CASE REPORT

Pellucid-like keratoconus [v1; ref status: indexed, http://f1000r.es/TLhefH]

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Abstract

Purpose: To study the tomographic features of pellucid-like keratoconus (PLK), and to report a new sign on the pachymetry map (PM) in pellucid marginal degeneration (PMD).

Patients and methods: A retrospective descriptive case series was performed in Damascus University in 2011. Clinical and tomographic findings of 15 eyes (9 patients) that had the claw pattern of the anterior sagital map (ASM) were reviewed. Patients were distributed into two groups: (1) 4 eyes were considered PMD since they had inferior corneal thinning on both slitlamp biomicroscopy and PM; (2) 11 eyes were considered as PLK since they did not show inferior corneal thinning. Patients were studied using slitlamp biomicroscopy and Scheimpflug-based tomography (Pentacam HR). The ASM, anterior elevation map (AEM) and PM were analyzed and compared to study the “kissing birds” sign, the “bell” sign, and cone location.

Results: Patients’ average age was 25.93±8.05 (16–44 years). In total, 60% of patients were male. In group 1, the AEM in the best fit sphere (BFS) mode revealed no kissing birds sign, and the cone was central in 1 eye (25%) and paracentral in 3 eyes (75%). PM showed the bell sign in 4 eyes (100%). In group 2, the AEM in the BFS mode revealed the kissing birds sign in 2 eyes (18.2%), and the cone was central in 1 eye (9.1%), paracentral in 8 eyes (72.7%) and peripheral in 2 eyes (18.2%). PM didn’t show the bell sign in any eye.

Conclusion: The claw pattern on the ASM is not a hallmark of PMD; it can be seen in PLK. Cone location does not relate to diagnosis. The “bell” sign on the PM is a deferential diagnostic sign in PMD.

Article Status Summary

Referee Responses

Referees	1	2	3
v1 published 15 Nov 2012	✓ report 1	✗ report 4	✓ report 1

1 **Virender Sangwan**, Kallam Anji Reddy Campus India

2 **Michael W Belin**, University of Arizona USA

3 **Natalie Afshari**, Duke University School of Medicine USA

Latest Comments

No Comments Yet

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Introduction

Pellucid marginal degeneration (PMD) is an idiopathic, progressive, non-inflammatory, ectatic corneal disorder characterized by a peripheral inferior band of corneal thinning in a crescent-shaped pattern[1], although PMD cases with areas of superior thinning have been reported[2].

Similarities between PMD and keratoconus (KC) have led some ophthalmologists to consider PMD to be a peripheral form of KC[2,3]. Distinguishing between the two entities is of potential clinical importance since they differ markedly in prognosis and management. The management of PMD is unique since PMD is a progressive disease despite the fact that it is encountered in the third to fifth decade of life. Accordingly, corneal cross linking should still be one of the treatment options. When intracorneal rings (ICRs) implantation is indicated in the management of PMD, caution should be paid to the location of the inferior segment, since it passes through the inferior thinned area. Hence the need to calculate the depth of implantation depending on the thinnest point on the resumed passage, rather than on the thickness of the site of incision, in order to avoid deep corneal penetration.

In PMD, corneal tomographic analysis reveals a flattening in the vertical meridian, inducing a significant against-the-rule (ATR) astigmatism and a significant steepening around the area of maximum thinning[3]. This corneal configuration corresponds to a tomographic map that shows the classical claw pattern (Figure 1).

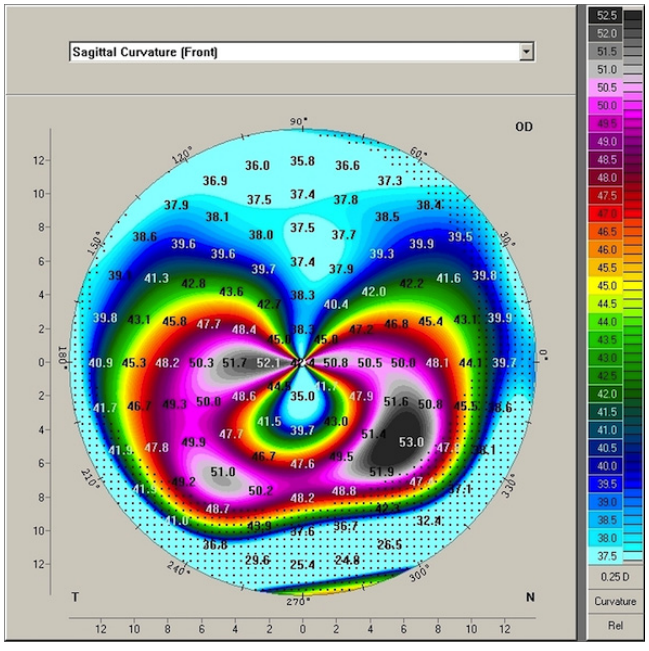

Figure 1. The crab claw or butterfly pattern on the anterior sagital curvature map. Notice the marked flattening of the cornea along the vertical meridian and the marked steepening of the inferior corneal periphery, which extends into the mid-peripheral inferior oblique corneal meridians associated with against-the-rule astigmatism.

Although corneal tomography is an important tool for the diagnosis of this corneal pathology, it should not be used as the only diagnostic criterion because it has been shown that this pattern is not always associated with the diagnosis of PMD; it might be seen with some other corneal ectatic disorders[4]. Therefore, pachymetric and biomicroscopic findings must also be considered for a reliable diagnosis.

In our study, we are reporting a "bell-shaped" sign on the pachymetry map (PM) in PMD which corresponds to the inferior thinning of the cornea observed by the slitlamp biomicroscopy. We are also reporting cases of KC with claw pattern on the sagital map but with neither the bell sign on the PM nor the inferior thinning on slitlamp biomicroscopy, identifying these cases to be "pellucid-like keratoconus (PLK)".

Patients and methods

A retrospective descriptive case series was performed in Damascus University in 2011. Clinical and tomographic findings of 15 eyes (9 patients) were reviewed and qualitatively analyzed. Inclusion criteria consisted of having a classic crab claw pattern on the anterior sagital curvature map (see Figure 1) taken by Pentacam® HR corneal tomographer (OCULUS Optikgeräte GmbH, Germany).

Patients were distributed into two groups based on clinical and tomographic findings. The diagnosis of PMD was based on both corneal tomography and clinical findings of peripheral corneal thinning in an arcuate or crescentic pattern on slitlamp biomicroscopy (Figure 2 and Figure 3). Therefore, group 1 was considered PMD; group 2 was given the name of "pellucid-like keratoconus (PLK)" since the inferior thinning was not observed in this group. The PMD group (group 1) consisted of 4 eyes that had the claw pattern on the anterior sagital map (ASM), and an inferior corneal thinning on slitlamp biomicroscopy. The PLK group (group 2) consisted of 11 eyes that had the claw pattern on the ASM, but without an inferior corneal thinning on slitlamp biomicroscopy.

Anterior sagital curvature, anterior elevation, and pachymetry maps were qualitatively analyzed and compared between groups 1 and 2. The anterior elevation map was studied using the best fit sphere

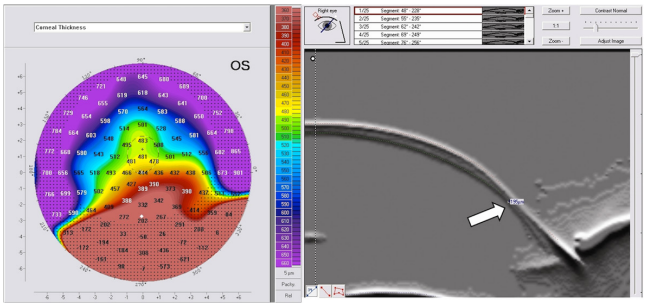

Figure 2. Inferior corneal thinning on corneal pachymetry in PMD. A: Bell sign on the thickness map, it is an indicator of inferior corneal thinning; compare values between the inferior part of this cornea with other peripheral parts. B: Scheimpflug image showing the thinning (white arrow).

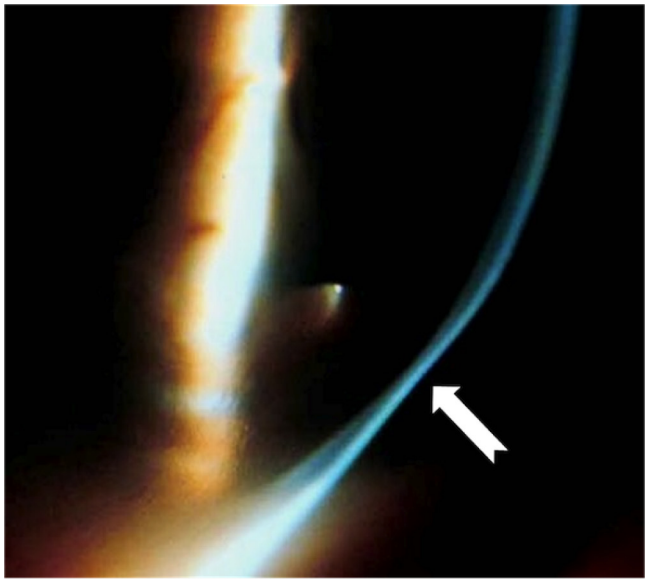

Figure 3. Inferior corneal thinning on Slitlamp biomicroscopy in PMD. Notice the abrupt narrowing of the slit beam in the area of inferior thinning (white arrow).

(BFS) float mode to localize the cone and to identify the kissing birds sign. The cone was considered central, paracentral or peripheral when the apex of the cone was within the central 3 mm zone, within 3–5 mm zone or out of the central 5 mm zone respectively (Figure 4).

Results

Patients' average age was 25.93±8.05 (16–44) years. A total of 60% of the patients were male. Average age of patients was 23±7.31 (16–33) years in group 1, and 27±8.03 (19–44) years in group 2.

Table 1 summarizes the results in both groups regarding the crab claw pattern, kissing birds sign, cone location, bell sign and inferior thinning on slitlamp biomicroscopy. In group 1, the cone was paracentral or central in 75% or 25% of cases respectively. Claw pattern, bell sign and inferior thinning were found in 100% of cases.

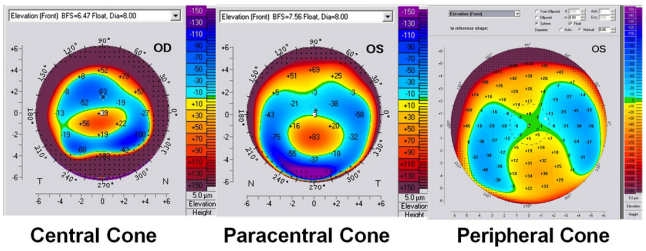

Figure 4. Classification of cone location on the anterior elevation map with the BFS float mode. The cone is central when its apex is within the 3 mm central circle, paracentral when it is in between 3 and 5 mm central circles, and peripheral when it is out of the 5 mm central circle.

In group 2, the kissing bird sign was present in 18.2% of cases, the cone was paracentral, peripheral or central in 72.7%, 18.2% or 9.1% of cases respectively. Bell sign and inferior thinning were absent.

Case report

A 27–year-old woman presented to the outpatient department with the complaint of blurred vision in both eyes. The uncorrected distance visual acuity (UDVA) was 0.7 (decimal) in the right eye (OD) and 0.2 (decimal) in the left eye (OS). On examination, the manifest refraction was -1.75D Cyl @ 65° OD, and -6.00D Cyl @105° OS. Corrected distance visual acuity (CDVA) was 1.0 (decimal) OD, and 0.7 (decimal) OS. Slitlamp biomicroscopy of the right eye revealed a clear cornea and normal features. The left eye showed inferior peripheral band of thinning extending from the 4 o'clock position to the 8 o'clock position. The intraocular pressure was normal in both eyes. Fundus examination revealed normal features. There were neither systemic nor local associations.

Corneal tomography (Figures 5a and b) revealed inferior perilimbal steepening with the crab claw pattern on the anterior sagital curvature map in both eyes. The anterior elevation map with BFS float mode revealed paracentral cones without the kissing birds sign in both eyes, the PM showed that the bell sign was present in the left eye and absent in the right eye. Therefore, diagnosis of PLK OD and PMD OS was made.

Table 1. Clinical and topographical findings in groups 1 and 2.

		Crab claw (anterior sagital map)	Kissing birds (anterior elevation map with BFS float mode)	Cone location (anterior elevation map with BFS float mode)			Bell sign (thickness map)	Inferior thinning on slitlamp
				Central	Paracentral	Peripheral		
Group 1: 4 eyes (PMD)	No. of eyes	4	0	1	3	0	4	4
	%	100	0	25	75	0	100	100
Group 2: 11 eyes (PLK)	No. of eyes	11	2	1	8	2	0	0
	%	100	18.2	9.1	72.7	18.2	0	0

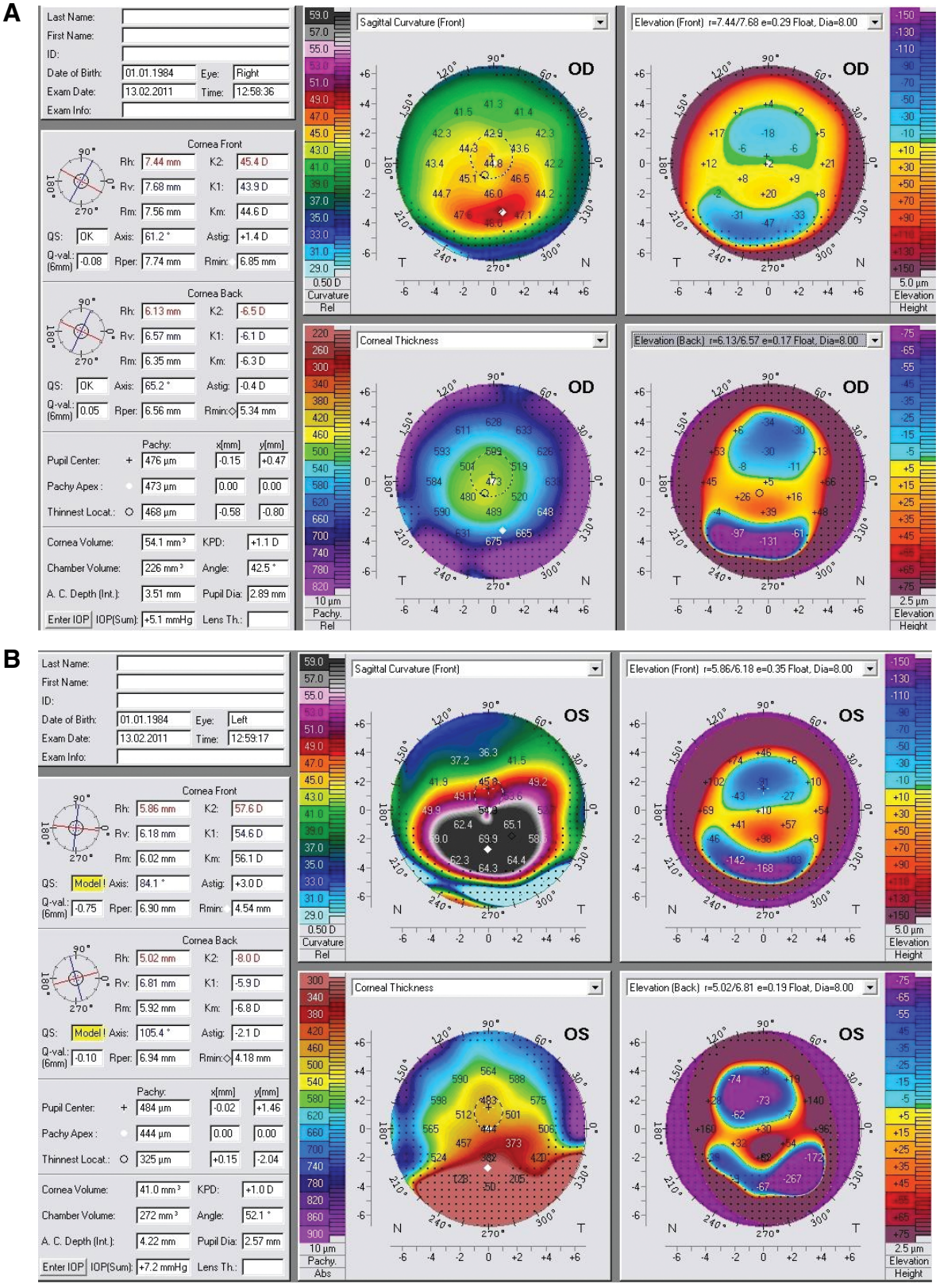

Figure 5a and b. A case with PMD in one eye and PLK in the other. (**A**): The right eye is PLK; notice the absence of the bell sign. (**B**): The left eye is PMD; notice the presence of this sign.

Discussion

In addition to careful clinical examination, corneal tomography has been recognized as an important and sensitive tool in detecting and managing ectatic corneal disorders, such as PMD and KC[5,6]. According to some reports, the typical sagital curvature map of the condition shows marked flattening of the cornea along a vertical axis and a steepening of the inferior cornea peripheral to the site of the lesion[3]. On the other hand, Lee BW et al. reported cases of ectatic corneal disorders with the same pattern[4]. Therefore, careful studying of corneal tomography with the main three maps is mandatory for diagnosing PMD and differentiating it from other ectatic corneal disorders. The main three maps consist of the anterior sagital curvature map, anterior elevation map and PM.

In our study, the crab claw pattern on the ASM was the inclusion criteria. Thereafter, cases were divided according to the presence or absence of the inferior corneal thinning on slitlamp biomicroscopy. Group 1 was identified as PMD when the thinning was present, and group 2 was identified as PLK when this sign was absent. We studied the other main maps qualitatively in both groups.

On the anterior elevation map, the kissing birds sign can be seen, and theoretically it should be always seen in PMD since it is a peripheral ectatic corneal disorder. In our study, we found that the presence of this sign was related to cone location. When the cone was peripheral, this sign was present; when the cone was central or paracentral, this sign was absent. This sign appeared when the BFS float mode was used, while it disappeared when switching to the best fit toric ellipsoid (BFTE) float mode, as shown in Figure 6. In group 1, the cone was central or paracentral in 100% of cases, and the kissing birds sign was absent in 100% of cases. In group 2, the cone was peripheral in 18.2% of cases, in which the kissing birds sign was present.

Cone location can be identified on the elevation maps or on the tangential curvature map, but not on the sagital curvature map[7]. In our study, we found no correlation between cone location and the differentiation between the two groups; i.e. the cone could be central, paracentral or peripheral in both entities. Therefore, neither the kissing birds sign nor the peripheral cone was a hallmark of PMD. Figure 7 is a PMD case without the kissing birds sign.

The PM showed thinning in PMD corneas towards the inferior part of the cornea, which was consistent with what was observed by the slitlamp biomicroscopy; a peripheral band of thinning of the inferior cornea from the 4 o'clock position to the 8 o'clock position and the light slit became very narrow abruptly in the inferior part of the cornea, which was the hallmark of the disease. This thinning was characterized with a special sign on the PM that can be called "bell" sign as shown in Figure 2. This sign was present in 100% of cases in group 1 and was absent in 100% of cases in group 2. Therefore, the hallmark of PMD was the bell sign on the PM, and this sign was due to inferior corneal thinning encountered in PMD.

On the other hand, there was no correlation between cone location and the thinnest area of the ectatic cornea. In group 1, although the bell sign was present and was an indicator of inferior thinning,

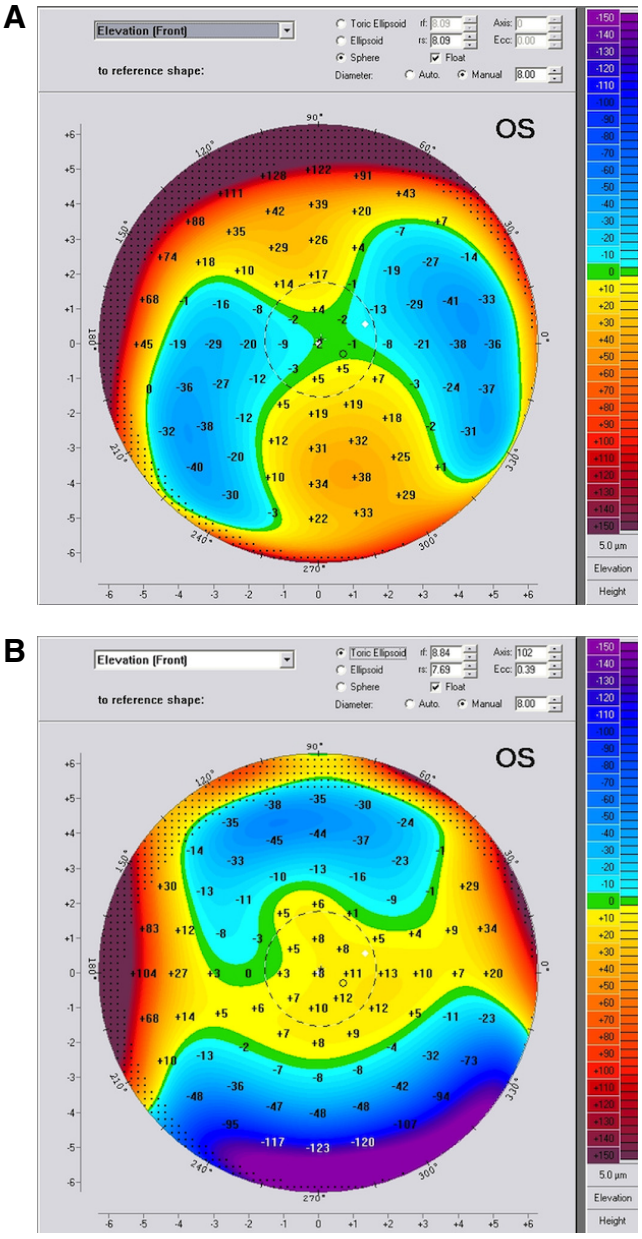

Figure 6. The kissing birds sign on the anterior elevation map. When it exists, it appears on the BFS float mode (**A**), and disappears when switching to the BFTE mode (**B**).

the cone was central or paracentral in 100% of cases. On the other hand, the bell sign was absent in 100% of cases in group 2 although there were 2 cases (18.2%) with peripheral inferior cones.

In regard to the location of the thinnest point of the cornea, theoretically, the thinnest point should be inferiorly displaced in both PMD and in KC, but the amount of displacement on the *Y* coordinate should be much larger in PMD, especially in advanced cases (Figure 8: white arrows). This is not always true. In very advanced PMD, the

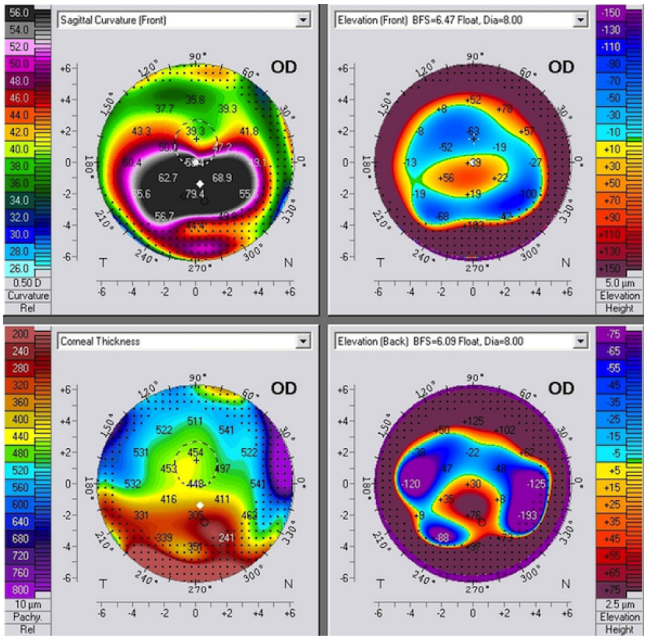

Figure 7. A PMD case without the kissing birds sign. Notice the bell sign on the corneal thickness map.

cornea is severely distorted, particularly in the inferior part of the cornea, and the tomographers may miss many data from this part, and extrapolate the area with dark dots indicating that an important part of the data was missing in this area, as shown in Figure 9.

It is well known in the literature that PMD usually starts later in life than KC: it presents in the third to fifth decade of life[3,5]. In our study, the mean age of presentation was the third decade in both PMD and PLK groups. There was one patient with bilateral PMD and his age was 16 years at presentation. In a series of 58 patients reported by Sridhar and Mahesh[8], there was an 8–year-old patient with PMD. Young ages were also seen in group 2; the age of the patients ranged from 19 to 44 years. Thus, patients' age is not an important issue when differentiating PMD from KC.

PMD and KC are bilateral ectatic corneal disorders, although they can be asymmetric in severity between both eyes[8]. Nevertheless, some studies reported the occurrence of PMD in one eye and KC in the fellow eye[9]. In our study, we reported a case with PMD in one eye and PLK in the fellow eye.

Regarding the case report, taking into account the clinical features, high ATR astigmatism, typical corneal tomography features and the presence of the bell sign on PM supported by peripheral corneal

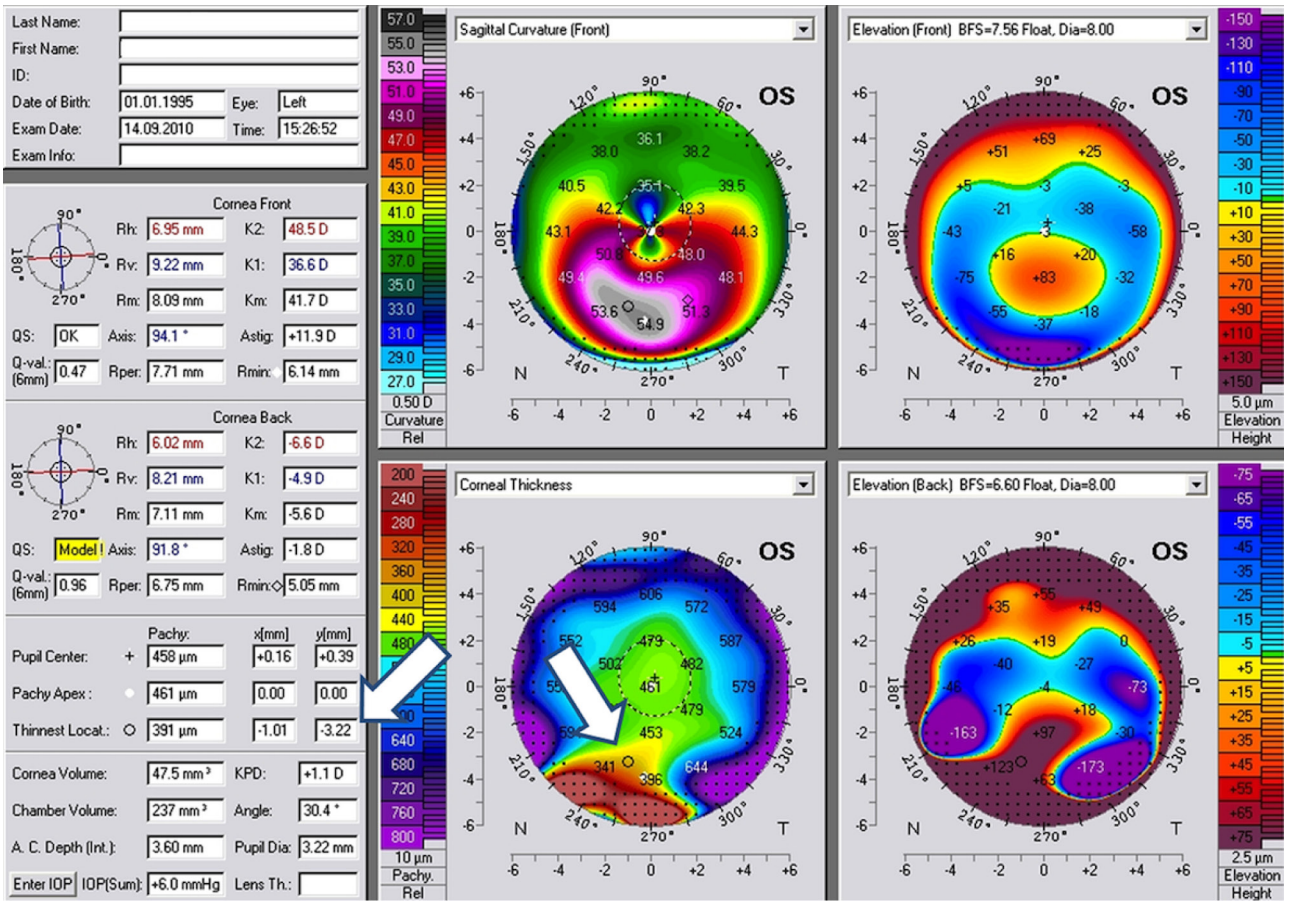

Figure 8. A case of severe PMD. The thinnest location is largely inferiorly displaced (white arrows).

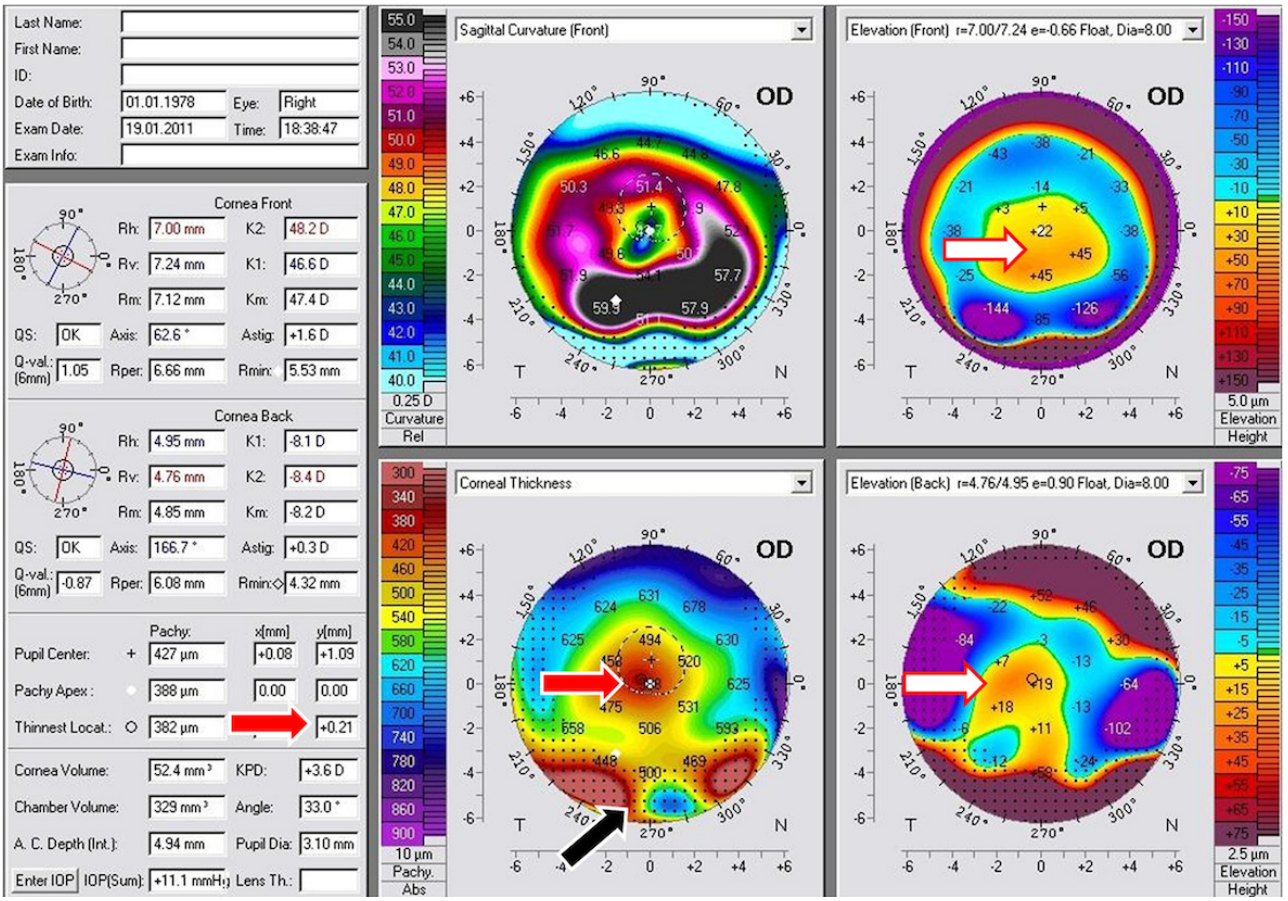

Figure 9. Corneal tomography of a severe PMD case with extrapolated data. The red arrows point to the thinnest location. The white arrows point to cone location. Black arrow points to a black dotted area suggesting that some data were missing in the inferior cornea due to severe corneal distortion.

thinning, a diagnosis of PMD was made in the left eye and PLK in the right eye.

In conclusion, we reported cases of KC mimicking the tomographical appearance of PMD, and we called that pellucid-like keratoconus (PLK). Moreover, identifying features of PMD on corneal tomography and slitlamp biomicroscopy is very important since there are some tomographical similarities between PMD and PLK, especially in early stages of the former which misguides doctors to misinterpret PLK as PMD. We also reported a sign on the PM in PMD, and we called it the "bell" sign.

Finally, the similarity between the two entities, in addition to what we found in the reported patient who has both entities, may suggest that PLK could represent a closely related disorder to PMD, just as Forme Fruste KC is to KC.

Author contributions

Mazen Sinjab contributed to the study concept and design, analysis and interpretation of data, critical revision of the manuscript, and provided statistical expertise and supervision. Lara Youssef contributed as a co-author, helping with data collection and drafting of the manuscript.

Competing interests

No competing interests were disclosed.

Grant information

The author(s) declared that no grants were involved in supporting this work.

Acknowledgements

The article was presented at the 2011 ESCRS conference in Vienna.

References

1. Rabinowitz YS: **Keratoconus.** *Surv Ophthalmol.* 1998; **42**(4): 297–319.
PubMed Abstract | Publisher Full Text

2. Sridhar MS, Mahesh S, Bansal AK, *et al.*: **Superior pellucid marginal corneal degeneration.** *Eye (Lond).* 2004; **18**(4): 393–9.
PubMed Abstract | Publisher Full Text

3. Maguire LJ, Klyce SD, McDonald MB, *et al.*: **Corneal topography of pellucid marginal degeneration.** *Ophthalmology.* 1987; **94**(5): 519–24.
PubMed Abstract

4. Lee BW, Jurkunas UV, Harissi-Dagher M, *et al.*: **Ectatic disorders associated with a claw-shaped pattern on corneal topography.** *Am J Ophthalmol.* 2007; **144**(1): 154–6.
PubMed Abstract

5. Karabatsas CH, Cook SD: **Topographic analysis in pellucid marginal corneal degeneration and keratoglobus.** *Eye (Lond).* 1996; **10**(Pt 4): 451–455.
PubMed Abstract | Publisher Full Text

6. Sridhar MS, Mahesh S, Bansal AK, *et al.*: **Pellucid marginal corneal degeneration.** *Ophthalmology.* 2004; **111**(6): 1102–1107.
PubMed Abstract | Publisher Full Text

7. Holladay JT: **Detecting Forme Fruste Keratoconus With the Pentacam.** Supplement to Cataract & Refractive Surgery Today 2008: 11–12.
Reference Source

8. Wagenhorst BB: **Unilateral pellucid marginal degeneration in an elderly patient.** *Br J Ophthalmol.* 1996; **80**(10): 927–8.
PubMed Abstract | Publisher Full Text | Free Full Text

9. Kayazawa F, Nishimura K, Kodama Y, *et al.*: **Keratoconus with pellucid marginal corneal degeneration.** *Arch Ophthalmol.* 1984; **102**(6): 895–6.
PubMed Abstract | Publisher Full Text

Current Referee Status:

Referee Responses for Version 1

Natalie Afshari, Department of Ophthalmology, Duke University School of Medicine, Durham, NC, USA

Approved: 27 November 2012

Ref Report: 27 November 2012
The authors of the case report introduce the idea of pellucid like keratoconus (KC). They report cases of KC with a claw pattern on the sagital map but with neither the bell sign on the Pellucid nor the inferior thinning on the slit-lamp biomicroscopy. This idea is new and therefore worthy of publication.

I have read this submission. I believe that I have an appropriate level of expertise to confirm that it is of an acceptable scientific standard.

Competing Interests: No competing interests were disclosed.

1 Comment

Author Response

Mazen Sinjab, Damascus University , Syria
Posted: 27 Nov 2012

Dear Professor Afshari,

During my past 16 years of practice as a refractive surgeon, I have seen more than 3000 cases of keratoconus and ectatic corneal disorders at my center and in the refractive clinic in the Damascus University hospital, and many of those cases were referred to me for a second opinion. This number seems to be large, but the cause behind it was that in Syria such diseases are relatively common due to close intermarriage, environment and to the high prevalence of vernal keratoconjunctivitis; this registers an incidence higher than the international one which is 1:2000, to be about 1:400 in Syria, and in some well known regions in Syria one can say 1:100. PMD consists of about 20% of the seen cases, and it is seen not only in adults but also in young patients, which gives the disease a more aggressive nature. The same can be said about KC, I have seen many cases with ages younger than 10 years old (the youngest was 4 years old!). Due to such a high number of cases that I have seen, I could recognize cases that lie in between KC and PMD although both entities have their own tomographic features; and that was the idea behind my study.
Competing Interests: No competing interests were disclosed.

Michael W Belin, Department of Ophthalmology, University of Arizona, Tucson, AZ, USA
Not Approved: 26 November 2012

Ref Report: 26 November 2012
The authors appear not to be aware of the current literature which clearly distinguishes the tomographic patterns of inferior keratoconus from Pellucid. The limitation of curvature analysis and the importance of the pachymetric map have been previously well documented. The authors reference list is outdated and lacks articles that clearly describe the tomographic findings of true Pellucid: Walker RN *et al.* (2008), Belin MW *et al.* (2011) and Belin MW *et al.* (2012).

The anterior curvature patters described as “crab claw” or many other descriptors are non-specific and highlight the limitations of curvature when trying to imply shape. Adding additional descriptors (e.g. bell shape) is counterproductive.

The authors use the term “tomographic” without making a distinction between anterior curvature (classic topography) and tomography which includes both anterior and posterior corneal elevation and corneal thickness maps.

The pachymetric “bell sign” is also a poor descriptor and is seen in this example because of the scale chosen. If the authors had chosen an appropriate scale that covered the full range of thickness, they would see the more classic inferior band that has been previously described and published.

The case report is fraught with mistakes. First the authors keep stating that the best-fit-sphere was used but the machine is set for a toric-ellipsoid. The pachymetric map has not reliable data inferior, which is why no thickness readings are produced. One cannot diagnose true PMD in this case.

All in all, the paper is poorly researched and the maps show a lack of understanding of the displays.

I have read this submission. I believe that I have an appropriate level of expertise to state that I do not consider it to be of an acceptable scientific standard, for reasons outlined above.

Competing Interests: I serve as a consultant to OCULUS GmbH which produced the Pentacam

4 Comments

Author Response

Mazen Sinjab, Damascus University , Syria
Posted: 12 Dec 2012

Dear Prof Belin, Thank you so much for your valuable comments.

Before I make any changes in my article, I would like to ask some questions. I have read the article “Scheimpflug Photographic Diagnosis of PellucidMarginal Degeneration” by Walker RN *et al.* (2008), I will refer to it as article number 1; and the article “What’s in a name: Keratoconus, Pellucid Marginal Degeneration and Related Thinning Disorders” by Belin MW *et al.* (2011), I will refer to it as article number 2. Unfortunately I could not have the third article which is “Simplified Nomenclature for Describing Keratoconus” by Belin MW *et al.* (2012). I would be very thankful if

you could provide me with it and it would be so kind of you.
The questions are:
1. In article 2: the author stated: “in our original review article PMD was described as a typically bilateral, clear, inferior, peripheral corneal thinning disorder, characterized by a narrow band of corneal thinning (often approaching 20% normal thickness) separated from the inferior limbus by a relatively uninvolved area 1 to 2 mm in width.” However, nothing was mentioned in this article and in article 1 about the new definition of PMD and whether it is bilateral or can be unilateral. So the question is: can PMD be unilateral?

2. In article 2 the author stated: “Ertan and Bahadir in 2006 described the use of the femtosecond laser and intrastromal rings to correct PMD. While no corneal thickness maps were presented, the posterior elevation maps were typical of KCN and not classic PMD.” But may I ask: what is typical of KCN and not classic PMD on the posterior elevation map? The same question about what the author mentioned about Kubaloglu and associates.

3. How do you define that the cone is central, paracentral or peripheral on the elevation map with the BFS mode? I mean when do say the the cone is central (within 3 mm, 4mm, 5mm….) since this was not mentioned in both articles?

4.In article 2, the author recommended that “extrapolated data should not be displayed” and thus the 9mm scale should be used, therefore according to this recommendation, the peripheral part of the cornea will not be displayed or estimated and the pachymetric changes of PMD will not be evaluated?! At the same time, the authors use the extrapolated areas in many of their talks during conferences and articles!

5. In article 1: how could the authors recognize the thinning in case of focal edema? And how could they depend on Placido or even Scheimpflug in this very distorted cornea? They also stated:” a bilateral superior flattening” in figure 4 but only OS was displayed; What about OD, is it possible to see the flattening on OD despite edema even if it was focal and inferior?

6. In article 1, page 2: the author stated:” thinning within 2 mm of the limbus, and a more normal corneal thickness inferior to the band of thinning.” But all of that lie in the extrapolated area which should not be displayed as the author recommended in article 2!

Sincerely Yours, Mazen
Competing Interests: No competing interests were disclosed.

Referee Response

Michael Belin, Universtiy of Arizona, USA
Posted: 13 Dec 2012

Dr. Sinjab, I sent you the requested article via this publisher.

In reference to the findings of KCN vs PMD on the posterior elevation, Inf KCN shows the classic positive islands of elevation when using the standard BFS (sphere). Elevation maps are not

overly useful in true PMD because the cornea is so flat in the central 8.0 mm zone that the reference surface is so flat as to not accentuate the pathology. This is why the pach map is the most important (opened up to show the full cornea)

As far as which is a central cone, vs paracentral, vs peripheral. I do not believe there is anything established but I use the central 3.0 for “central” and outside 6.0 for “inferior” but it is often just a visual description.

In general we limit our display to 9.0 mm as for routine screening this is easier. That is not the case for peripheral disease. In cases of true PMD, you have to show the entire cornea. It is true that the more peripheral the data the more likely it will be extrapolated but if the Pentacam produced a thickness reading it is useful for visual diagnosis. If the number is produced, but extrapolated, it means it did not meet the strict machine criteria. If no number (thickness) is produced then the map needs to be repeated as there is no useful data.

In article 1 the thinning was easily seen in the other eye as this was bilateral. The Scheimplfug images were still reliable in the eye with the hydrops except for the far periphery so the central and paracentral flattening could still be seen. The reason both eyes were not shown in all displays is a limit of the journal for case reports.

Competing Interests: No competing interests were disclosed.

Author Response

Mazen Sinjab, Damascus University , Syria
Posted: 13 Dec 2012

Thank you very much Prof. Belin, and many thank to this journal that gave me the opportunity to learn from you. You were so kind to send me the article.

But may I ask you: could PMD be unilateral?

Sincerely Yours, Mazen

Competing Interests: No competing interests were disclosed.

Referee Response

Michael Belin, Universtiy of Arizona, USA
Posted: 14 Dec 2012

UNILATERAL – I don't know. I would guess that if PMD (as is KCN) is ultimately a disorder of collagen (i.e. genetic / biochemical) that the collagen would be abnormal in both eyes though clinical presentations may be unilateral or highly asymmetic.

Competing Interests: No competing interests were disclosed.

Virender Sangwan, L V Prasad Eye Institute, Kallam Anji Reddy Campus, Hyderabad. Andhra Pradesh, India
Approved: 19 November 2012

Ref Report: 19 November 2012
This is a well done study with a very good documentation of the case.

I have read this submission. I believe that I have an appropriate level of expertise to confirm that it is of an acceptable scientific standard.

Competing Interests: No competing interests were disclosed.

1 Comment

Author Response

Mazen Sinjab, Damascus University , Syria
Posted: 19 Nov 2012

Thank you Virender for your endorsement.
Competing Interests: No competing interests were disclosed.