Surgical Management of Keratoglobus: A Case Report

Diksha Prakash* Samir Kumar**, OPS Maurya**, Abhishek Chandra***

Abstract

Keratoglobus is a rare noninflammatory corneal thinning disorder characterised by generalised thinning and globular protrusion of the cornea. 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. Managing this condition may be challenging due to lack of definitive procedures.

Introduction

Keratoglobus is the rare form of corneal ectasia group of disorders characterised by corneal thinning, protrusion, and scarring. Though primarily considered a congenital disorder[1,2], in recent years, there have been reports of acquired forms of keratoglobus. The congenital form of the disorder is always bilateral, exact genetics has not been described. It is assumed to be autosomal recessive, as described by Poliquenet al.[3]

Acquired form has been seen associated with disorders of the connective tissue such as Ehlers–Danlos syndrome, Marfan syndrome, and osteogenesis imperfecta and others such as Leber's congenital amaurosis, vernal keratoconjunctivitis, chronic marginal blepharitis, idiopathic orbital inflammation and dysthyroid eye disease[4].

Clinical findings include globular protrusion of the cornea associated with diffuse thinning from limbus to limbus. The thinning is commonly maximal at the periphery and may be up to one-fifth the normal corneal thickness. Vogt striae and Fleischer's rings are not associated with keratoglobus.[5,6] As a result of the thinning and protrusion, there is highmyopia with irregular astigmatism, which is the main cause of poor vision in these patients, and is difficult to treat with refractive correction. The diagnosis of keratoglobus is essentially a clinical one owing to the characteristic clinical findings, corneal pachymetry may supplement in cases with clinical dilemma.

Case report

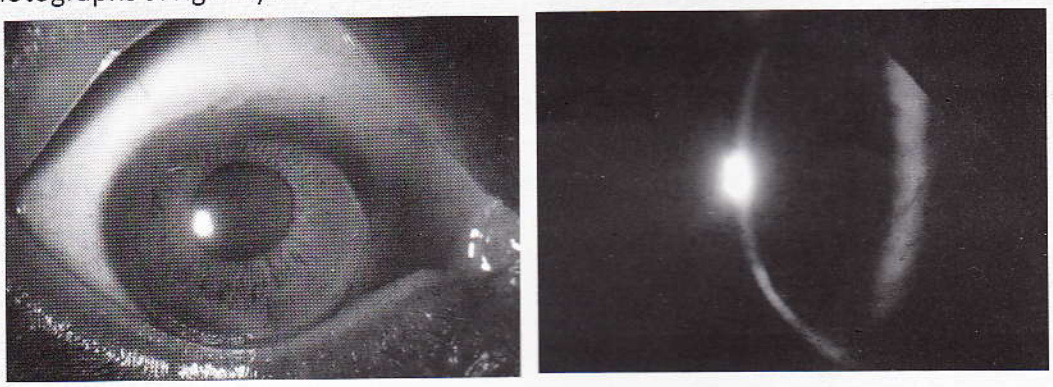
52 year old male farmer presented to us with complaints of both eye gradual progressive, painless diminution of vision for near and distance for last 20 years. No significant past or family history.

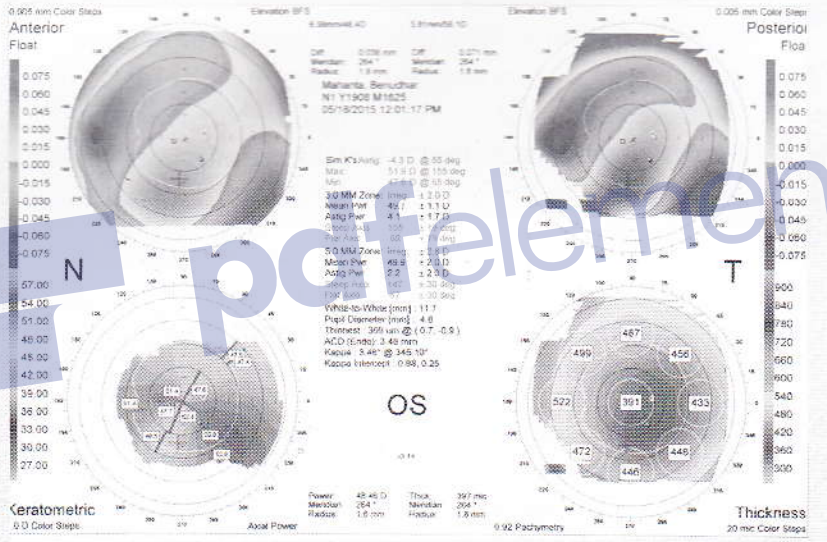
Unaided visual acuity in right eye was counting finger at 1 meter which improved to 20/50 with pin hole. Retinoscopy finding was -12.00 D sph / -10.00 D cyl @140°. The vision in left eye was 20/60 improving to 20/30 with pinhole. Retinoscopy finding was -2.50 D sph / -5.00 D cyl @40°. Near vision was N10 with +1.25 D in both eyes.

* Fellow, L.V. Prasad Eye Institute, Bhuvaneshwar
** Deptt. of Ophthalmology, Institute of Medical Sciences, BHU, Varanasi
*** Director, Chandra Eye Care, Lanka, Varanasi

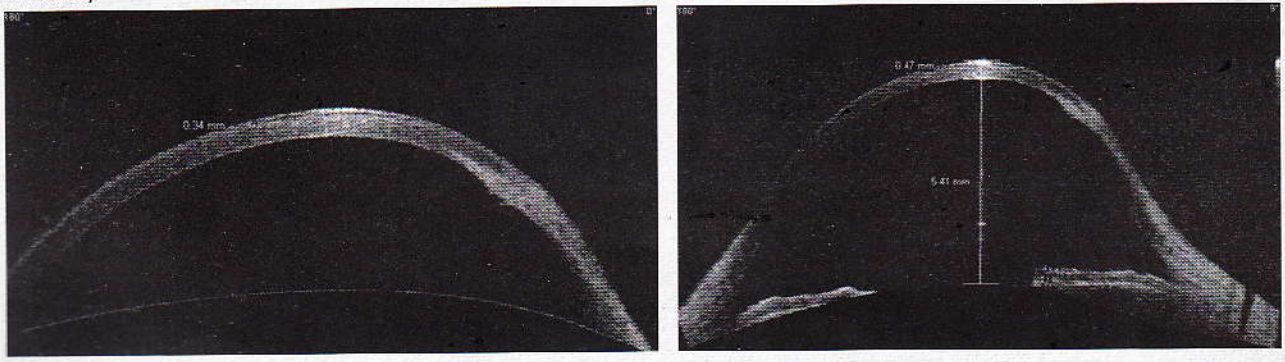
Slit lamp examination of right eye shows diffuse globular corneal thinning with generalized ectasia and inferior scarring as shown in the picture below. Left eye also showed globular protrusion. Rest of the ocular findings were within normal limit.

Clinical photographs of right eye





Orbscan of left eye showing generalized steepening with thinned out cornea (Right eye scan could not be obtained)



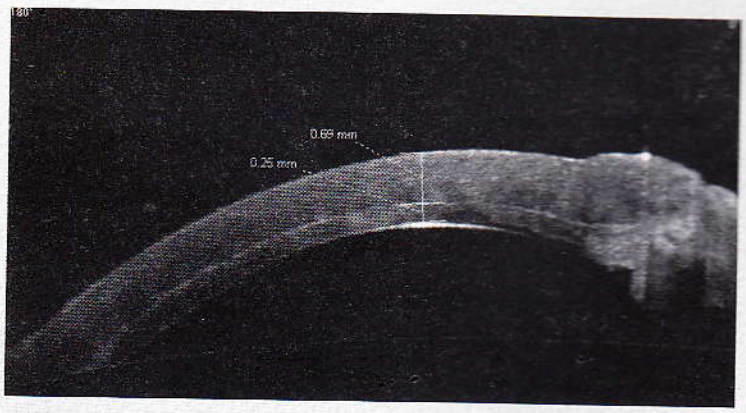
Anterior segment OCT photographs of right eye showing generalized thinning and globular protrusionof cornea

Based on following, clinical diagnosis of both eye keratoglobus was made.

Our plan of management in this patient was lamellar keratoplasty in form of tuck-inlamellar keratoplasty (TILK)[7]. A Hessburg-Baron vacuum trephine of 9.5 mm was centered and used to make an initial partial thickness groove of approximately 200 microns and anterior partial thickness lamellar dissection was done. 360 degree stromal pocket was created in peripheral cornea using a crescent knife. A full-thickness graft of10.0 mm diameter was punched, and the donor endothelium was scraped off. The peripheral part of the donorwas thinned from the posterior aspect to create a bevel-shapedflange in the periphery to complement the pocket created in thehost. The inner aspect of the donor button rim was held with aPierse Hoskins forceps and this was trimmed with the help of a fine-curved Vannas scissors to create a bevel or flange of approximately 1 mm width all around the rim. This button was placed overthe host bed. A paracentesis was done in the recipient cornea at the 9 o'clock position with a MVR blade to flatten the central dome ofthe host bed. The flange of the graft was tucked into the peripheralpocket of the host created previously, and the graft was suturedtightly with sixteen radial, interrupted, 10–0 nylon sutures.

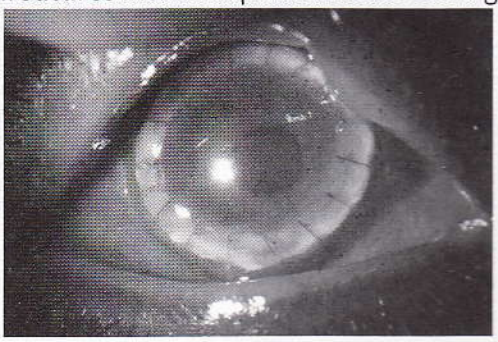
On postoperative day 1, patient was doing fine with vision of PL+ and PR accurate. Air bubble was in AC. Graft host junction was well apposed with all sutures intact. We started on prednisolone acetate 1% eye drop six times/day and ofloxacin 0.3% eye drop four times a day. Patient was asked to review after 1 week.

Clinical photograph at post op day 1

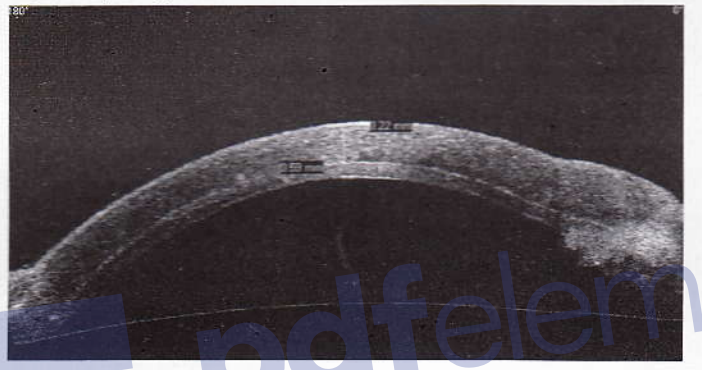


Anterior segment - OCT at post op day 1 showing graft thickness of 690 microns and bed thickness of 250 microns.

After 1 week, vision in right eye was counting fingers at 1m improving to 20/400 with pin hole. Graft host junction was well apposed with all sutures intact. Topical steroids were gradually tapered and patient was asked to review after 1 month.

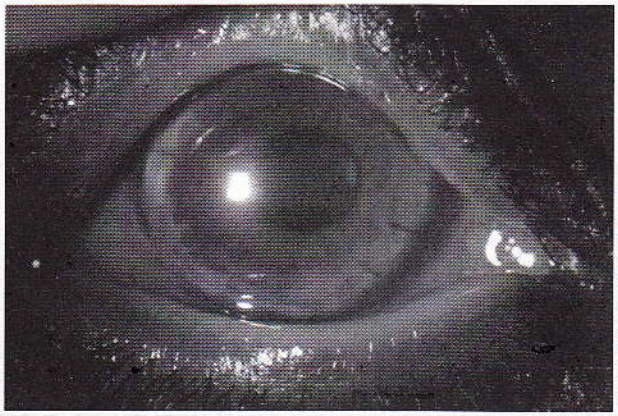


Clinical photograph at post op day 7.

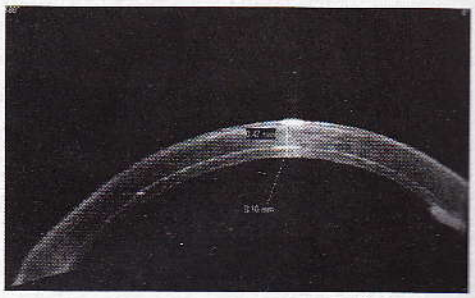


Anterior segment - OCT at post op day 7 showing graft thickness of 590 microns and bed thickness of 220 microns.

At 3 months follow up, best corrected visual acuity (BCVA) in right eye was 20/100 (+3.50 D sph / -6.00 D cyl @40°).



Clinical photograph at 3 months



Anterior segment - OCT at post op 3 months showing graft thickness of 470 microns and bed thickness of 160 microns

Discussion

Various methods for the management of keratoglobus has been described. Conservative management[8] includes patient counselling for use of eye protection because of thin cornea associated with high risk of perforation, spectacle wear to correct high myopia and astigmatism in the early stages. Contact lens fitting may be difficult because of corneal shape and itself is associated with risk of trauma inflicted by contact lens.

Surgical management includes penetrating keratoplasty, epikeratoplsty and lamellar keratoplasty[9]. Though penetrating keratoplasty is conventional, it is limited by peripheral graft host thickness disparity, large irregular astigmatism and poor visual outcomes. Taking large limbus to limbus corneal graft is an option but is limited by loss of immune privilege, limbal stem cell disruption and angle structure disruption.

The technique of tuck-in keratoplastyby Vajpayee et al, effectively reduces the corneal ectasia, not only centrally but also peripherally, and decreases the corneal-induced myopia and astigmatism and thus visually rehabilitates the patient in a single stage. It provides tectonic support, so that the structural integrity of the globe is maintained. Since no surgical intervention is done on the overlying limbal area or the angle, it preservesthe host stem cells and prevents any damage to the trabecularmeshwork of the host. Being an extraocular procedure, problems of graft rejection and vision-threatening complicationssuch as endophthalmitis are avoided.

References

- Cavara V. Keratoglobus and keratoconus: a contribution to the nosological interpretation of keratoglobus. Br J Ophthalmol 1950; 34: 621–626.
- Gregoratos ND, Bartsocas CS, Papas K. Blue sclerae with keratoglobus and brittle cornea. Br J Ophthalmol 1971; 55(6): 424–426.
- Pouliquen Y, Dhermy P, Espinasse MA, Savoldelli M. Keratoglobus. J Fr Ophthalmol 1985; 8(1): 43–45.
- Jacobs DS, Green WR, Maumenee AE. Acquired keratoglobus. Am J Ophthalmol 1974; 77(3): 393–399.
- Feder RS, Kshettry P. Noninflammatoryectatic disorders. In: Krachmer JH, Mannis MJ, Holland EJ (eds) Cornea: Fundamentals, diagnosis and management 3rd Vol. (Elsevier Mosby: New York, 2005, pp 955–974.
- Cameron JA. Keratoglobus. Cornea 1993; 12(2): 124–130.
- Vajpayee RB, Bhartiya P, Sharma N. Central lamellar keratoplasty with peripheral intralamellar tuck: a new surgical technique for keratoglobus. Cornea 2002; 21(7):657–660.
- Wallang BS, Das S. Keratoglobus.Eye (Lond).2013 Sep;27(9):1004-12.
- Lockington D, Ramaesh K. Use of a novel lamellar keratoplasty with pleat technique to address the abnormal white-to-white diameter in keratoglobus. Cornea 2015 Feb;34(2):239-42.