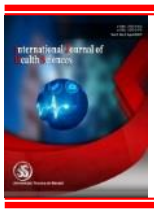

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Managing a Middle-Aged Patient with Bilateral Neglected Keratoconus: A Rare Case Report

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corneal topography;
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Abstract

The Orbscan's benefits include providing a thickness map and being noncontact for keratoconus screening. However, several developing countries have delayed keratoconus management due to a lack of resources and qualified examiners. Here we report a case of underdiagnosed and mistreated by high astigmatism, leading to a neglected case of keratoconus. A 34-year-old woman is bothered by her developing blurred eyesight and habit of scratching her eyes. The streak retinoscopy examination revealed the right eye was 5/20 cc S+7.75 C-6.00 Axis 150 became 5/15 and 5/20 cc S+7.25 C-6.00 Axis 80 became 5/15 for the left eye. The Placido test revealed irregular lines. The Schirmer test showed dry eyes. Slit-lamp examination revealed irregular thin cornea, Munson sign, Rizzuti's sign, and Vogt's striae were positive. The manual keratometer showed remarkably high corneal powers. The corneal topography showed characteristics of keratoconus. Scleral contacts with AS-OCT guiding were inserted into both eyes. Each eye's final BCVA improved to 5/12 and 5/6.5, respectively. The patient attained better visual performance and comfort. Detecting keratoconus by corneal topography and scleral contact lens fitting improved visual acuity. A skilful practitioner and accessible sources are essential to prevent these neglected cases.

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1 Introduction

The progression of corneal ectasia and cone-shaped forward centre thinning that characterizes the early stages of keratoconus results in high irregular myopic astigmatism and impaired vision. The early phases of keratoconus were challenging to identify before advanced technologies were available. Furthermore, the incidence and spread of the disease are perpetuated by subpar research techniques used in various medical facilities and subpar diagnostic and therapeutic approaches (Abuallut et al., 2022). A complex multifactorial aetiology causes keratoconus. The role of genetics remains unclear. High astigmatism, mechanical eye rubbing, ultraviolet exposure, and atopy may contribute to keratoconus development (Alió et al., 2010). Keratoconus begins around puberty and advances slowly. However, the symptoms remain the same throughout time (Rathi et al., 2015). Corneal topography is a sophisticated diagnostic method that has become the gold standard for detecting and diagnosing keratoconus early (Abuallut et al., 2022; Amano et al., 2006).

Several corneal topographic instruments have various features that provide essential screening and diagnosing information for keratoconus, such as The Orbscan (Bausch & Lomb Inc., Rochester, NY, USA), Pentacam (Mirzajani et al., 2019), and Scheimpflug (Yi & Min, 2022). Thinning and central corneal thickness, average K reading, and anterior and posterior corneal elevations are all represented numerically (Mirzajani et al., 2019). The instruments may provide numerous anterior corneal surface indices, providing a complete image of the entire cornea from the flattest to steepest readings (Abuallut et al., 2022).

To increase visual acuity in keratoconus, there are several different contact lens alternatives, intracorneal ring segment (ICRS) implantation, corneal collagen cross-linking (CXL), lamellar keratoplasty, and penetrating keratoplasty. The most popular and successful option for keratoconus is a rigid gas permeable (RGP) lens on the cornea. Scleral lenses, on the other hand, are appropriate for people with severe keratoconus (Kreps et al., 2019). In several studies, a scleral contact lens has been demonstrated to increase visual acuity in keratoconus patients and achieve comfortable fits without touching the ocular surface (Albert et al., 2022). These lenses provide the best chances for correcting vision for uneven corneas due to their geometry; they can delay or even avoid surgery and lower the risk of corneal scarring (Jacobs et al., 2021; Rathi et al., 2015). However, keratoconus management is delayed in several developing country regions due to a lack of resources and qualified examiners (Colin et al., 2000). We present a case of neglected bilateral keratoconus in a middle-aged patient who lived in a remote environment. The patient signs the approval for using included clinical records and images (Laditka et al., 2003).

2 Case Description

A 34-year-old woman with persistent eye rubbing, tired eyes, and growing vision distortion visited the outpatient clinic of a tertiary referral hospital. Over the last ten years, the patient has switched eyeglasses, but her vision has not gotten any better. Morning rhinitis allergies had previously affected the patient. There is no underlying disease or keratoconus in the family. Throughout the entire general evaluation, the patient's vital signs remained normal. The initial visual acuity of each eye was 5/20 and 5/15. Both were difficult to be corrected. The streak retinoscopy examination revealed that the right eye with S+7.75 C-6.00 Axis 150 degrees became 5/15, and the left with S+7.25 C-6.00 Axis 80 degrees became 5/15. The Schirmer test on both eyes was below 10 mm, showing dry eyes.

From the clinical findings of both eyes, a V-shaped lower lid margin was seen when the patient looked downwards: Munson's sign (Figure 1a). The slit-lamp examination revealed Rizzuti's sign (Figure 1b) and Vogt's triae (Figure 1c). A small corneal scar (macula) was seen at the visual axis of both eyes (Figure 1d). The Placido disk presented irregular distorted ring lines closer to the cone's middle (Figure 1e). Fundus photography examination showed a tigroid fundus appearance on both eyes. An automated refracto-keratometer RK-700 was performed twice. However, the K readings were undetected in both eyes.

Alternatively, a manual refracto-keratometer (Inami Ophthalometer) was performed. The right eye showed K1 reading 53 Diopter and base curve 6.6mm and K2 reading 52 Diopter and base curve 6.5mm. The left eye showed a K1 reading of 52 Diopter and a base curve of 6.5 mm, while the K2 reading was 52 Diopter and a base curve of 6.5 mm (Figure 1f). The indented biometry measured long axial length for both eyes, 26.96 mm, and 27.05 mm for the right and left, respectively.

Several measurements, including the central corneal thickness, anterior float shape, colour zone, anterior and posterior elevation best fit sphere, horizontal visible iris diameter (HVID), K1 and K2 reading, can help to identify the characteristics of keratoconus from corneal topography analysis based on the Orbscan II (Bausch & Lomb, Rochester, NY, USA). The right anterior and posterior float of the cornea (Figure 1g,h). The cornea's left anterior and posterior float (Figure 1i,j). These measurements were described in Table 1.

Table 1
Corneal topography analysis

Corneal Topography Analysis	Right eye	Left eye
Central corneal thickness	291 um	277 um
Anterior float shape of keratoconus	globus	globus
Red colour	Central zone	Central zone
Green & blue colour	Peripheral zone	Peripheral zone
Anterior elevation best fits the sphere	6.61mm (51 Diopter)	undetected
Posterior elevation best fits the sphere	4.99 mm (67.7 Diopter)	undetected
Corneal power	120.80 D	128.71 D
HVID	11.67 mm	11.67 mm
K1 reading	64.74 D at axis 20 degrees	69.52 D at the axis 4 degrees
K2 reading	76.59 D at axis 110 degrees	72.96 D at axis 94 degrees

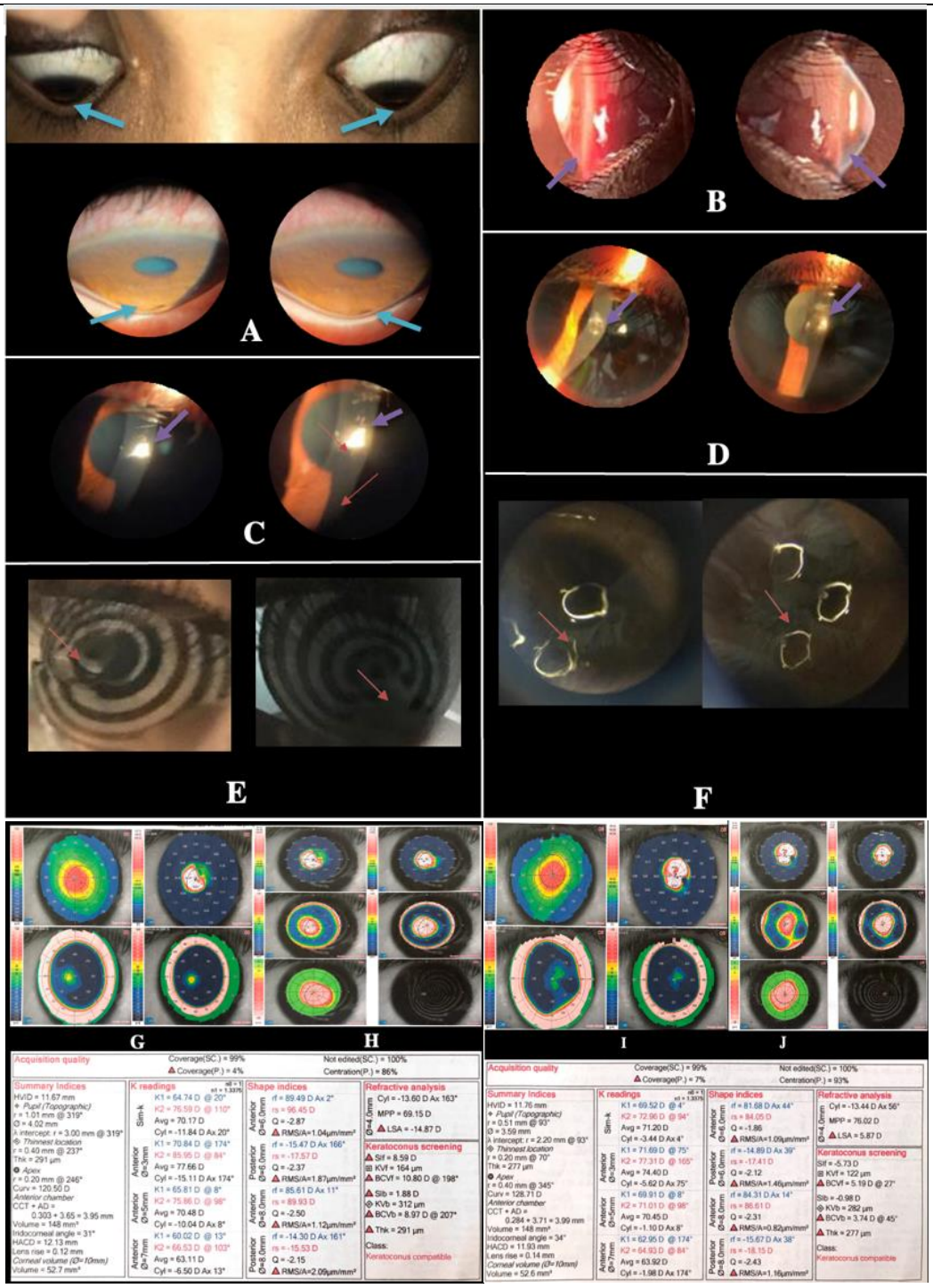

Figure 1. Eye examination images of the patient A) Munson sign of both eyes. B) Rizutti's three signs of both eyes. C) Vogt striae of both eyes. D) Corneal scar. E) Placido disk of both eyes 4 showed irregular distorted ring lines grouped more closely in the central region of the five corneas. F) Manual keratometer showing both eyes' K1 and K2 readings and base curve. Corneal topography of the right eye taken by Orbscan II (G and I) showed G) Anterior Seven float, I) Posterior float. Corneal topography of the left eye taken by Orbscan II (J and K) showed J) Anterior float, K) Posterior float

The patient was fitted with scleral gas permeable contact lenses (Figure 2a). Some evaluations had to be taken care of for the contact lens fitting, such as choosing the correct base curve, diameter, CT, sagittal depth, and corneal clearance. The evaluation is described in Table 2. The value of corneal clearance of both eyes is within the normal limit (Figure 2b). While fitting the scleral lenses and dyeing them with fluorescein drops, it was evaluated on the slit lamp that there was no hyperemia, vascular shrinkage, anoxia, and air bubble on both eyes. The 3 points touch of the scleral contact lens evaluation, including apical touch, paracentral clearance, and peripheral clearance on both lenses, fitted perfectly on both eyes. The final BCVA using the scleral contact lens was improved significantly with the addition of trial lenses. The right eye was added with C-2.50 Axis 90 degrees became 5/12, and the left eye was added with S+1.75 C-5.00 Axis 80 degrees became 5/6.5. The patient achieved better visual performance and comfort.

Table 2
Scleral contact lens fitting evaluation

Scleral contact lens fitting	Right eye	Left eye
Base curve	7 mm	7.25
Diameter	16.6 mm	16.6 mm
CT	0.20	0.20
	(-8.25 Diopter)	(-6.50 Diopter)
Sagittal depth	5.10 mm	4.99 mm
Corneal clearance	227 um	249um

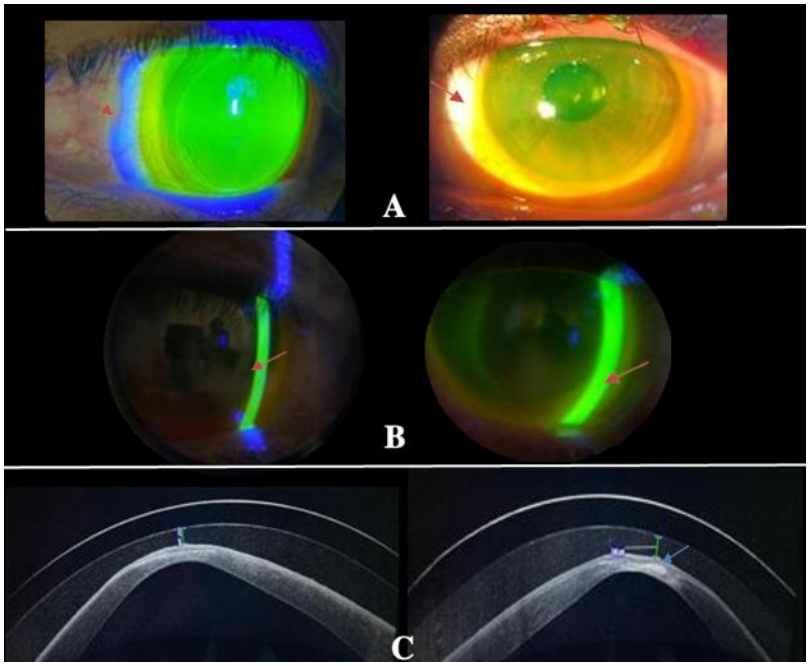

Figure 2. Scleral contact lens fitting. A) Right eye fitting and the corneal clearance evaluation under the slit lamp B) Left eye fitting and the corneal clearance with fluorescein dye evaluation under the slit lamp (A and B) showed 3 points touch of the scleral contact lens evaluation, including apical touch, paracentral clearance, peripheral clearance on both lenses had fitted perfectly, no hyperemia, vascular shrinkage, anoxia, and air bubble on both eyes. C) Corneal clearance of both eyes seen in AS-OCT showed the value of corneal clearance of both eyes is within normal limit

3 Discussions

Patients with keratoconus, commonly present in their teens or twenties, complain of increasing distorted and blurred vision due to myopia and high astigmatism. This patient also had frequent changes of spectacles due to its visual acuity measurement being unstable for some time (Ernawati et al., 2022). Symptoms of keratoconus include photophobia, glare, and monocular diplopia. The early symptom is Rizzutti's sign, a conical reflection of light on the nasal cornea when it is shined temporally. Near the cone's base, the Fleischer ring might be round or have iron deposits within the epithelial layer. Spontaneous tears in the Descemet membrane can cause hydrops. Due to its stretch, the stroma may have fine, nearly stressed lines or striations (Vogt lines). Munson's sign is angulation of the lower lid on downgaze caused by a corneal protrusion in advanced keratoconus (Albert et al., 2022). During retinoscopy, a scissors reflex is a warning indicator. The corneal scar was due to a continuous protrusion of the cornea. This patient manifested most symptoms, such as the Munson sign, Rizutti sign, Vogt lines, and the corneal scar that influenced the visual function. A manual Placido disk examination was used for early detection of anterior corneal curvature in high astigmatism. The ring lines were irregular and grouped at the apex of the cone. This patient was suspected of neglected keratoconus (Abuallut et al., 2022; Alio, 2016).

Several corneal topographic devices have various features that provide essential screening and diagnosing information for keratoconus, such as The Orbscan (Bausch & Lomb Inc., Rochester, NY, USA), Pentacam (Mirzajani et al., 2019), and Scheimpflug (Yi & Min, 2022). Thinning and central corneal thickness, average K reading, and anterior and posterior corneal elevations are all represented numerically. The maps are colour-coded and include numerous indices of anterior corneal surfaces, allowing for a comprehensive picture of the entire cornea, from the flattest to steepest readings. It has also been used to track progress, cross-link collagen, and fit contact lenses (Alio, 2016). This patient was evaluated by The Orbscan (Bausch & Lomb Inc., Rochester, NY, USA), fit detected keratoconus by providing data on anterior and posterior elevation, best-fit sphere, and a corneal pachymetry map, and corneal power. Detecting fruste keratoconus and keratoconus suspects can also be done by calculating the corneal thickness spatial profile and corneal volume distribution (Kreps et al., 2019). The treatment and management of keratoconus are determined by the stage of the condition. According to Standard Amsler-Krumeich Keratoconus classification; stage I presents as eccentric steepening, myopia/astigmatism< 5.00 D, Mean K< 48.0 D, stage II as myopia/astigmatism 5.00- 8.00D, Mean K< 53.0 D, absence of scarring, minimal apical corneal thickness > 400 um, stage III as myopia/astigmatism 8.00 – 10.00 D, Mean K > 53.0 D, absence of scarring, minimal apical corneal thickness 300-400 µm, and stage IV as refraction not possible, mean 16 K > 55.0 D, central corneal scarring, minimal apical corneal thickness <300 µm. 14 Based on the duration of the condition and the result from corneal topography of the patient, the characteristics of the keratoconus was at stage IV.

Contact lenses, intracorneal ring segment (ICRS) implantation, corneal collagen cross-linking (CXL), lamellar keratoplasty, and penetrating keratoplasty are all options to improve visual acuity in keratoconus. Due to the patient's inability to wear spectacles, contact lenses were scheduled to be fitted (Djuraev et al., 2021). High astigmatism, keratoconus severity, topography analysis, occupation, allergy history, ocular surface or dry eye history, and cost were used to determine lens type (Alpysbaev et al., 2021). The most popular and successful option for keratoconus is a rigid gas permeable (RGP) lens on the cornea. Scleral lenses, on the other hand, are appropriate for people with severe keratoconus (Rathi et al., 2015). Scleral contact lenses are designed to fit severely uneven corneas or those who have undergone keratoconus surgery, and they may be a viable alternative to surgical procedures. The goal is to keep the ring/suture area safe from touch while protecting the high vault from corneal abnormalities and asymmetries (Rathi et al., 2017). The AS-OCT-guided scleral lenses allow the practitioner to examine the exact distance between the lens and the cornea and the relationship between the anterior corneal profile and the posterior contact lens surface. The suggested contact lens vault review after 4 hours of lens wear. Avoiding issues and reducing the corneal thickening caused by scleral contact lens usage is necessary (Vincent & Fadel, 2019). The haptic alignment, limbal clearance, and central clearance are all evaluated during scleral lens fitting (Rico-Del-Viejo et al., 2017). Because of the large diameter, there is less lid interaction and less detectable lens movement. The lens rests on the sclera instead of the cornea. A non-preserved saline solution was used to fill the lens to cover astigmatism (Rathi et al., 2015). Some advantages that could be given are stability, good centration, and

improved visual acuity. The larger optical zone maintains stability and better visual outcome than other contact lenses (Rico-Del-Viejo et al., 2017).

4 Conclusion

The techniques employed for The Orbsan's corneal topography can be used for different keratoconus assessments around the globe. The most effective diagnostic method for keratoconus is a corneal topography examination. The results of the streak retinoscopy examination showed that the left eye had a 5/20 cc S+7.25 C-6.00 Axis 80 became 5/15, and the right eye had a 5/20 cc S+7.75 C-6.00 Axis 150 became 5/15. Patients with high astigmatism are more likely to develop keratoconus. Wearing a scleral contact lens significantly enhances visual acuity. A scleral contact lens with AS-OCT guidance was placed in both eyes. The final BCVA improved dramatically to 5/12 for each eye and 5/6.5 overall. The patient's comfort and visual performance improved. The detection of keratoconus using corneal topography and scleral contact lenses increased visual acuity. The patient needs a knowledgeable practitioner and easy access to resources to stop these untreated situations. Therefore, it is crucial to be aware of early diagnosis, have access to diagnostic equipment, and have a thorough understanding of how to manage keratoconus to avoid ignoring cases of the condition.

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