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REVIEW

Keratoglobus

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Abstract

Keratoglobus is a rare noninflammatory corneal thinning disorder characterised by generalised thinning and globular protrusion of the cornea. It was first described as a separate clinical entity by Verrey in 1947. Both congenital and acquired forms have been shown to occur, and may be associated with various other ocular and systemic syndromes including the connective tissue disorders. Similarities have been found with other noninflammatory thinning disorders like keratoconus that has given rise to hypotheses about the aetiopathogenesis. However, the exact genetics and pathogenesis are still unclear. Clinical presentation is characterised by progressive diminution resulting from irregular corneal topography with increased corneal fragility due to extreme thinning. Conservative and surgical management for visual rehabilitation and improved tectonic stability have been described, but remains challenging. In the absence of a definitive standard procedure for management of this disorder, various surgical procedures have been attempted in order to overcome the difficulties. This article reviews the aetiological factors, differential diagnosis, histopathology, and management options of keratoglobus.
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Introduction

The noninflammatory corneal ectasia are a group of disorders characterised by corneal thinning, protrusion, and scarring. Keratoglobus forms a rarer subset of this group.[1] In the past, it was considered synonymous with megalocornea and congenital glaucoma. However, in 1947, Verrey[2], through detailed descriptions of his patients, was able to show that it was a distinct clinical entity. This was further supported by Cavara[3] in 1950. The exact cause remains largely unknown although various theories have been proposed based on its similarities with other more common noninflammatory ectasia such as keratoconus. In fact, these similarities have brought about confusion as to whether the disorders comprising this group are separate clinical disorders, or rather a spectrum of the same disease process.

Aetiological factors

Keratoglobus is primarily considered a congenital disorder present since birth.[3–5] However, in more recent years, there have been reports of acquired forms of keratoglobus. The congenital form of the disorder is always bilateral. The exact genetics of the disorder have not been studied in detail and no definite inheritance pattern has been described. It is assumed to be autosomal recessive, as described by Poliquen *et al*.[4,5] It has also been associated with disorders of the connective tissue such as Ehlers–Danlos syndrome, Marfan syndrome, and Rubinstein–Taybi syndrome (Table 1).[6] Initially, there were reports of keratoglobus in relation to 'blue sclerae'.[5,7,8] These blue sclera syndromes were actually thought to be manifestations of the aforementioned syndromes, including osteogenesis imperfecta.[9,10] The seemingly blue sclera is caused by a thinned and more transparent sclera, maximally at the ciliary body. Ehlers–Danlos syndrome type VI, in particular, is distinct for its ocular manifestations.[11] These include corneal abnormalities of cornea plana, keratoconus and keratoglobus, blue sclera, and ocular fragility. Other systemic features in this type are the presence of hypermobile joints, skeletal abnormalities like scoliosis, pectus excavatum, a marfanoid habitus, and hearing loss. Skin laxity and fragility is not a characteristic finding, unlike in other types of Ehlers–Danlos syndromes, and lysyl hydroxylase activity may be normal.[11,12] Keratoglobus has also been described in cases of Leber's congenital amaurosis.[13]

Table 1 Reported associations of keratoglobus

	Systemic features	*Ocular features*
(A) Connective tissue disorders		
(i) Ehlers–Danlos syndrome type VI[5,7,8,11,12,51,58]	Musculoskeletal abnormalities, hyperflexible joints, hearing loss, marfanoid habitus	Blue sclera, keratoconus, cornea plana
(ii) Marfan syndrome[5,7,8,59]	Musculoskeletal defects: dolichostenomelia, arachnodactyly, prognathism, high-arched palate, kyphoscoliosis, pectus excavatum, pectus carinatum, joint laxity, cardiovascular abnormalities	Ectopia lentis, microspherophakia, lens opacities, ocular colobomas, cornea plana, keratoconus, open-angle glaucoma, retinal tears and detachments, blue sclera, strabismus, iris muscle hypoplasia, high myopia
(iii) Rubenstein-Taybi syndrome[6,60,61]	Mental retardation, postnatal growth deficiency, microcephaly, broad thumbs and big toes, congenital heart defects, joint hypermobility, obesity	Lacrimal duct problems, high myopia, nystagmus, pupillary abnormalities, microcornea/microphthalmia, corneal abnormalities, congenital glaucoma, iris atrophy, congenital cataract, ectopia lentis, microphakia, ocular coloboma, chorioretinal dystrophy, optic atrophy, optic disc anomalies
(iv) Osteogenesis imperfecta[9,10,62]	Multiple fractures at birth, early deformity of lower extremities, coxa vera, osteopaenia, rhizomelia	Blue sclera (types I–III)
(B) Leber's congenital amaurosis[13]	Mental retardation	Congenital retinal blindness: severe visual loss, wandering nystagmus, amurotic pupils, pigmentary retinopathy. Oculodigital sign of franceschetti, ptosis, strabismus, high hyperopia, high myopia, keratoconus, microphthalmos, lenticular opacities, macular coloboma, maculopathy, disc oedema, retinal vascular attenuation
(C) Syphilis[18,63–65]	Primary syphilis: chancre Seconday syphilis: maculopapular rash, lyphadenopathy, constitutional symptoms, mouth ulcers, gastrointestinal, ophthalmological, hepatic disease Tertiary syphilis: obliterative endarteritis that can affect any organ	Papillary/granulomatous conjunctivitis, scleritis/episcleritis, interstitial keratitis, cataract, anterior uvetis, roseola, vitritis, chorioretinitis, retinitis, neuroretinitis, retinal vasculitis, inflammatory disc oedema, papilloedema, optic atrophy, uveitic glaucoma, cranial nerve palsies, Argyll–Robertson pupil
(D) Dysthyroid eye disease[15,66,67]	Hyperthyroidism, hypothyroidism	Ocular surface disease; Orbital congestion: lid oedema, chemosis, exophthalmos, dilated episcleral vessels, optic neuropathy; Inflammatory myopathy
(E) Posterior polymorphous dystrophy	—	Dystrophic endothelial cells, polymorphous corneal opacities, vesicles at Descemet's membrane level, peripheral anterior synechiae, glaucoma, Terrien's marginal degeneration, keratoconus
(F) Acquired		
(i) Vernal keratoconjunctivits[14,68–70]	Atopic conditions	Giant papillae ('cobble-stone' appearance), Trantas' dots, conjunctival hyperaemia, corneal shield ulcers, corneal neovascularisation, keratoconus, cataract, steroid-induced glaucoma
(ii) Chronic marginal blepharitis	—	Lid margin thickening, scaling at lid margin, ulcerative blepharitis, marginal keratitis
(iii) Idiopathic orbital inflammation	—	Proptosis, cranial nerve palsy (Tolosa Hunt syndrome), uveitis
(iv) Post-traumatic	—	Corneal scar

The acquired forms of keratoglobus have been described in association with vernal keratoconjunctivitis, chronic marginal blepharitis, idiopathic orbital inflammation,[14] and dysthyroid eye disease.[15] In the case of vernal keratoconjunctivitis and chronic marginal blepharitis, the corneal ectasia may be related to frequent eye-rubbing.[14,16] This has been thought by some authors to be a factor in the development of keratoconus, although the exact mechanism of such an association has not been proven.[17] Overlaps in aetiological factors between keratoconus, pellucid marginal degeneration, and keratoglobus, such as their manifestations in connective tissue disorders and various acquired forms has further fuelled the speculation about their being different spectrums of the same disease. Also, there are reports of keratoconus and keratoglobus,[3,14,18] as well as pellucid marginal corneal degeneration (PMCD) and keratoglobus[19,20] being clinically documented in the same patient over time. Poliquen *et al*[4] classified keratoglobus into two types, one being congenital and the other acquired. He described the acquired type as being a severe form of keratoconus. Topographical analysis of pellucid marginal degeneration and keratoglobus by Karabatsas and Cook[19] includes a case report of a patient diagnosed with pellucid marginal degeneration in one eye and who was later found to have developed a keratoglobus-like picture on the follow-up. This led them to hypothesise that the natural history of pellucid marginal degeneration may be in the development of keratoglobus by circumferential extension of the peripheral gutter. Similarly, Cameron[14] described a case report documenting kerotoconus progressing into a keratoglobus-like picture. However, a constant association and progression between these ectatic disorders has not been described to validate these hypotheses. In the reports of keratoglobus associated with thyroid ophthalmopathy[15] and orbital inflammatory disease,[14] the authors hypothesise ischaemia of the

anterior segment secondary to these conditions resulting in diffuse, progressive ectasia of the cornea. Case reports of keratoglobus with syphilis, a post-traumatic case and posterior polymorphous dystrophy have also been described in the literature.[18,21]

Clinical findings

Keratoglobus is a bilateral ectatic disorder of the cornea, principally characterised by a globular protrusion of the cornea associated with diffuse thinning from limbus to limbus. The age of onset is at birth. The thinning is commonly maximal at the periphery and may be up to one-fifth the normal corneal thickness (Figure 1a). Other corneal parameters, however, are normal, including a normal corneal diameter that is an important criterion in differentiating it from conditions such as buphthalmos. Patients generally present with clear corneas unless they undergo acute episodes of hydrops and scarring. Vogt striae and Fleischer's rings are not associated with keratoglobus.[1,14,18]

As a result of the thinning and protrusion, there is high myopia with irregular astigmatism, which is the main cause of poor vision in these patients, and is difficult to treat with refractive correction. Other than poor vision

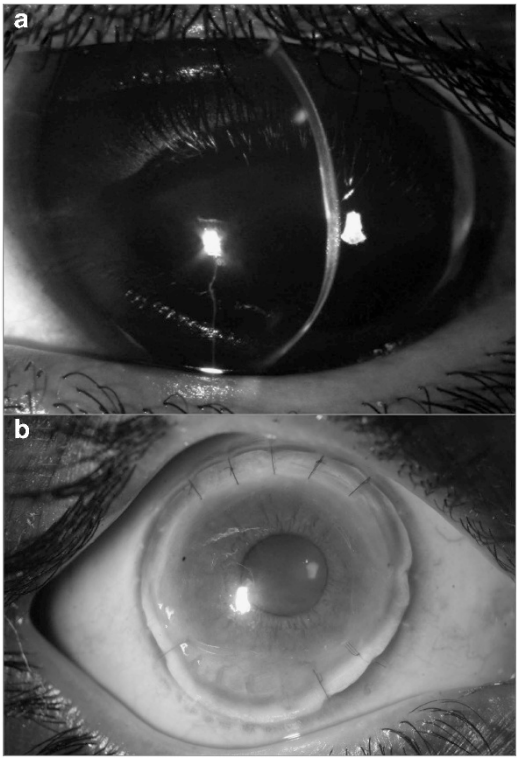

Figure 1 Slit-lamp photograph of a patient with bilateral keratoglobus (a) showing thinning (maximum at the periphery) and bulging of cornea, (b) after 'tuck-in' lamellar keratoplasty (TILK).

patients are generally asymptomatic. However, owing to extreme thinning and fragility of the cornea, many cases may initially present with corneal perforations, either spontaneous or following minimal trauma.[7,10,14,18] In such cases, the diagnosis of keratoglobus must be kept in mind. This is especially important as surgical closure in these cases is difficult because of the extreme thinness of the cornea that leads to cut through of the sutures and inability for wound closure. As mentioned earlier, spontaneous tears in Descemet's membrane may occur resulting in acute presentations of pain, tearing, photophobia, and sudden diminution of vision of acute hydrops.[14,22,23] The loss of endothelial cell barrier leads to fluid accumulation within the stroma. Resolution may take months, with variations from 5 to 36 weeks reported in other corneal ectatic diseases. A study of anterior segment optical coherence tomography of cases involving acute hydrops described two stages in resolution.[24] The first was reattachment of Descemet's membrane, followed by endothelial migration across the tear. There are no other associated ocular abnormalities with keratoglobus.

The diagnosis of keratoglobus is essentially a clinical one owing to the characteristic clinical findings. Confusion might arise in less severe cases where there may be difficulty in differentiating the condition from other ectatic conditions. Investigational modalities include ultrasonic pachymetry that would demonstrate reduced corneal thickness and corneal topography by Orbscan TECHNOLAS Perfect Vision GmbH (Munich, Germany) that would show diffuse thinning.[17,19,25,26] However, these investigational modalities may not be possible to carry out in cases of more severe corneal distortion. There is limited literature describing exact topographical pictures of keratoglobus. A description by Karabatsas and Cook[19] of a patient with both pellucid marginal degeneration and keratoglobus described the videokeratography image, showing irregular astigmatism with irregular power distribution of the eye with keratoglobus. There was a peripheral arc of increased power or steepening resulting in flattening of 'arching' of the bow tie configuration of topography. They felt that the peripheral arc of increased power reflected a progression of PMCD by extension of the inferior peripheral thinning circumferentially.[19] Another case report of keratoglobus in association with posterior polymorphous dystrophy showed Orbscan findings of generalised steepening of both anterior and posterior curvatures, with irregular astigmatism and asymmetric bow tie pattern.[21]

Systemic evaluation might point towards connective tissue disorder. These include features mentioned earlier of blue sclera, joint hypermobility, skeletal abnormalities, hearing loss, abnormal dentition, high-arched palate, as

well as other features specific to each syndrome. It has been noted in few case series that keratoglobus in association with blue sclera syndromes is more probable to undergo spontaneous perforation or after minimal trauma, and hence the name 'brittle cornea'.[7,14]

Histopathology

The anatomical features that have been described for keratoglobus corneas are frequent disruptions or complete absence of Bowman's layer, stromal thinning and disorganisation, and breaks or thickening of Descemet's membrane (Figure 2).[4,14,27] Stromal thinning is most marked at the periphery or mid-periphery, as seen clinically. Poliquin *et al*[4] described different histopathological findings for the acquired type of keratoglobus, which is characterised by an essentially normal Bowman's layer that undergoes focal breaks with superficial stromal ectasia secondary to other corneal pathology. The histopathalogical findings are similar to that of keratoconus.

Immunohistochemical studies of keratoconus have been done, but are limited in cases of keratoglobus. A study done by Meghpara *et al*[27] described the immunohistochemical features of nine corneal buttons of keratoglobus patients in comparison with keratoconus and normal cornea. They found decreased expression of proteinase inhibitor alpha-1-PI, and increased expression of the transcription factor Sp1 in the corneal epithelial cells. This imbalance leads to an alteration in tissue degradation processes within the cornea. In addition, they found increased expression of matrix metalloproteinases (MMPs) 1, 2, and 3 within the epithelial cells. The MMPs are responsible for the degradation of some component of the extracellular matrix and are transcriptionally upregulated by various inflammatory mediators and inhibited by tissue inhibitors of metalloproteinases.[28] Their role in wound repair and remodelling is well known.[29] Similar findings were observed in keratoconus. However, in keratoconus, there was an increased expression of only MMP 1, which was maximal at the centre, corresponding to the area of maximal thinning in this disease. In comparison, increased Sp1 and MMP 1, 2, and 3 expression was found diffusely throughout the cornea, but maximally at the mid-periphery and corresponded to areas of underlying Bowman's layer disruptions. The increased expression of pro-degradation products and decreased expression of inhibitory substances is most probably a key pathogenic factor in causing ectasia.

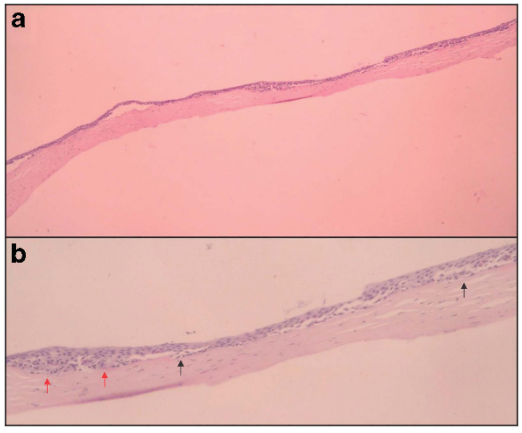

Figure 2 Photomicrograph of (a) lamellar corneal button showing diffuse stromal thinning (stain: haematoxylin and eosin, ×40), (b) central cornea showing thinned corneal epithelium with intraepithelial oedema and separation of epithelium from epithelial basement membrane. There is discontinuity of Bowman's layer (marked between the two red arrows and two black arrows; stain: haematoxylin and eosin, ×400).

Differential diagnosis

The main differential diagnosis of a case of keratoglobus is the other noninflammatory ectatic disorders. These include keratoconus, pellucid marginal degeneration, and posterior keratoconus.[1,17,29] In addition, confusion might arise in young patients in differentiating keratoglobus from congenital glaucoma and megalocornea.

In a case of megalocornea, the main differentiating feature is the increased corneal diameter (usually over 12.5 mm) with absence of any corneal thinning. This is in contrast to keratoglobus where corneal diameters are normal and there is profound, diffuse thinning. There is therefore an absence of any corneal protrusion, astigmatism, hydrops, or scarring in cases of megalocornea. Congenital glaucoma may also present with moderate protrusion of the cornea, hydrops, and mild astigmatism with myopia. However, the hallmark would be raised intraocular pressure and possible glaucomatous optic nerve changes that would be absent in the case of keratoglobus. There is also no corneal thinning and corneal diameters may be increased. In congenital glaucoma, the myopia would result primarily from the increased anterior and posterior axial length, whereas in keratoglobus it would be mainly because of the increased corneal curvature.

Keratoconus is different from keratoglobus in the age of presentation. Whereas keratoglobus presents at birth, keratoconus develops around puberty and may progress until 40–50 years of age. Keratoglobus is considered a non-progressive or minimally progressive disorder. The corneal thinning in keratoconus is most commonly seen in the inferior paracentral aspect of the cornea. The protrusion is commonly described as conical in shape,

with maximal thinning at the apex. This is in contradiction to keratoglobus that presents with diffuse thinning and a globular protrusion. Scarring is more commonly found in keratoconus, and there are the presence of Vogt's striae and Fleischer's ring. In case of pellucid marginal degeneration, the age of presentation is around 20–40 years. Thinning involves the inferior aspect of cornea, as a band of 1–2 mm width and extending from 4 to 8 o'clock. Protrusion occurs superior to this area of thinning leading to characteristic topographical patterns. Scarring and hydrops may also occur.

Treatment

Treatment of keratoglobus remains challenging to date. Conservative therapy is refractive correction for high myopia, but is limited by the high irregular astigmatism. Use of contact lens and newer scleral lenses have been described, but is still a matter of debate because of the theoretical risk of perforation on contact lens insertion and removal over corneas that are known to perforate on even trivial trauma.[14] There are no specific studies or literature on contact lens fitting in cases of keratoglubus. The extreme protrusion and irregularity makes fitting complicated, with a need for balance between optical and lens stability. Customised fitting, on patient to patient basis, of sclera lenses, small diameter rigid gas permeable (RGP) lenses, reverse geometry hydrogel lenses, as well as large diameter inverse geometry RGP lenses have been described for corneal ectasias.[30–35]

Of prime importance for these patients is counselling for use of protective eye wear, and avoidance of contact sports owing to the high risk of perforation. However, in children, enforcement of use of protective glasses is difficult making them susceptible to injury.

Treatment of acute hydrops can be both conservative and surgical. Conventional treatment is nonspecific, and involves the use of patching, bandage contact lens, topical hypertonic saline, and cycloplegics to reduce the oedema.[22–23] In recent years, reports on faster resolution with use of intracameral gas had brought a shift towards surgical intervention of acute hydrops. Use of air, sulphur hexafluoride, and perfluoropropane has been described.[36–41] The gas is thought to act as a mechanical barrier to fluid entry into the corneal stroma, as well as a tamponading agent in re-opposing the rolled, detached edges of the Descemet's tear. This is theoretically felt to aid in endothelial migration across the tear.[39] Various studies have shown a significantly faster resolution time with use of these agents as compared with conventional therapy in acute hydrops with keratoconus as well as a few case reports in eyes of kerotoglobus.[36,40–41] A study by Basu *et al*[41] found that the resolution time did not show much difference between conventional and intracameral perfluoropropane treatment in cases of pellucid marginal degeneration and keratoglobus. They felt that this may have been due to location of breaks in these cases as well as the usually larger, more extensive breaks associated with keratoglobus.

Surgically, there is no known standard procedure for management of the condition owing to its rarity, and therefore scarcity of reports of consistent surgical results. The aim of surgical intervention was earlier restricted to the repair of perforations. As mentioned earlier, this commonly resulted in poor outcomes because of the nature of the perforations that are usually large and stellate, and owing to the fragility of the thinned cornea that prevented stable placement of sutures that would cut through or 'cheese-wire'.[14] Attempts to overcome these problems have been reported in few case reports using intracameral air or perfluoropropane as tamponading agents to achieve wound closure in adjunct to suture closure.[42]

Conventional penetrating keratoplasty in these patients is also not possible because of the thinned cornea and peripheral graft–host thickness disparity that prevents adequate wound closure. Patients also are left with extreme irregular astigmatism and ultimately poor visual outcome or tectonic stability. Attempts to overcome these problems have led to reports of modifications of the penetrating keratoplasty procedure (Table 2). These include the use of large limbus to limbus donor corneal grafts to avoid placement of the graft–host junction at the thinned mid-periphery, thereby creating better stability. However, this led to loss of the immunological privelege within the avascular cornea, increasing chances of graft rejection, and therefore requiring long-term immunosuppression and its antecedent complications.[43,44] There is also a case report on the use of midsize (9 mm diameter), eccentrically placed, paracentral graft with the aim of centralising the graft at the point of maximum thinning of host cornea in order to avoid the graft–host junction at this point.[45]

Lamellar keratoplasties have also been attempted in these cases, namely epikeratoplasty. This is a type of onlay lamellar procedure. Following posterior dissection of the conjunctiva and removal of host epithelium, a donor corneoscleral lenticule devoid of endothelium and Descemet's membrane is placed over the host cornea and sutured over it at the periphery to sclera.[9] Epikeratoplasty was first described by Kaufman and Werblin[46] in treatment of aphakia, myopia, and keratoconus in children. It is considered a safe and easy procedure, especially in eyes with thinned corneas making extensive host corneal manipulation risky. However, such large onlay grafts disrupt and overlie the host limbal stem cells that results in delayed

Table 2 Surgical Procedures for Keratoglobus

Surgical procedures	*Advantages*	*Drawbacks*
Penetrating keratoplasty procedures		
Limbus-limbus donor grafts	Avoids graft–host junction at thinned mid-periphery	Loss of immune privilege, Limbal stem cell disruption, Angle structure disruption
Lamellar keratoplasty procedures		
Epikeratoplasty	Easy to perform, Good tectonic stability, Corneal flattening effect, Reduced myopia and astigmatism	Limbal stem cell disruption, Persistent epithelial defect
Epikeratoplasty with 360 host peripheral intrastromal pocket for peripheral donor lenticule	No limbal stem cell disruption, No angle structure disruption, Provides stable host bed for possible secondary penetrating keratoplasty	Interface opacities and intraepithelial cysts
Epikeratoplasty followed by secondary penetrating keratoplasty	Visual rehabilitation following host bed stabilisation by epikeratoplasty	2-stage procedure
'Tuck-in' lamellar keratoplasty	Good tectonic stability, No limbal stem cell disruption, No angle structure disruption	Technically difficult, Interface opacities
Pentacam-based deep anterior lamellar keratoplasty	3D topographical analysis preoperatively, Decreased endothelial rejection rate	Technically demanding
Corneoscleral rim	Easy to perform, Buttress over thinned corneal periphery for tectonic stability Slowed progression of mid-peripheral thinning Allows for delay in further surgical intervention Decreased immune reaction	Temporary measure

re-epithelialisation of the graft or persistent epithelial defects that are the commonest complications of the procedure.[47,48] This in turn may lead to increased chances of infection and the ensuing complications. Javadi *et al*[49] have described a variation in the technique to avoid limbal stem cell damage. They described formation of a 360 degree peripheral lamellar intrastromal pocket in the host cornea into which the donor corneal lenticule is inserted peripherally, thereby avoiding any manipulation at the host limbus. Other complications reported include interface opacities and intraepithelial cysts that would affect the final visual outcome.[50] However, the procedure provides a stable host bed for a secondary penetrating keratoplasty for visual rehabilitation, should it be required. Such a procedure has been described by Macsai *et al*,[51] and Jones and Kirkness.[52] Surgical stability and visual outcome reported though are much better as compared with a conventional penetrating keratoplasty in these eyes.

Another procedure described for surgical management of keratoglobus is corneoscleroplasty, modified to avoid the host angle structures.[53] This involves a central full thickness penetrating keratoplasty with a lamellar peripheral corneoscleral dissection from the edge of the full thickness keratoplasty to ~14 mm. The deeper tissue of the corneoscleral graft is dissected appropriately in the periphery to fit over the lamellar host sclera dissection peripherally. The authors of this procedure report both a good structural integrity and visual outcome in two patients of ectatic corneal disorders with avoidance of secondary glaucoma. However, delayed epithelialisation and immunological graft reactions were complications encountered with the procedure necessitating long-term immunosuppression.

In attempts to overcome the problems in surgical management, many modifications in technique have been described by various authors. Another such procedure described by Vajpayee *et al*[54] is 'tuck-in' lamellar keratoplasty. This involves lamellar dissection of the central cornea and lamellar dissection of intrastromal pockets in the periphery into which the peripheral flange of the donor corneal lenticule is tucked in (Figure 1b). The tucked in peripheral flange especially provides structural stabilisation at the point of maximal thinning. Lamellar dissection of an already thinned cornea is technically demanding, however, with chances of perforation during the procedure. Interface opacities associated with lamellar procedures might also be expected, but similar to epikeratoplasty, the initial procedure stabilises the host bed for a future penetrating keratoplasty, should it be required. A mean follow-up of 1.7 years postoperatively in 12 patients undergoing this

procedure by Kaushal *et al*[55] showed improvement in best-corrected visual acuity, with significant reduction in the spherical equivalent and astigmatism. There were no cases of interface haze either. The same procedure has been used in both pellucid marginal degeneration and keratoconus by the authors, limiting the peripheral flange to the inferior 180 degrees where maximal thinning is present.

In 2005, Kanellopulous *et al*[56] reported use of a corneoscleral rim over the thinned corneal periphery of a patient with keratoglobus that acted as a buttress while avoiding any manipulation of the central visual area. They felt that this technique slowed the progressive expansion of the thinned mid-periphery, was easy and safe to perform, with minimal chances of immunological reaction, and helped to delay the need for further surgical intervention.

More recently, pentacam-based big bubble deep anterior lamellar keratoplasty was carried out in a series of 50 patients, one of which was a case of keratoglobus.[57] Use of pentacam provides a three-dimensional image or thickness profile of the entire cornea preoperatively, which is crucial in the assessment of depth of lamellar trephination for better outcome. However, deep dissection to Descemet's membrane level is surgically demanding and difficult, more so in ectatic conditions. They report an overall conversion rate of penetrating keratoplasty of 16% using this technique. Details on a case to case basis, such as for the single case of keratoglobus, is not reported.

There is no standard surgical procedure in the management of keratoglobus. Reports of surgical results are limited by the rarity of the condition. Individual procedures have their own advantages and disadvantages, and choice of procedure would depend on individual surgeons' choice and technical ability.

Conclusion

Keratoglobus is a rare noninflammatory ectatic corneal disorder with limited literature on the subject. Various case reports and case series by different authors have brought out the fact that it is a distinct corneal disorder with characteristic clinical findings and aetiological associations. Although primarily congenital, an acquired form has also been recognised secondary to other corneal pathology, giving rise to a keratoglobus-like picture. Histological and immunohistochemical studies have shown a degradative process causing stromal thinning in both keratoglobus and keratoconus, although the exact pathogenesis is still unclear. This further emphasises the possible connection between different noninflammatory ectatic conditions, leading some authors to believe they are phenotypic variations of the same underlying condition. Treatment remains a challenge both in visual rehabilitation as well as maintaining structural integrity of the cornea. Various surgical procedures have been described, but no treatment standard exists because of limited patient reports and follow-up of newer surgical procedures.

Conflict of interest

The authors declare no conflict of interest.

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