CASE REPORT

First presentation of keratoconus in a geriatric patient: Diagnosis and treatment of late progression

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This case report presents an atypical, late presentation of keratoconus in a geriatric patient with successful management using a transepithelial corneal crosslinking (CXL) procedure. The treatment-naive 77-year-old patient presented to the clinic after an assessment by an optometrist for recent onset of reduced vision that was initially thought to represent early cataract development. Corneal topography confirmed keratoconus, and subsequent review identified further progression. Crosslinking was performed using a transepithelial approach. The patient was followed for 3 years postoperatively. By the final visit, the uncorrected distance visual acuity had improved and the corrected distance visual acuity remained at 20/20 bilaterally. Corneal topographic parameters were considered stable. This case shows that keratoconus can present outside the previously established parameters and that CXL might be an effective option in older patients showing signs of progression. This might have additional benefits for future intraocular lens power calculations.

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Keratoconus is a corneal disease with typical onset during puberty and progression through the third decade of life. Progressive keratoconus can have a profound effect on vision-related quality of life, manifesting as increasing irregular astigmatism and loss of corrected vision.

The treatment of keratoconus has historically involved the use of spectacles and contact lenses to optimize visual acuity until a corneal graft procedure was required. Although progression of the disease remains rare in older patients, corneal crosslinking (CXL) now provides an effective option to prevent significant deterioration and minimize the need for future corneal surgical procedures. Corneal crosslinking is performed after confirmation of disease progression, with the majority of cases done in younger individuals to maximize visual acuity and the quality of vision.

Gokul et al. recently identified a subset of keratoconic patients who continued to have corneal topographic progression after 40 years of age. The relatively small sample precluded the authors from positively identifying contributing factors, although more significant disease was proposed as a major variable. This suggests that a cohort of keratoconus patients might benefit from late CXL to stabilize vision, in particular taking into account the probable need for future cataract surgery and intraocular lens (IOL) power calculations.

We describe an unusual case of keratoconus in which the first presentation and progression occurred in the eighth decade of life. This case identifies the potential benefit of CXL in managing keratoconus in elderly patients.

CASE REPORT

A 77-year-old woman presented reporting decreased visual acuity at distance and near. The patient had been referred by her regular optometrist, who had noted a recent history of visual and refractive progression that was suspicious for keratoconus in the absence of significant cataract. The patient denied a history of eye rubbing or ongoing irritation of eyes; however, she stated that her grandson had recently been diagnosed with keratoconus. The patient had a history of hyperlipidemia and hypertension well managed with medication.

The patient had no history of contact lens use or surgical intervention in either eye. The uncorrected-2 distance visual acuity (UDVA) was 20/60 correcting to 20/15-2 with +2.00 –0.50 × 90 in the right eye and 20/20 correcting to 20/20 with +0.50 –1.50 × 130 in the left eye. Corneal mapping with scanning-slit corneal topography (Orbscan, Bausch & Lomb, Inc.) was suggestive of keratoconus; the maximum keratometry (K) measurements were 47.7 diopters (D) and 50.1 D in right eye and left eye, respectively. The endothelial cell count (ECC) (SP-3000F, Topcon Corp.) was 2181 cells/mm² and 2251 cells/mm², respectively. Pachymetry at the thinnest point was 468 μm in the right eye and 448
μm in the left eye (Figure 1). Direct ophthalmic examination using a slitlamp showed increased corneal curvature in both eyes (left eye worse than right eye) and mild Vogt striae in the left eye. Fundus and ocular adnexa examination showed no further significant features or abnormalities. The patient was counseled against eye rubbing and asked to return in 6 months to monitor the progression and discuss possible treatment options.

At 6 months, the patient reported further reduction in vision at distance and near. The UDVA was unchanged in the right eye; however, the UDVA in the left eye had worsened from 20/30 at the initial visit to 20/70. The subjective refraction showed refractive progression, with the right eye requiring +2.25 -1.25 × 105 and left eye +1.50 -2.25 × 120 to achieve 20/15 bilaterally. Scanning-slit corneal topography showed that the maximum K values had increased from the initial visit to 49.3 D in the right eye and 52.0 D in the left eye (Figure 2). Scheimpflug corneal topography (Pentacam, Oculus Optikgeräte GmbH) imaging was also taken because the scanning-slit corneal topographer was scheduled to be removed from the clinic after this visit. The maximum K values were similar between Scheimpflug tomography mapping (48.9 D right eye; 51.5 D left eye) and scanning-slit corneal topography (Figure 3). This change was repeatable with multiple scans taken at the initial visit and the 6-month visit. Because the topographic and refractive parameters indicated marked progression of the keratoconus, the need for intervention was discussed with the patient. All other parameters were unchanged from the initial visit.

After discussion about the available therapeutic and surgical options, the patient had bilateral epithelium-on CXL using hypotonic riboflavin (0.1% concentration). Both eyes had an initial 40-minute soak to ensure full uptake of the riboflavin before a 30-minute exposure to ultraviolet (UV) light at 3.15 mW/cm². The procedure was uneventful, and no adverse events were recorded. Six months after CXL, the UDVA was 20/40 correcting to 20/15 with +2.50 -1.00 × 70 in the right eye and 20/60 correcting to 20/15 with +3.00 -2.25 × 100 in the left eye. Scheimpflug corneal topography showed significant bilateral flattening of the cornea compared with the final preoperative visit; the change in corneal cylinder at the 5.0 mm zone was 2.2 D in the right eye and 1.1 D in the left eye (Figure 4). The maximum K measurements were reduced by 1.2 D and 1.9 D, respectively. The patient reported stable vision with little fluctuation or strain at distance or near.

Eighteen months after CXL, the visual parameters remained unchanged in the right eye. However, the left eye appeared to continue to improve, with a UDVA of 6/6 despite a refraction of +1.75 D. Topography parameters remained stable with little change in the maximum K value (0.4 D) or corneal cylinder. The patient presented 3 years after CXL describing a gradual deterioration of distance visual acuity. The UDVA was 20/30 correcting to 20/15 with +2.75 -1.25 × 85 in the right eye and 20/50 correcting to 20/20 with +2.50 -0.75 × 75 in the left eye. Topography showed minimal change from the previous follow-up.
Slitlamp examination showed clear corneas bilaterally and nuclear sclerotic changes. After a discussion regarding further options, it was decided that no additional treatment was necessary at that time.

**DISCUSSION**

This unusual presentation of progressive keratoconus in an older patient shows that keratoconus can progress at any time in life. As this case shows, CXL might continue to provide benefits in slowing the progression of the disease and minimizing the loss of visual acuity in patients outside the typical treatment range.

Keratoconus is typically first diagnosed in younger demographics, with onset normally occurring during the second decade and third decade of life. Naderan et al. examined demographic information of patients at the first presentation of keratoconus; the mean age of first presentation was 21.03 years \( \pm \) 6.17 (SD) (maximum age 49 years). One of the first large-scale studies to examine the demographic associations with keratoconus at presentation was the Collaborative Longitudinal Evaluation of Keratoconus Study. The mean age of presentation (39.3 \( \pm \) 10.9 years) was older than in other similar studies. This was likely a result of the inclusion criteria, which required clinical signs (eg, corneal scarring, Vogt striae) that might not be as obvious in younger patients. More recently, Gokul et al. followed a small group of keratoconus patients for progression. Notably, almost one half of the cohort (48.1%) was older than 40 years at baseline. The authors found that up to 37.0% of patients had an increase in topographic parameters of 1.00 D or more in at least one eye across the follow-up period, suggesting that further late change is plausible. No patient in this cohort had a history of contact lens use, removing this as a potential variable. Our examination of the current literature did not find a case outlining first presentation and progression in a geriatric patient, in particular without confounding factors such as contact lens use or eye rubbing.

According to the Delphi Panel, consistent change in at least 2 topography or tomography parameters (eg, steepening of anterior and/or posterior corneal surfaces) outside the standard deviation of machine measurement error is considered evidence of progression, although the interval between follow-up visits should be titrated to age and the severity of the diagnosis. Patient follow-up in the case reported here found progression in visual, refractive, and topographic parameters, suggesting that CXL was an appropriate option for our patient despite her age.
An alternative procedure is corneal or lamellar transplantation. A study by Duman et al.\textsuperscript{9} assessed the outcomes of corneal transplantation in geriatric patients and found that visual outcomes are often poor and the regraft rates higher in older age groups. The risks for tissue rejection and complexity of the procedure and those associated with invasive surgery in an older patient contributed to the decision to consider CXL for our patient as a first-line treatment. As a noninvasive procedure, it was a preferable alternative to surgical correction. Our results support the treatment decision, showing that CXL can be effective at halting disease progression and subsequent vision loss in older patients.

Given the age of the patient and the likelihood of cataract surgery within the near future, the choice of an epithelium-on procedure was made to decrease the risk for damage to the cornea during the surgery and in the immediate postoperative period. Several studies\textsuperscript{1,10–13} have shown that use of a hypotonic riboflavin solution combined with a longer initial soak period can be an effective alternative to the standard Dresden protocol, which in general requires the cornea to be thicker than 400 $\mu$m to prevent exposure of the endothelium to the UV radiation. Although our patient’s corneas were thicker than 400 $\mu$m bilaterally, the structural integrity of the corneas was likely to be weaker given the age of the patient as well as a decreased number of endothelial cells associated with an aging cornea. The ECC was 2181 cells/mm\textsuperscript{2} in the right eye and 2251 cells/mm\textsuperscript{2} in the left eye compared with the average ECC of 2341 cells/mm\textsuperscript{2} for age ranges 70 to 79 years as established by Raczyńska et al.\textsuperscript{14} and Galgauskas et al.\textsuperscript{15} These measurements do not show a clinically significant decrease from normal limits; however, because the ECCs in our patient were lower than in younger patients who have CXL, we were very cautious with the UV light exposure. Thus, an epithelium-on approach was deemed a safer option for the patient.

Although this case provides promising results, there are limitations. First, the use of different topographic scanners between the visits after surgery adds a slight degree of variability to the postoperative data. In addition, the degree of corneal flattening and changes to topography after CXL might have been amplified as a result of the device change. Further case reports and studies of keratoconus presenting in geriatric populations would be useful in determining a more reliable model of disease progression in older populations and help establish a more refined treatment protocol.
Although the clinic team acknowledges that corneal data before the first presentation would be of benefit, this information was not available for this patient and the patient was not aware of previous mapping.

Also, when our patient was in her youth, the clinical evaluation of keratoconus, including diagnostic imaging, was not comparable to that with modern techniques. Therefore, previous clinical notes, even if available, would have been of limited use and value. The main benefit of previous corneal maps would have been to establish whether the patient had forme fruste keratoconus that suddenly progressed later in life. Although this information does not have a bearing on treatment, it could show the importance of following subclinical presentations of keratoconus over long periods. It is possible that the patient had undiagnosed forme fruste keratoconus that progressed later in life; however, at the time of presentation, it had manifested to progressive keratoconus. Differentiating between forme fruste keratoconus and keratoconus relies on a combination of corneal mapping results, direct slitlamp examination, and evidence of progression over time. In addition, the visual quality and corrected visual acuity can be normal in eyes with forme fruste keratoconus, while true keratoconus presents with decreased visual quality and a gradual loss of corrected vision. The initial on corneal mapping of this patient showed significant inferior steeping with an abnormally thin cornea. The patient reported a recent gradual reduction in her distance vision as the reason for presentation. This indicated progression, and slitlamp examination of the cornea showed increased corneal curvature and the presence of Vogt striae, both clear criteria for keratoconus.

Although keratoconus typically first presents in younger demographics, this case is an unusual example of first presentation at a significantly older age. Despite the age of our patient, CXL might still be an effective treatment to help prevent progression and maintain vision. This might have additional benefits for future cataract surgery by improving the stability and accuracy of biometry values and thereby IOL power calculations.

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