CASE REPORT

Keratoglobus: The Everest of Cones

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Abstract

Keratoglobus is a rare bilateral non-inflammatory corneal ectasia characterized by the diffuse thinning of the entire cornea with increased thinning in the periphery and the protrusion of the cornea. This case outlines the disease as well as treatment and management options.

Keywords: Case report, Cornea ectasia, Corneal degeneration, Corneal diseases, Corneal thinning, Keratoconus, Keratoglobus.

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Introduction

Keratoglobus is a non-inflammatory corneal ectasia that presents similarly to other non-inflammatory corneal conditions with ectasia of the cornea, protrusion, reduced visual acuity, and potential scarring. The initial presentation can easily be mistaken for other similarly presenting conditions, and careful examination should be performed to ensure an accurate diagnosis.

Case Description

A 17-year-old Asian male presented with a referral for examination and scleral contact lens fitting. He complained of blurry vision and a history of ocular trauma. The patient denied any pain, dryness, itching, or tearing. The patient had a history of significant ocular trauma involving his left eye (OS) after being hit with a racquet in the eye. He underwent a ruptured globe repair and subsequently developed endophthalmitis and significant opacification with neovascularization (Fig. 1).

His entering uncorrected visual acuity was 20/400 in the right eye (OD) and light perception in the OS; his pinhole acuity was 20/50 in the OD, and no improvement was found in the OS. The patient's last auto refraction was −9.75−4.75 x 0.28 in the OD only; the OS could not be tested. His pupil in the OD was round and reactive to light. A slit lamp evaluation revealed normal adnexa, lids, lashes, puncta, bulbar conjunctivas, and palpebral conjunctivas in the OD and OS. The right cornea protruded and had 2+ striae, and the left cornea had significant opacification and neovascularization. The lens and iris appeared normal in the OD. The patient's OS developed phthisis bulbi due to significant trauma. The posterior pole was not examined (Fig. 2).

Topography was performed in the OD only and yielded steep K 41.4D, flat K 35.6D, and 5.5D of astigmatism. Pachymetry yielded thinning from limbus to limbus, maxing out at 197 µm and ranging to 380 µm.

Multiple attempts to utilize scleral lens fitting sets failed due to the cornea's irregularity and a significantly steepened corneal scleral junction. The patient's OD was fit using Rose K rigid gas permeable (RGP) lenses, yielding a vision potential of 20/50. The small-diameter RGP lenses were unsatisfactory for a longer-term fit. The patient was then molded using the Eyeprint Pro fitting system to obtain the best possible fit.

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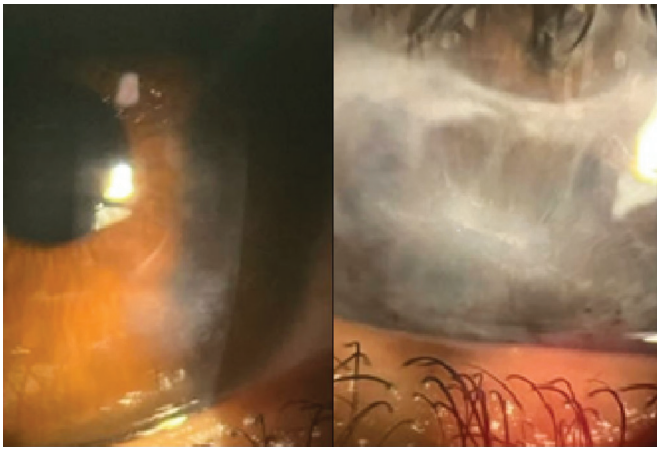

Fig. 1: Bilateral colored photograph: Take with a slit lamp camera of each cornea. Intended to show the present state of each eye at the initial time of scleral fitting

Discussion

Keratoglobus is defined as an ectatic corneal condition that involves thinning from limbus to limbus with the associated protrusion of the globe and more extensive thinning in the periphery without

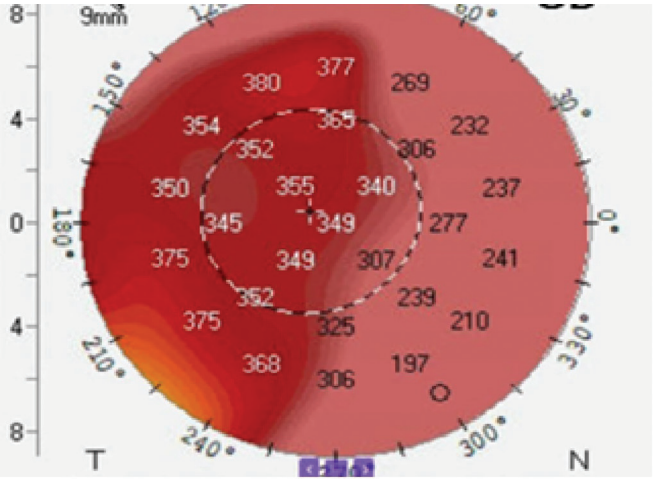

Fig. 2: Right eye: Pentacam corneal thickness analysis. Showing significant date of global corneal ectasia

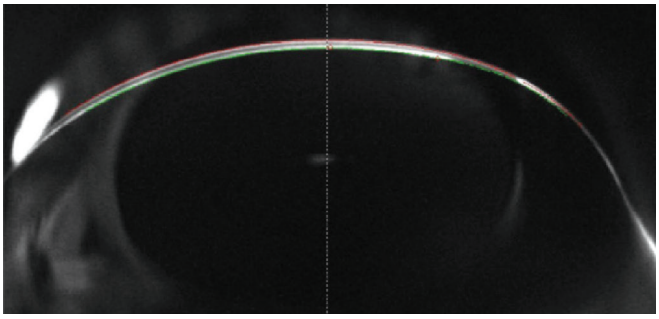

Fig. 3: Right eye: Pentacam Scheimpflug image. Showing the significant corneal ectasia across the entire cornea, and the severity of steepening for the corneal scleral junction

an enlarged corneal diameter.[1] Keratoglobus is a rare non-inflammatory condition that has 2 forms: Acquired keratoglobus and congenital keratoglobus. Both forms have been associated with other ocular and systemic connective tissue disorders. Patients with keratoglobus typically have varying amounts of visual impairment associated with high myopia, irregular astigmatism, the scarring of the cornea, and the possibility of a global rupture (Fig. 3).[2]

Acquired keratoglobus has been associated with vernal keratoconjunctivitis, chronic marginal blepharitis, idiopathic orbital inflammation, and dysthyroid eye disease. In the case of vernal keratoconjunctivitis and chronic marginal blepharitis, the thinning is thought to be from mechanical stress due to excessive eye rubbing.[1]

Congenital keratoglobus is more common than acquired keratoglobus, is present at birth, and is always bilateral. The precise genetics are not fully understood, and a clear inheritance pattern has not been established. The congenital form is assumed to be autosomal recessive. The congenital form has been associated with blue sclera syndromes, such as Leber congenital amaurosis, Ehlers–Danlos syndrome, Marfan syndrome, Rubinstein–Taybi syndrome, osteogenesis imperfecta, and other connective tissue disorders.[3]

Keratoglobus involves thinning throughout the cornea with the more extensive thinning in the periphery. The peripheral thinning may be as significant as one-fifth of a normal thickness.[1] Patients typically present with clear corneas; however, scarring may occur if hydrops or edema has occurred. Patients with keratoglobus do not see corneal diameter changes; intraocular pressure is normal to low; pupil reactions are unaffected, and accommodation and posterior pole findings are within the normal range.[3]

Histologically, it has been shown that corneas in keratoglobus experience disruptions in Bowman's layer or even the complete absence of Bowman's layer. Other problems include the thinning of the stroma and disruptions such as breaks or the thickening of Descemet's membrane.[1,2]

Diagnosing keratoglobus relies largely on presentation and findings during examination. Patients may present with complaints of decreased vision that does not improve with spectacle correction, with visual acuity being minimally reduced, or a severe issue like the inability to count fingers. Slit lamp evaluation is used to assess the cornea's overall health, assess the extent of its protrusion, and evaluate its diameter. Corneal pachymetry should be utilized for corneal thickness, topography, tomography, and anterior.

Segment optical coherence tomography (OCT) is achieved via detailed slit lamp evaluation.[1–3] Keratoglobus should be differentiated from diseases with a similar presentation, such as keratoconus, pellucid marginal degeneration (PMD), congenital glaucoma, and megalocornea.

Keratoglobus differs from keratoconus at the age of presentation: Keratoglobus at birth and keratoconus around puberty. The thinning is also different between them. Although both diseases are corneal ectasia conditions, keratoglobus involves thinning throughout the entire cornea, and keratoconus is typically a localized thinning of the cornea.[1,4] Pellucid marginal degeneration also presents later in life, typically from the ages of 20–40, and has a distinct "crab claw" topographical pattern due to the thinning inferior, typically at the 4 and 8 o'clock positions.[1]

Keratoglobus and congenital glaucoma present at birth; however, congenital glaucoma presents with elevated intraocular pressure, whereas keratoglobus is in the normal to lower range.[1,2] Megalocornea also presents at birth and may look similar to keratoglobus; however, neither corneal thinning nor an increase in corneal diameter is an issue.[1,3]

Despite the minimal success of spectacle correction, spectacles with adequate safety ratings should be prescribed, and the patient should wear them at all times due to the increased risk of corneal rupture and injury. Polycarbonate and CR39 lenses are suitable for protection and correction.[5] Soft contact lenses have not been shown to be successful in treating keratoglobus due to the cornea's irregularity; however, scleral contact lenses have been shown to improve patients' vision. Scleral contacts offer some protection to the cornea and improve vision better than simple spectacles.[6] Due to the cornea's high irregularity, more custom-fitting scleral lenses may be necessary.[7]

Patients who experience acute corneal hydrops should first be treated utilizing lubrication, prophylactic antibiotics, cycloplegics to manage pain, hypertonic solutions (such as Muro 128), and, if needed, intraocular pressure-reducing medications to reduce hydrodynamic forces on the eye. In some cases, a bandage contact lens may be utilized to help with pain as well.[8] In severe cases, intracameral gas injection may also be utilized.[8]

Similarly, to other corneal ectasias, evidence exists that corneal cross-linking can be used to manage keratoglobus. A report in

The Journal of Refractive Surgery Case Reports found success in stabilizing keratoglobus after treatment with corneal cross-linking for up to 32 months' post-treatment.[9]

Penetrating keratoplasties has not been highly successful in treating keratoglobus due to the excessive thinning peripherally at the limbus and the complications of suturing a graft onto such a thin cornea. Due to the rare nature of the condition, the number of cases to study has been limited.[1] A recent report by Cheng et al. showed success in treating keratoglobus utilizing whole lamellar keratoplasty with corneoscleral limbal.[10] No standard surgical procedure for keratoglobus currently exists, and as such, great consideration regarding each case is required prior to surgical intervention.

Conclusion

This case demonstrates the complexity of fitting a highly irregular cornea with conventional scleral lens fitting sets and the need to use a custom-molded fitting. Additionally, the case clearly demonstrates the need for protective eyewear for patients with keratoglobus and the risk of permanent vision loss following trauma.

Further research on genetics, inheritance patterns, and the link to many connective tissue diseases is needed to further understand the disease, the associated risks, and the benefits of various treatment options. The options include corneal cross-linking, keratoplasties, and the use of scleral contact lenses.

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