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Temporal keratoconus in a pediatric patient

Lyvia J. Zhang [a], Aisha S. Traish [a,b], Thomas H. Dohlman [a,b,*]

[a] Cornea Service, Massachusetts Eye and Ear, Department of Ophthalmology, Harvard Medical School, Boston, MA, USA
[b] Cornea Service, Boston Children's Hospital, Department of Ophthalmology, Harvard Medical School, Boston, MA, USA

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ABSTRACT

Purpose: To report a pediatric patient with bilateral temporal keratoconus.
Observations: A 14-year-old male presented with a two-year history of progressively worsening visual acuity in both eyes and suspicion for undiagnosed amblyopia in the right eye. Retinoscopy revealed a scissoring reflex in both eyes and corneal topography demonstrated high keratometry values (Kmax 57.9 D and 46.1 D in the right and left eyes, respectively), with relative temporal steepening approximately coinciding with the thinnest pachymetry in both eyes. Corneal cross-linking was recommended.
Conclusions and importance: Keratoconus can present as a temporal variant with relative temporal steepening and thinning. It is important to maintain a high index of suspicion for keratoconus in pediatric patients with subnormal visual acuities. Prompt assessment and diagnosis may prevent progression of keratoconus and development of amblyopia.

1. Introduction

Keratoconus is a condition characterized by degenerative thinning of the cornea that commonly develops in the second to third decades of life and progressively worsens over time.[1] Although it is generally an isolated condition, associations with other conditions including Down's syndrome, Leber's congenital amaurosis, Ehler's Danlos connective tissue disorders, Marfan's syndrome, and atopy have been reported.[1] The prevalence of keratoconus was reported to be 20.6 per 1000 in men and 18.33 per 1000 in women based on a 2020 meta-analysis.[2] The underlying etiology of keratoconus is poorly understood. Its onset is characterized by noninflammatory formation of a "cone" and irregular thinning of the corneal apex. Corneal ectasia may progress and be followed by inflammation and scarring of the corneal stroma.[3] Pediatric keratoconus is now more widely recognized and tends to progress more aggressively than in adults. This is generally attributed to rapid corneal collagen remodeling and may be associated with the higher incidence of ocular allergies in children.[4] Keratoconus is classically described as corneal steepening inferior to the visual axis. Topographically it generally originates as a symmetric bowtie pattern along the 90-degree axis and becomes an asymmetric bowtie with relatively greater inferior steepening.[5,6] Although inferior corneal steepening is classic, superior keratoconus has also been described.[7,8] Here, we describe a case of a pediatric patient with unusual temporal corneal thinning suggestive of 'temporal keratoconus', a presentation of keratoconus that, to our knowledge, has not been described to date.

2. Case report

A 14-year-old Portuguese-speaking male presented to optometry clinic with a two-year history of painless, progressively worsening visual acuity in the right eye. Due to his nonspecific presentation and lack of exam findings to this point, he had been diagnosed with amblyopia in the right eye and patching had not recommended due to his age. The patient wore prescription glasses and denied difficulty with performing his activities of daily living. He denied a history of ocular trauma, and his past medical history and family history were unremarkable. Of note, although he reported a history of rare eye-rubbing he had no history of allergy or atopy, and there was no family history of any ocular disease.

On examination, his best corrected visual acuity with current glasses was 20/70-2 in the right eye and 20/30-2 in the left eye (Table 1). Dilated cycloplegic refraction measured a best spectacle corrected visual acuity of 20/60 (Plano −7.00 x 080) in the right eye, and 20/30 (-0.75 -1.00 x085) in the left eye. Retinoscopy was performed with noted difficulty neutralizing the light reflex, along with a scissoring reflex in the right eye. Cover testing revealed a right exotropia. Slit lamp examination demonstrated a 'cone-like' shape to the patient's right cornea, with apical corneal thinning. The patient's progressive decrease in visual

* Corresponding author. Massachusetts Eye and Ear 243 Charles St, Boston, MA, 02114, USA.
E-mail address: Thomas_Dohlman@meei.harvard.edu (T.H. Dohlman).

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acuity, possible amblyopia in the right eye and scissoring on retinoscopy were concerning for a diagnosis of keratoconus. A contact lens fitting was scheduled in order to try and improve vision with a rigid gas permeable contact lens and the patient was referred to the Cornea service for further evaluation.

Upon presentation to Cornea clinic, the patient again reported difficulty seeing with his right eye. He denied eye pain, redness, itching, or eye rubbing. Slit lamp examination of the right eye revealed a conical appearance to the cornea and a nasal vertically oriented iron line. No Vogt's striae, apical scarring or frank Fleischer ring was appreciated. There was a subtle Munson's sign in the right eye localized to the temporal lower lid margin present on downward gaze (Supplemental Fig. 1). Slit lamp examination of the left eye revealed a clear cornea without Vogt's striae, apical scarring, or Fleischer ring. Corneal topography demonstrated asymmetric against-the-rule bow-tie astigmatism, with relative temporal steepening coinciding with the point of greatest corneal thinning in both eyes (Fig. 1). In the right eye, the Kmax was 57.9D, and the thinnest pachymetry was noted to be 450μm. There was 7.8 D of astigmatism with the steep axis at 175.1°. In the 3mm zone, the horizontal hemi-meridians of the bowtie-shaped apices showed K values of 51.6D (nasally) and 57.2D (temporally), with a difference of 5.6D (Table 1). Given these values and the striking bowtie-shaped astigmatism skewed temporally along the central visual axis on topography, the right eye was diagnosed with a temporal variant of keratoconus. In addition, the left eye showed signs of early keratoconus with relative temporal steepening, a Kmax of 46.1D and 0.3 D of astigmatism with the steep axis at 15.8° (Table 1). In the 3mm zone, the horizontal hemi-meridians of the bowtie-shaped apices showed K values of 42.4 D (nasally) and 45.6 D (temporally), with a difference of 3.9D (Table 1).

In summary, after over two years of decreased vision in both eyes, our patient was found to have relative temporal corneal steepening and thinning, with a Kmax of 57.9 D in the right eye and 46.1 D in the left eye. To prevent further progression of his keratoconus, corneal cross-linking was recommended in the right eye, to be followed by the left eye, as well as rigid gas permeable (RGP) and/or scleral contact lenses fitting. After discussing the risks and benefits of early intervention with corneal crosslinking, the patient elected to proceed with surgery.

3. Discussion

This report describes a 14-year-old male with a two-year history of relatively decreased vision in the right eye. The patient's exam findings of a scissoring reflex on retinoscopy, Munson's sign and conical-shaped cornea were all suggestive of a diagnosis of keratoconus. This diagnosis was confirmed upon performing corneal topography, which revealed asymmetric bow-tie, temporal corneal steepening coinciding with the point of greatest corneal thinning. Interestingly, rather than being vertical, the axis of astigmatism was horizontal, with greater steepening temporally. This latter finding confirmed a diagnosis of 'temporal' keratoconus. Keratoconus classically presents as a protruding corneal apex and area of steepening and thinning inferior to the central visual axis,[9,10] although cases of superior keratoconus have also been reported.[7,11] To our knowledge, no cases of temporal keratoconus have been reported in the literature to date.

Bilateral progressive thinning of the cornea in keratoconus results in irregular astigmatism and subsequent blurred vision. Normally the corneal collagen fibrils align predominantly in the superior-inferior and nasal-temporal axes. Compared to healthy eyes, keratoconus eyes contain fewer collagen fibrils as the corneal stroma thins. The relaxed corneal tissue lengthens as it cones outward, often leading to myopia and sub-normal visual acuity.[3,12] In patients in the amblyogenic age range, high astigmatism and/or keratoconus can lead to the development of amblyopia, as seen in the patient described here. Delayed workup and diagnosis compounds the problem by allowing progression of corneal pathology and protrusion, leading to poorer treatment outcomes.

The diagnosis of keratoconus is based on a combination of ocular history, slit lamp exam findings, keratometry, and characteristic patterns of corneal topography. Advanced imaging techniques such as the Pentacam, which was performed for our patient, offer 3-dimensional scans of the cornea with detailed analysis of the cone. The colored 'heatmap' on Pentacam offers a visual guide to the cornea's curvature. It can help readily locate the corneal apex and provide value in planning appropriate surgical interventions, although crosslinking is generally the most common intervention offered to pediatric patients (see below). The apex of the cone is most commonly characterized by the steepest keratometry measurement and overlies the thinnest location of the cornea in keratoconus. The apex is surrounded by concentric zones of decreasing steepness.[13,14]

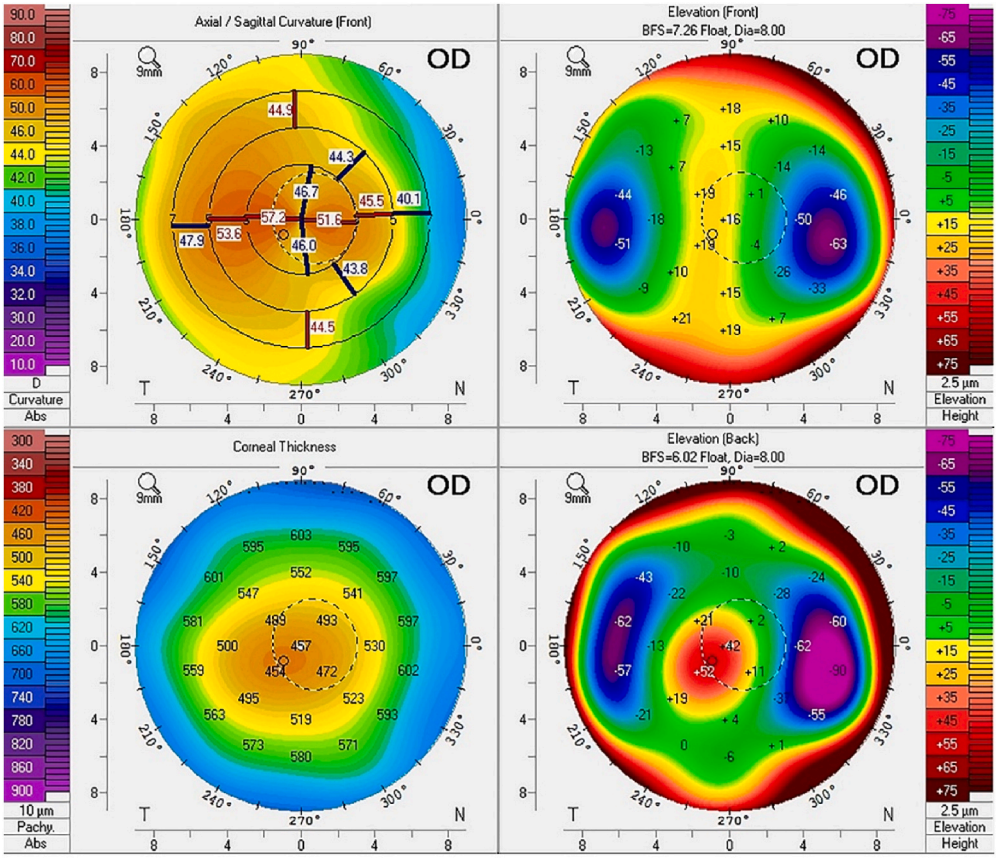
The Pentacam permits assessment of multiple measurements for the diagnosis of keratoconus. The inferior-superior asymmetry index (I-S) is one such measure and is defined as the difference between the average of five points in the inferior and superior hemi-meridians.[15] It can also be calculated by finding the difference between the steep axes of the inferior and superior hemi-meridians within the 6 mm (3 mm radii) zone.[16] A difference between I-S of greater than 1.9D is considered the diagnostic cut-off value of keratoconus.[15–17] The temporal keratoconus described here manifested as an against-the-rule, asymmetric bowtie astigmatism with temporal corneal steepening and thinning. We adopted the method of the I-S index and calculated the temporal-nasal asymmetry index (T-N) at the 3 mm radii zone, which was 5.6D in the right eye and 3.2D in the left eye. This exceeds the accepted (I-S) cutoff value of 1.9D and confirms the temporal steepening and diagnosis of 'temporal keratoconus'. It is also possible that this patient has central keratoconus. Given his exotropia and possible amblyopia with difficulty to fixate, causing an unusual presentation of keratoconus. While this is maintained on the differential, we believe the patient's asymmetric steepening in the horizontal meridian is most consistent with 'temporal' keratoconus.

Several interventions are used to prevent progression of abnormal corneal curvature and refractive error in keratoconus. Treatment varies depending on the severity of keratoconus and presence or absence of progression. Conservative treatment includes refractive correction such as with rigid gas permeable (RPG) contact lenses. Surgical interventions include corneal cross-linking, intracorneal ring segments and penetrating keratoplasty.[4] Keratoconus in the pediatric population tends to

Table 1
Clinical and topography data.

	Distance VAcc[a]	K1 (front)	K2 (front)	Astigmatism	Thinnest Point	Kmax	Temporal K (3mm)	Nasal K (3 mm)	T-N index[b]
OD									
Baseline	20/70 -2	–		–	–	–	–	–	–
Cornea Follow up	20/70	46.5D	54.3D	7.8D @175.1°	450μm	57.9D	57.2D	51.6D	5.6D
OS									
Baseline	20/30 -2	–		–	–	–	–	–	–
Cornea Follow up	20/25 -1	43.7D	44.0D	0.3D @15.8°	501μm	46.1D	45.6D	42.4D	3.2D

[a] VAcc = BCVA with spectacles; μm = micron.
[b] T-N index = temporal-nasal asymmetry index.



B. Left Eye (OS)

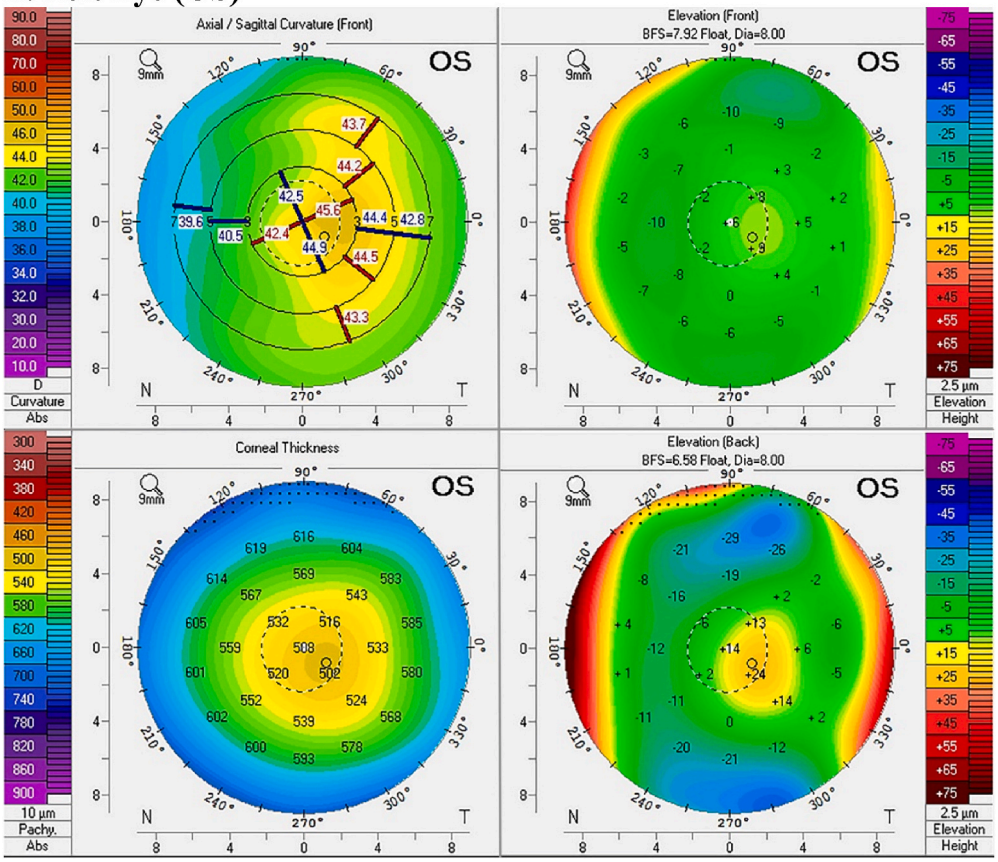


Fig. 1. Topography exam (Pentacam) in the A) right and B) left eyes.

progress more readily as compared to adults.[1] Therefore, early detection and intervention, such as with cross-linking, is considered the gold standard treatment in pediatric patients. Of note, in the United States, corneal cross-linking is FDA-approved for patients between the ages of 14 and 65.[18,19] In the case reported here, our patient had a two-year history of progressively worsening vision prior to his definitive diagnosis of keratoconus. After confirmation of keratoconus on topography, corneal crosslinking was recommended in both eyes.

4. Conclusions

In the present report we describe a unique temporal presentation of keratoconus, a condition characterized by progressive steepening and thinning of the central or paracentral corneal stroma that commonly leads to reduced vision and irregular astigmatism in pediatric patients. Delayed diagnosis and vision correction in patients with keratoconus can contribute to decreased vision, amblyopia and, if progressive, corneal hydrops and ectasia. Eye care specialists should maintain a high index of suspicion for keratoconus in cases of persistent and uncorrectable refractive error in pediatric patients, and should be aware of this temporal variant of keratoconus. Prompt workup including topography can detect the condition and allow for interventions to prevent progression and achieve an optimal visual outcome.

Patient consent

Written informed consent was obtained from the patient to publish case details.

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Authorship

All authors attest that they meet the current ICMJE criteria for Authorship.

Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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Appendix A. Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.ajoc.2023.101900.

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