: 20/20 OS: 20/50
IOP	NCT OD: 12 OS: 8	GAT OD: 15 OS: 15	iCare OD: 15 OS: 4
SLE	**OD**: No signs **OS**: No signs	**OD**: Gr 1 neo **OS**: Gr 1 neo; Vogt striae	**OD**: Gr 2 neo **OS**: Gr 2 neo; Vogt's striae
CL	Europa Toric scleral lenses	Europa Boston XO Center-Distance	sMap scleral lens fit – Europa Tangent
Dx	Presumed stable KC OS > OD. Function VA w/SL.	Progressive KC OS > OD. Monitor closely.	Progressive PMD OU. Refer for CXL consult
Other		Resolved sterile ulcer OS	Partial thickness lamellar hole OU

BCVA: Best Corrected Visual Acuity; IOP: Intraocular Pressure; SLE: Slit Lamp Exam; SL: Scleral Lens; CL: Contact Lens; NCT: Non-contact tonometry; GAT: Goldmann Applanation Tonometry

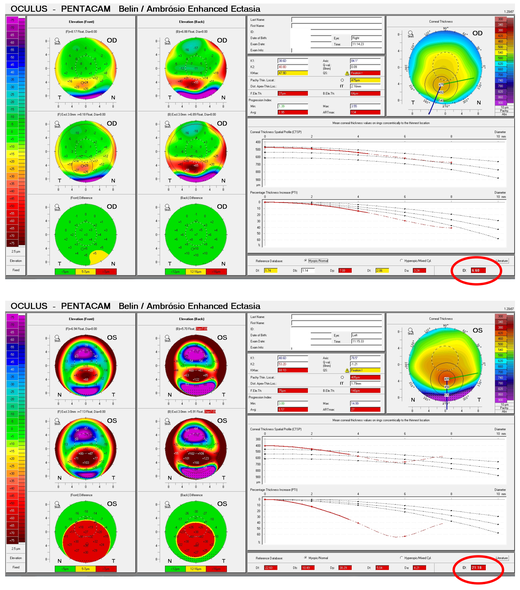

Figure 1. Oculus Pentacam Belin/Ambrosio enhanced ectasia display (BAD) of OD (top) and OS (bottom). Inferior displacement of the exclusion zone is observed OD>OS, although OS is a more severe disease state.

"D" scores are flagged in red OU indicating ectasia due to the significant deviation from the mean (red circle).

Corneal Tomography

Corneal tomography (Pentacam, Oculus) shows progressive PMD. Compared to baseline, Belin ABCD staging increases in severity OU and there is an increased number of flagged indices of irregularity OU.

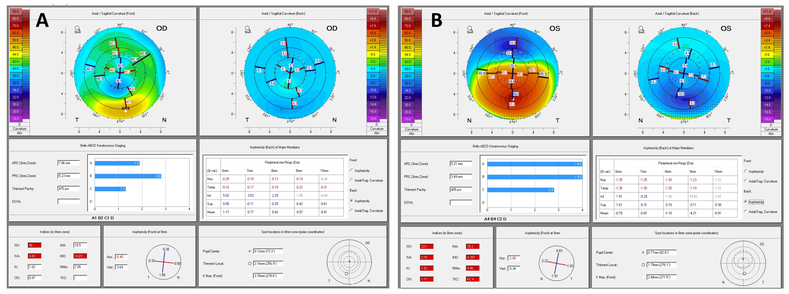

Figure 2. (2017) Corneal tomography images showing KC staging maps OD (A) and OS (B). Imaging shows a far inferior apex OD and a broader, more centralized cone OS.

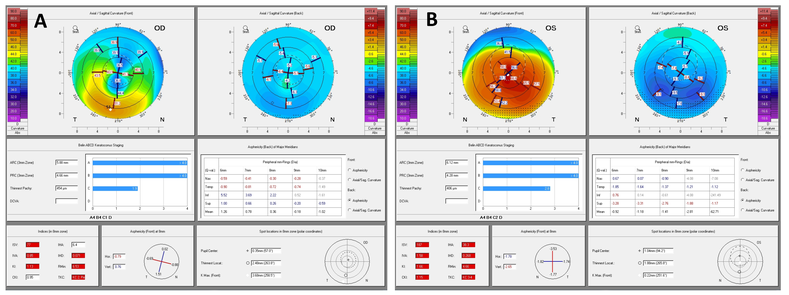

Figure 3. (2020) Corneal tomography images and KC staging OD (A) and OS (B). OD is becoming more indicative of PMD with inferior displacement of the apex visible.

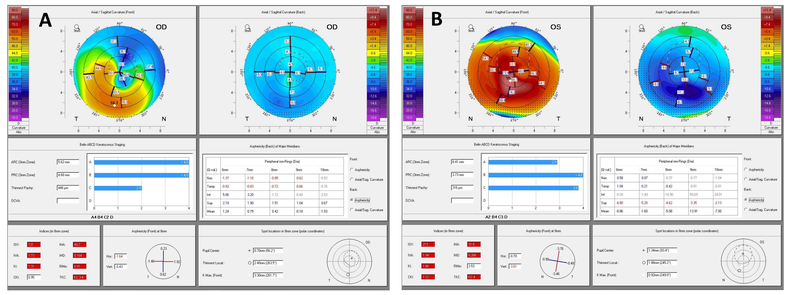

Figure 4. (2023) Corneal tomography images and KC staging OD (A) and OS (B). OD is following the characteristic "kissing-doves" pattern seen in PMD.

Table 2. Data collected from Pentacam imaging including maximum K values, thinnest pachymetry, and rate of change for thickness and dioptric measures. Indicative of significant progression OU.

YEAR	2017	2020	2023
Max K (D)	OD: 47.9 OS: 68.1	OD: 51.7 OS: 72.5	OD: 53.1 OS: 95.5
Min Pachs (um)	OD: 475 OS: 405	OD: 454 OS: 406	OD: 448 OS: 316
Per Year Change	Baseline	OD: 7 um; 1.3 D OS: 0.3 um; 1.5 D	OD: 4.5 um; 0.9 D OS: 14.8 um; 4.6 D

Discussion

- The risk of ectasia progression was overlooked in this patient due to her advanced age.
- The prognosis and treatment of PMD differs from KC in several ways:
  - PMD presents later in life (second to fifth decade) than KC (puberty to third decade).[3]
  - PMD progresses more slowly with less visual change.[4,5]
  - There is a lack of documentation and literature on progression of PMD.
  - Many insurance carriers do not specify "PMD" for medically necessary contact lens coverage.
- Early differentiation between PMD and KC is important due to different prognosis for progression
  - BAD map may be utilized to distinguish PMD from KC in earlier stages.
  - Exclusion zone of KC aligned centrally compared to inferior decentration with PMD.[6]
  - Be wary of placement of the exclusion zone when diagnosing KC.
- In this case, the right eye appeared to be clear and progressive with potential to benefit from corneal cross linking (CXL).
  - CXL not currently FDA approved for PMD, but studies show it may slow disease progression.[7]
- This also represents a case where bilateral macular holes were detected but may have been missed on previous exams due to the focus on the anterior segment.
  - The case belies the importance of comprehensive ocular health evaluation in patients with corneal disease.

Conclusions

- This case describes an atypical and severe progressive case of PMD in a patient with late onset of the disease. In the fifth decade of life, disease was onset and progressed to severe within 6 years of diagnosis.
- This case highlights the importance of careful diagnosis of corneal ectasias and assessment of progression in all demographics.
- This serves as a reminder to clinicians to carefully monitor progression at all visits and consider CXL regardless of age and suggests the need to better understand the differences between KC and PMD.

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