

Sclerokeratoplasty
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Cases

Case 1

40 year-old female from Peru presented to our Service with inflamed OS for 2 months duration. Her symptoms began as red painful OS, which progressively worsened with accompanying loss of vision. Initially, in Peru, the impression was that she had sclero-uveitis. However, oral and periocular prednisone, intra-muscular and topical Voltaren and oral cyclosporin did not improve her symptoms. Scleral biopsy, done in Peru, showed necrotizing scleritis and culture of the sclera grew *Pseudomonas*. She was then she was started on intra-venous ceftazidime and amikacin. Her condition continued to deteriorate.

The patient was referred to our Service at this time.

Past Ocular History: Pterygium excision of right eye

Past Medical History: Sacroiliitis

Vision 20/20 OD, CF OS

No afferent-pupillary defect

Slit Lamp Examination

Right Eye - within normal limits

Left Eye - as seen

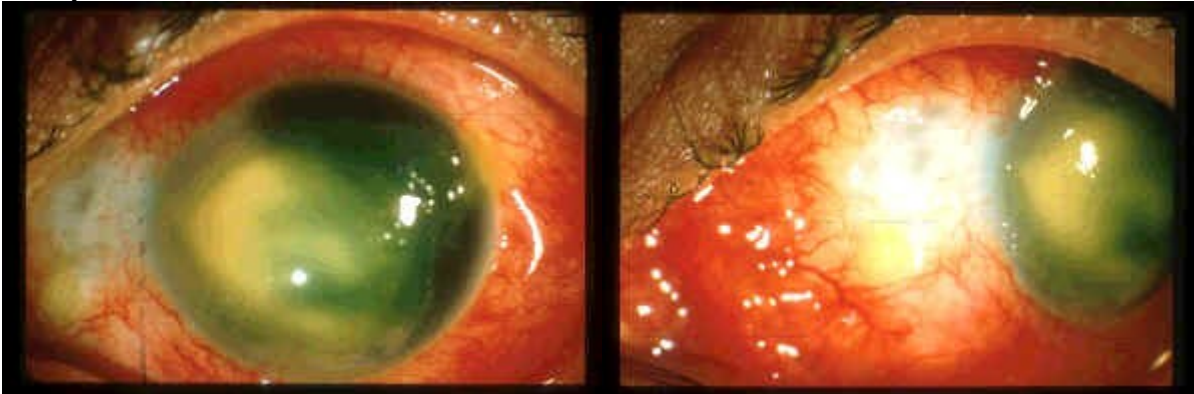


Fig 1

Fig 2

Serology: +ANA : Hep2 1:80 Speckled; Rat Liver 1:80 Speckled

Anterior chamber tap, scleral biopsy and culture: all Negative

The patient's condition continued to worsen.



Fig 3

Sclerokeratoplasty was performed for therapeutic and diagnostic purposes.



Fig 4

PCR of cornea: positive for HSV

The patient was started on intra-venous acyclovir

Graft remained clear at 4 weeks

Vision 20/60 OS

Maintained on oral acyclovir, Pred-forte and artificial tears

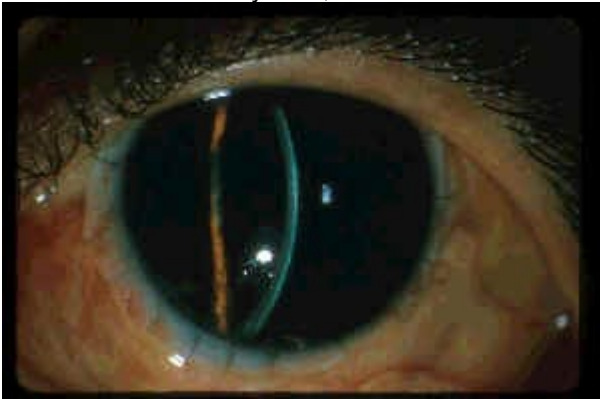


Fig 5

Case 2

72 year-old female referred for evaluation of corneo-scleral melt of OS. She has 40 years history of rheumatoid arthritis, with recent increase in joint activity. Patient also has history of HSV keratitis in OS and had three corneal transplants in the past.

Systemic Meds:

Methotrexate 15mg/wk sub-cutaneous

Naprosyn 375mg 3 times a day

Acyclovir 200 mg 5 times a day

Vision 20/25 right eye, LP left eye

Slit Lamp Examination

Right eye - 2+ nuclear sclerosis cataract, otherwise within normal limits

Left eye - as seen



Fig 6
Perforation was noted and glue application with bandage contact lens placement was done.



Fig 7
The patient was begun on fortified antibiotics.

Work up of the immune status revealed the following values.

ESR = 74
CRP = 4.0 (0-0.5)
sIL-2r = 1000 (0-820)
RF = 24.1 (0-20)

The clinical condition worsened, and she was started on intra-venous cyclophosphamide pulse. Sclerokeratoplasty was performed.



Fig 8



Fig 9

Discussion

Definition

Sclerokeratoplasty is the use of a corneoscleral graft of varying size, shape and position to end or improve those treatment resistant diseases of the cornea that resulted in dissolution of tissue and the production of an anatomic defect that is incompatible with continued survival of the globe. This definition is adopted from Taylor and Stern's paper from 1980.

Historical background

Sclerokeratoplasty was first described by Girard in 1956. At the meeting of the Pan-American Association of Ophthalmology, he presented 12 cases of sclerokeratoplasty he had performed. In the literature, however, Barraquer is usually given the credit for describing the surgery. In 1961, Barraquer published an article about his experience with the procedure, which he called a variation of total penetrating keratoplasty. In Taylor and Stern's paper in 1980, they described their experience with 50 cases of reconstructive keratoplasty, a few of which were sclerokeratoplasty. In 1998, Panda et al. published 2 articles of the use of sclerokeratoplasty for treatment of refractory corneal ulcer and anterior staphyloma.

Indications

The goals of the procedure are three fold. The procedure can maintain or provide structural integrity of the globe. It can remove or debulk the diseased corneal and scleral tissue. And when needed, the removed tissue can be used for diagnostic purpose.

Clearly, this is a radical surgery that should not be taken lightly. Indications for the procedure are listed here: Infection (bacterial, viral, fungal or parasitic); trophic ulcers; severe collagen vascular disease such as our second case where the cornea and sclera were melting relentlessly; severe dry eyes to the extent of severe damage to anterior segment; and obviously trauma. In addition any condition that results in damage to anterior segment that cannot be repaired by ordinary means.

Technique

The technique is straightforward. Complete conjunctival peritomy is performed. Then excision of the entire cornea, and if necessary, the surrounding sclera is performed. Reformation of the angle may be needed in some cases. Secure the donor graft including a 1 to 2 mm rim of sclera.

Potential Post-op Complications

Possible post-operative complications are glaucoma, hypotony, graft rejection, continued disease process, and refraction error

Glaucoma after sclerokeratoplasty is likely due to alteration or damage to the angle structures. Zimmerman et al. advocated use of full thickness suture to reduce posterior gaping of the wound, which can lead to secondary angle closure. Taylor and Stern recommended cyclodialysis when it was still performed as the procedure for glaucoma. Cohen, Kenyon and Dohlman described using iris tightening suture as a technique to prevent anteriorization of the iris and subsequent angle closure in PK, which can be applied here. Cobo et al performed lamellar dissection of the sclera in both the host and the donor, thus avoiding the trabecular meshwork during the surgery.

Hypotony is also a potential complication, but is less frequent compared with elevated intraocular pressure. Some of these eyes have sustained damage, which results in ciliary shutdown before the surgery. And obviously sometimes, there can be wound leak.

Report of frequency of graft rejection is variable. In the original 12 patients described by Girard, he described all 12 as having opaque cornea from rejection within one year, but he stressed that all of the eyes were viable. In contrast, in Panda et al series of ten patients only one had a rejection episode, over one year post-op, which was successfully treated.

Recurrence of the underlying condition can be a problem because of the medical condition or the infection is poorly controlled. The transplanted tissue is subjected to the same condition and likely will have the same consequence, if the underlying condition is not treated. In the study of sclerokeratoplasty for refractory bacterial ulcers, by Panda et al., patients received concomitant broad spectrum systemic and topical antibiotics, and none of the 10 patients had recurrence of their ulcer.

Refraction, though a minor problem for patients with these severe problems, can still be debilitating in the long run. The difficulty with refraction lies in the inability to predict the correction after surgery. Interestingly, astigmatism did not seem to be a major concern, perhaps due to the rigidity of the graft provided by the scleral rim.

Prognosis

Prognosis after sclerokeratoplasty is good, especially when considering the alternatives: endophthalmitis, enucleation, evisceration, or phthisis. Patients may require further interventions, but the eyes generally remain viable. Important factors in the prognosis are the underlying conditions and their treatment.

Summary

Sclerokeratoplasty is a justifiable procedure, when the ocular condition is so severe that other treatment modalities appear futile. Post-operative complications must be anticipated and steps should be taken to prevent them. Most importantly, the underlying condition must be treated concomitantly.