An Alternative Surgical Procedure for the Management of Keratoglobus

A. John Kanellopoulos, MD, and Lawrence H. Pe, MD†*

Purpose: To describe an alternative surgical technique in stabilizing peripheral corneal thinning in keratoglobus corneas with severe ectasia.

Methods: A 27-year-old man with keratoglobus and severe corneal ectasia in his remaining functional eye was managed by surgically placing an allograft corneoscleral ring around the limbus to support the mid-peripheral thinned cornea. Nylon 10-0 interrupted sutures were used only at the limbus. The allograft corneoscleral ring had an outer diameter of 14.5 mm and an inner opening of 7.0 mm. Conjunctiva was draped over and sutured to the donor corneoscleral ring graft.

Results: Cornea and vision are stable at 18 months' follow-up.

Conclusions: The peripheral suturing of a corneoscleral ring graft over the periphery of the cornea maybe an acceptable technique for providing tectonic tissue support and stabilizing eyes with keratoglobus.

Key Words: keratoglobus, allograft, bilateral corneal thinning, corneal ectasia

(*Cornea* 2005;24:1024–1026)

Keratoglobus is classified as one of the noninflammatory corneal ectatic disorders. It is a bilateral disease that is characterized by the globular protrusion of the cornea. This typical clinical appearance is the result of generalized corneal thinning that is most marked in the periphery.[1–3] In some cases, it is associated with abnormalities of connective tissue synthesis, such as seen in Ehlers-Danlos syndrome (EDS) type VI, the ocular-scoliotic form, and Rubinstein-Taybi syndrome.[4–6] Spectacle correction is usually the first step in the management of these patients and provides additional protection against trauma. A good contact lens fit is difficult to achieve and increases the likelihood of trauma to the eye. When functional vision cannot be obtained by these means or when the cornea appears to be at risk of rupture, surgery is undertaken. This can be done by means of a lamellar keratoplasty, epikeratoplasty, a wide-diameter penetrating keratoplasty, or a 2-stage procedure that initially involves a lamellar keratoplasty followed by a penetrating keratoplasty.[7–11]

CASE REPORT

A 27-year-old man of Cypriot descent presented with deteriorating vision on his right eye over the past 3 months and was not improving with spectacle correction. We made a diagnosis of keratoglobus based on the finding of a globar enlarged and thinned cornea coupled with clinical characteristics of EDS type VI (hyperextendable joints and skin). On examination, there were no signs of localized peripheral corneal thinning or a gutter that would lead us to suspect Terrien's marginal degeneration or other peripheral corneal thinning syndromes. This was the patient's only good eye since he had had 2 previous penetrating keratoplasties and retinal detachment surgery on his left eye, which, at the time of consult, had a poor visual prognosis and visual acuity of only hand movement.

On examination of the right eye, the best spectacle corrected vision (BCSVA) was 20/70, refraction −1.50 sph with −6.75 cyl × 77 degrees. The horizontal corneal diameter was 14.5 mm and had pachymetry measurements of 438 μm centrally and 250–544 μm peripherally. The cornea eye showed severe mid-peripheral thinning and ectasia (Fig. 1).

MATERIALS AND METHODS

Under peribulbar anesthesia, 360-degree conjunctival peritomy and superficial epithelial removal in the host right eye were performed. The epithelial removal was accomplished by using a Bard Parker no. 15 blade. This was done to expose a bed to which the graft could adhere to. An allograft corneoscleral ring was prepared by trephining the central 7 mm off a donor corneoscleral button with the Hanna trephine. Half of the deeper cornea was then manually removed in a 360-degree arcus from the inner ring opening using Westcott scissors to create an inner ring bevel. The outer diameter of the corneoscleral ring was sized to the host limbus (14.5 mm). A total of sixteen 10-0 nylon interrupted sutures (Alcon Laboratories, Forth Worth, TX) were placed in the outer corneoscleral rim to secure the allograft to the host sclera adjacent to the corneal limbus. All sutures were placed in the periphery of the donor corneoscleral ring and none on the inner rim to avoid any complications in the center of the cornea, which was still clear at this point. Following this, the previously recessed conjunctiva was draped and sutured over the graft (Fig. 2). A 24-mm diameter bandage contact lens (BCOR 9.20, plano) (Kelvin, Cantor and Nissel Ltd., U.K.) was placed over the eye, and the patient was given topical ofloxacin 0.3% (Exocin; Allergan, Westport, Ireland)

Received for publication June 8, 2004; revision received December 17, 2004; accepted December 18, 2004.

From the *LaserVision.gr Eye Institute, Athens, Greece; and †Manhattan Eye, Ear and Throat Hospital and Department of Ophthalmology, New York University Medical School, New York, New York.

Donor tissue provided by the Eye Bank for Sight Eye Restoration, New York, NY.

Reprints: A. John Kanellopoulos, MD, Laservision.gr Eye Institute, Pirgos Athinon, Mesogeion 2 & Vasilissis Sofias, Ampelokipoi 11527, Greece (e-mail: laservision@internet.gr).

Copyright © 2005 by Lippincott Williams & Wilkins

FIGURE 1. Right eye preoperatively shows globular cornea with areas of edema on the superior and nasal quadrants.

and prednisolone acetate 1% (Pred Forte; Allergan, Inc., Irvine, CA) QID for 1 month.

RESULTS

The patient is seen regularly and is at present in his 18th postoperative month on the right eye. The corneoscleral ring graft has taken well and the cornea appears stable (Fig. 3). BSCVA OD is 20/50 with a refraction of −2.00 sph combined with −6.00 cyl at 88 degrees.

DISCUSSION

Surgery for keratoglobus patients is often difficult because the peripherally thinned cornea is not amenable to a standard diameter (8.0–8.5 mm) penetrating keratoplasty. Previous authors advocate doing a large-diameter keratoplasty that is about 9.0 mm or more to include the abnormal cornea, but this is also prone to problems of damaging the anterior chamber angle and peripheral vascularization of the graft.[3,9]

Lamellar keratoplasty and epikeratoplasty have also been done in these cases to provide tectonic support and to address the optical quality as well. Some problems with this technique are the development of interface opacities and incomplete epithelialization.[8,9]

Others describe an intricate 2-stage approach that initially involves doing a lamellar graft or epikeratoplasty, and, subsequently, once the lamellar graft has stabilized (usually after a period of 4 to 6 months) is followed by a conventional diameter penetrating keratoplasty.[9–11] Thus, this later technique is technically difficult and uses 2 corneoscleral grafts per eye, one for the lamellar graft and another for the keratoplasty.

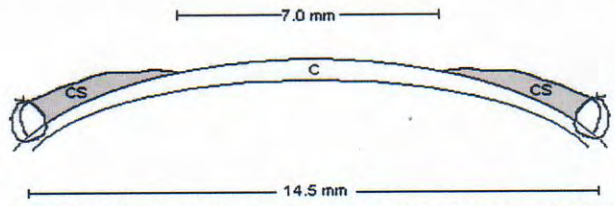

FIGURE 2. Cross-sectional diagram of eye with corneoscleral ring graft (CS). C, cornea.

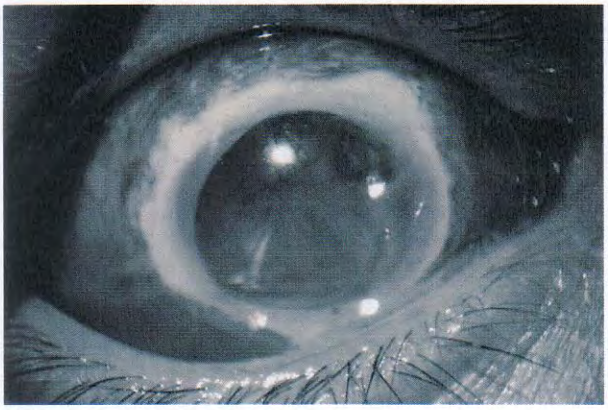

FIGURE 3. Right eye 18 months postoperatively.

It was our clinical assessment that there was impending spontaneous or traumatic rupture in this eye, and we decided to use an alternative technique, described to the best of our knowledge for the first time herein. Our purpose was to reinforce the mid-peripherally thinned cornea without the potential risk of central visual compromise in this functionally monocular patient that the traditional 2-step penetrating graft carries. This procedure would also carry little concern of allograft rejection. Furthermore, the central cornea is not disturbed during this procedure. It was our opinion that the corneoscleral graft had enough rigidity to support the peripherally thinned cornea without placing sutures in the inner rim of the graft. By securing this corneoscleral ring on the host sclera (the graft was sutured only to the host sclera), we believe that this halts the progressively "expanding" mid-periphery and periphery of the host cornea and also offers tectonic support over the dangerously thinned periphery. Finally, this technique is nonpenetrating and therefore carries less surgical risk.

The increase in visual acuity in this patient may be secondary to the flattening effect induced by the graft, especially on the thinned cornea on the superior-nasal quadrant resulting in less irregular astigmatism, although it did not directly reflect in a significant refractive change in the sphere and cylinder correction of the patient. There was also resolution of the corneal edema that was present in the preoperative period, which may have also contributed to the gain in visual acuity.

The patient has been advised to use protective eyewear at all times as this is his only remaining functioning eye, and although the procedure might have added tectonic support to the peripheral cornea, the danger of traumatic rupture still exists.

In summary, the placement of a tectonic mid-peripheral corneoscleral ring, as described, maybe an acceptable technique in stabilizing eyes with keratoglobus and possibly delay or even eliminate the need for a penetrating keratoplasty.

© 2005 Lippincott Williams & Wilkins

REFERENCES

1. Smolek MK, Klyce SD. Is keratoconus a true ectasia? An evaluation of corneal surface area. *Arch Ophthalmol.* 2000;118:1179–1186.
2. Feder RS. Non inflammatory ectatic disorders. In: Krachmer JH, Mannis MJ, Holland EJ, eds. *Cornea and External Diseases: Clinical Diagnosis and Management Volume II.* St. Louis: Mosby, 1997:1102–1103.
3. Wilson FM II. Congenital anomalies of the cornea and conjunctiva. In: Smolin G, Thoft RA, eds. *The Cornea. Scientific Foundations and Clinical Practice*, 3rd ed. Boston: Little, Brown; 1994:535–553.
4. Nelson ME, Talbot JF. Keratoglobus in the Rubinstein-Taybi syndrome. *Br J Ophthalmol.* 1989;73:385–387.
5. Cameron JA. Corneal abnormalities in Ehlers-Danlos syndrome type VI. *Cornea.* 1993;12:54–59.
6. Beers MH, Berkow R. eds. Musculoskeletal and connective tissue disorders. *In: Beers MH, Berkow R, eds. The Merck Manual, 17th ed. Merck & Co.* Inc., 1999:2404.
7. Vajpayee RB, Bhartiya P, Sharma N. Central lamellar keratoplasty with peripheral intralamellar tuck: a new surgical technique for keratoglobus. *Cornea.* 2002;21:657–660.
8. Cameron JA, Cotter JB, Risco JM, et al. Epikeratoplasty for keratoglobus associated with blue sclera. *Ophthalmology.* 1991;98:446–452.
9. Cameron JA. Keratoglobus. *Cornea.* 1993;12:124–130.
10. Jones DH, Kirkness CM. A new surgical technique for keratoglobus-tectonic lamellar keratoplasty followed by secondary penetrating keratoplasty. *Cornea.* 2001;20:885–887.
11. Macsai MS, Lemley HL, Schwartz T. Management of oculus fragilis in Ehlers-Danlos type VI. *Cornea.* 2000;19:104–107.

© 2005 Lippincott Williams & Wilkins