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Modified Surgical Epikeratoplasty for Keratoglobus: Anatomic and Visual Findings

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ABSTRACT

Introduction: Keratoglobus is a controversial issue and still remains as one of the ambiguous corneal disorders; it can be managed by different surgical techniques successfully; yet the risk of globe rupture is high in these patients due to corneal weakness. In cases of progressive involvement, a large corneal graft is needed due to extreme thinness of the cornea. Recent adventures in surgical techniques have led to the introduction of new managements particularly in Keratoglobus treatment such as the "tuck procedure" in which a 12 mm corneo-scleral graft is miniaturized at its peripheral margin. A pocket is formed at the limbus of the recipient and the donor graft is "tucked" into it. We present a case of epikeratoplasty in Keratoglobus management.

Case: A 45-year-old man with no history of a systemic disease underwent epikeratoplasty with the diagnosis of advanced bilateral keratoglobus since three years and six months before in the left and right eye, respectively. Pachymetry and best-corrected visual acuity (BCVA) were recorded before and six months after surgery. The highest recorded BCVA was 5/200 before the operation, whereas it was measured 6/10 (left eye) and 4/10 (right eye) at six-month follow-up examinations. Pachymetric findings improved from 244 to 773 and 212 to 744 (thinnest points) in the left and right eyes, respectively.

Conclusion: Epikeratoplasty is a relatively safe, effective and reversible extraoccular procedure in the management of keratoglobus. It can also be performed to flatten the cornea and protect it against acute corneal hydrops and perforation.

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Introduction

Keratoglobus is a minimally or non-progressive, noninflammatory corneal ectasia characterized by a spherical protrusion of the cornea with or without changes in corneal diameter and peripheral corneal thinning without any neovascularisation (1). The cornea is naturally clear; however, acute corneal edema can be caused by Descemet's membrane rupture and perforation even after minimal trauma (2). Keratoglobus is often bilateral and associated with several ocular conditions such as the blue sclera syndrome (3) leber's congenital amaurusis (4), pellucid marginal degeneration (5), orbital pseudotumor, chronic blepharitis, chronic eye rubbing and also post PK (1) Moreover, it also has comorbidity with joint hypermobility and consanguinity. The acquired form of this disorder has been reported to coincide with vernal kerato-conjunctivitis (1), hyperthyroidism (6), advanced

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keratopathy (keratoconus), trauma and exophthalmos (7). The etiology of the keratoglobus has remained unclear (3) and there are limited number of reports on the pertinents' genetic background in this regard (4).

Case presentation

A 45-year-old man was referred to our clinic. His left and right eyes were operated on three years and six months before, respectively. Prior to epikeratoplasty, a BCVA of 5/200 was recorded whereas pachymetric measurements summed up to 244 (left eye) and 212 (right eye). Corneal topography and keratometry were virtually impossible in both eyes because of the protrusion and severe corneal ecstasia. An informed consent was obtained from each patient after the nature of the surgical procedure was explained. This study was approved by Mashhad University of Medical Sciences Research Council Ethic Committee.

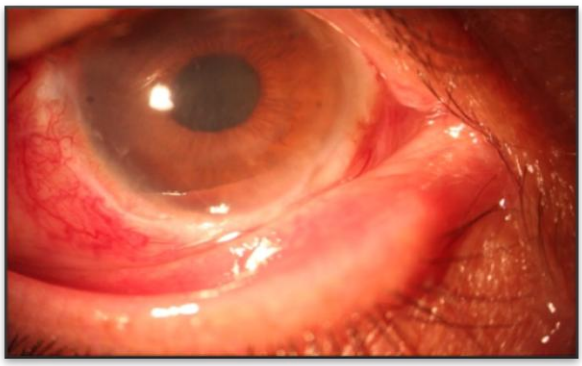


Figure 1: Right eye, six months after surgery

The operation was performed under general anesthesia, practicing intensive care to avoid trauma and perforation throughout the preparation while a single surgeon operated on both eyes. The donor's corneoscleral graft was prepared, cut to 50% of its original scleral thickness anteroposteriorly, the donor endothelium was also removed. The recipient's epithelium was removed thoroughly using alcohol and blade 15, followed by 360-degree peritomy.

A blade 15 degree was used to incise the limbus circumference and undermine the sclera, creating a 2 mm deep groove posteriorly to minimize trauma to the stem cells of the recipient.

In order to create a hypotonic globe, hence minimize the risk of corneal ectasia, aqueous humour was partially drained through a 1 mm incision posterior to the limbus using a 15-degree blade. Subsequently, the donor's cornea was stabilized on site and four temporary sutures were applied at 12, 3, 6 and 9 o'clock positions. The thinned sclera belonging to the donor was placed in the groove while any necessary recalibration was performed to remove the redundant tissue; therefore, it was possible to bring the sclera thoroughly inside the prepared groove. This procedure maximized epithelization as the limbus stem cells provided a full coverage. Finally, the graft was permanently fixed by applying 24 sutures (silk 8.0 or nylon 10.0) followed by attaching the conjunctiva to the limbus (vicryl 8.0).

After surgery, the patient was treated with oral stroid (Prednisolone 1 mg/kg for 2 weeks), topical steroid, antibiotics and frequent lubricant application. Follow-up examinations were performed after one and two weeks and again after one, three and six months for both eyes and twice each year thereafter for the left eye only.

Topical steroids (prednisolone acetate 1%) were prescribed after 3 to 6 months postoperatively to eliminate the chance of immature suture vascularization and loosening and to reduce the chance of graft rejection. Topical antibiotics were applied postoperatively by the time re-epithelization was complete (1 week post-surgery). It should be noted that post-operation managements were the same for both eyes and the sutures were removed at months 3 to 6, unless loosened or vascularized.

Figure 1, illustrates the right eye at the 6th month follow-up examination. As can be seen in this figure, there is a slight wrinkling of the recipient in the interface. At four month after the operation, there was a vascularization of the peripheral cornea at 6 and 12 o'clock in the left eye, therefore, injection of avastin was performed to induce complete regression. It should be noted that, no recurrence of vascularization was observed up to the time of reporting this study. Figure 2, shows the haziness of the corneal interface in the midperiphery and lipid deposition due to the regression of the vascularization.

Three years follow-up examinations for the left eye showed a pachymetry reading of 773 (thinnest point), whereas the same index was measured as 744 in the right eye after 6 months (Figures 3a-b), with a BCVA of 6/10 (left eye) and 4/10 (right eye).

Corneal re-epithelization was complete one week after the surgery.

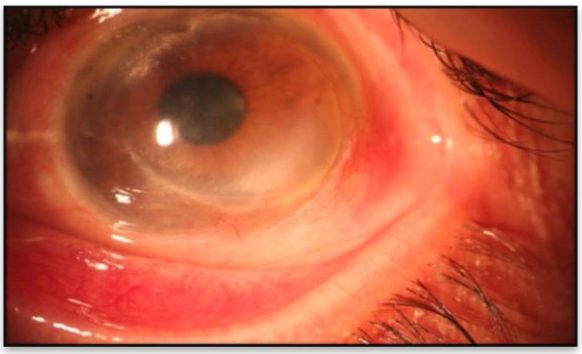
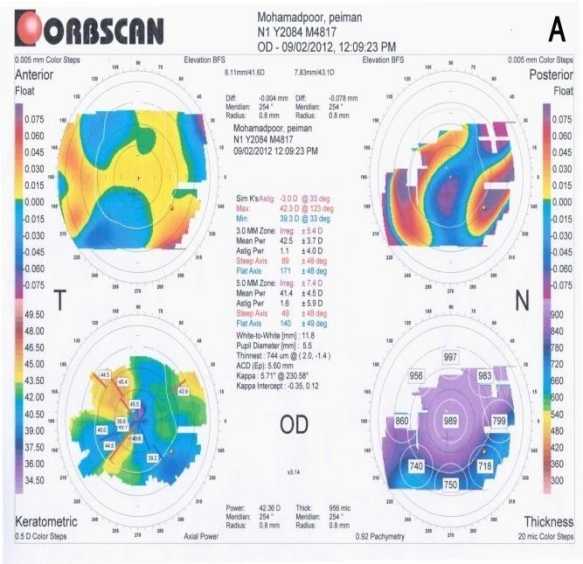


Figure 2: Left eye, three years post-surgery



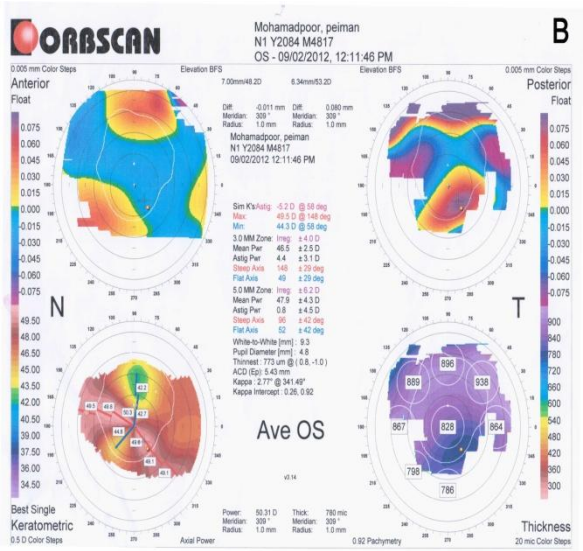


Figure 3a-b: Orbscan results a) right eye six month post-operation b) left eye three years post-operation

Discussion

Epikeratoplasty is a form of lamellar graft in which a human cornea created lens is attached to the anterior surface of the cornea which could result in some changes in the anterior curvature and the refractive power (8), this procedure has a number of advantages; mainly preservation of host endothelium, decreased graft rejection rate, normal post-operative activity, and minimum risk of traumatic dehiscence of the globe. In the current study, the donor corneal stem-cells were removed and the donor sclera was placed in the recipient's groove; therefore, immunosuppressive and long-term oral steroid treatments were not required.

However, it should be noted that in comparison with the penetrating keratoplasty (PK) technique, interface opacification and irregularity with astigmatism are more common in lamellar keratoplasty (9,10). Epikeratoplasty makes it possible to prevent adverse effects mainly corneal perforation, even in case of minor trauma, given the extreme thin cornea often seen in keratoglobus.

It is noticeable that spontaneous perforation is also common in keratoglobus, which may cause permanent eye loss, occasionally (11).

Unless BCVA is satisfactory, post-epikeratoplasty PK will also be possible. Common complications include incomplete corneal re-epithelization requiring bandage contact lens dressing plus tarsoraphy, and interface opacification due to epithelial overgrowth and recipient's corneal closed fold (12,13). In previous studies of Epikeratoplasty management, the corneoscleral graft size was 1 mm more than the recipient corneal diameter (12), or usually set as a fixed size of 12.5 mm (14). In the current study, corneoscleral graft was fixed with four temporary sutures on the recipient cornea and was adjusted in each quadrant relative to the recipient groove. We believe that the aforementioned procedure had a great impact on the accuracy of the graft adjustment and suturing and improved the corneal epithelization. However, the procedure is time-and labor-intensive, requiring full attention throughout the entire procedure particularly sclera groove making.

Our patient fortunately had acceptable corneal opacity and did not have any signs of graft rejection.

These features give more weight to epikeratoplasty as a treatment modality in keratoglobus patients before corneal perforation.

Acknowledgment

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