Case Report

Deep Anterior Lamellar Keratoplasty and Peripheral Lamellar Keratoplasty for a Case of Severe Peripheral Ulcerative Keratitis

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Keywords

Deep anterior lamellar keratoplasty · Keratoplasty · Peripheral lamellar keratoplasty · Peripheral ulcerative keratitis · Rheumatoid arthritis

Abstract

We experienced a rare case of severe peripheral ulcerative keratitis in a patient undergoing surgery combined with deep anterior lamellar keratoplasty (DALK) and peripheral lamellar keratoplasty (LK). A 63-year-old Japanese woman was referred to our hospital for the treatment of visual disturbance caused by peripheral ulcerative keratitis in the left eye. Although the inflammation subsided with topical and oral administration of steroids, peripheral ulcerative keratitis worsened 4 weeks after the medical treatment. Surgery combining DALK and peripheral LK, including the corneal limbus, was performed as treatment. Two weeks after the surgery, a double anterior chamber appeared, but it disappeared spontaneously. There was no postoperative rejection or intraocular pressure elevation. One year and 6 months after the surgery, the inflammation did not recur, the cornea remained transparent, and the thickness of the cornea was maintained. In conclusion, combined DALK and peripheral LK may be a surgical option for treating severe peripheral ulcerative keratitis.

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Introduction

Peripheral ulcerative keratitis (PUK) is characterized by progressive ulceration of the limbal region of the cornea, causing an epithelial defect, progressive stromal degradation, and thinning [1]. In severe cases, accompanying scleritis, particularly the necrotizing form, is observed [2]. PUK progresses and involves most of the corneal circumference, ultimately including the central cornea, and causes marked corneal edema [1, 3]. Serious complications of PUK include corneal perforation and loss of vision [4, 5].

Medical management with immunosuppressants plays a significant role in the initial stages of treating PUK [3]. However, progressive cases require surgical management. Various surgical modalities have been described for managing PUK, such as resection of the perilimbal conjunctiva, lamellar grafting, and amniotic membrane transplantation [5, 6]. Complex surgical methods are required in severe cases with a high risk of corneal perforation [6]. We herein report our successful surgical results for a case of severe PUK undergoing surgery combined with deep anterior lamellar keratoplasty (DALK) and peripheral lamellar keratoplasty (LK).

Case Report

A 63-year-old Japanese woman was admitted to our hospital complaining of visual impairment, conjunctival redness, and irritation of her left eye. She presented with worsening pain and redness in her left eye during the previous 6 months. Her visual acuity was 10/200 in the right eye and 4/200 in the left eye, and the intraocular pressures (IOPs) were 15 and 20 mm Hg, respectively. A slit-lamp examination of the left eye showed peripheral corneal thinning extending from 8 to 4 o'clock and necrotizing anterior scleritis of adjacent sclera (Fig. 1a). Rubeosis iridis associated with inflammation was observed in the iris, and hyphema was observed in the anterior chamber. The left fundus was invisible due to hyphema and mature cataract. The right fundus examination revealed a myopic choroidal neovascularization leading to central vision loss. No pathogens or viruses were detected by the smear test or polymerase chain reaction from the lesion, and the culture results were negative.

The inflammatory workup revealed elevated erythrocyte sedimentation rate (45 mm/h; normal range <15 mm/h) and C-reactive protein (2.1 mg/dL; normal range <0.3 mg/dL). The serologic workup revealed elevated rheumatoid factor (26 U/mL; normal range <15 IU/mL) and anti-nuclear antibody (160 U/mL; normal range <40 U/mL). The remainder of her serologic (ANCA, anti-SS-A/RO, and anti-SM-A/RO) and infectious workup was negative.

She consulted a rheumatologist and confirmed joint symptoms. She was diagnosed as rheumatoid arthritis based on blood tests and joint symptoms. The diagnosis of RA was based on clinical findings according to the American College of Rheumatology/European League Against Rheumatology (ACR/EULAR) classification criteria. She was diagnosed as PUK associated with rheumatoid arthritis.

Following this diagnosis, the patient was treated with topical prednisolone acetate 1% eye drops 4 times daily. Prophylactic topical antibiotic eye drops and oral 25 mg steroid daily were added. The immunosuppressant could not be used because it was under scrutiny for liver dysfunction. Two weeks after the medical treatment, the inflammation seemed to have subsided. However, 4 weeks after the treatment, the ulcer base had widened and deepened with significant thinning. Necrotizing anterior scleritis spread along the affected cornea (Fig. 1b). Marked edema appeared in the central cornea. Although there was no perforation of the cornea, the nasal peripheral cornea had exposed Descemet's membrane (Fig. 1c). Immediate surgical treatment was required to prevent perforation.

Fig. 1. Before surgical treatment. **a** Slit-lamp photograph showing peripheral corneal thinning and necrotizing anterior scleritis in her left eye at a visit to our hospital. **b** Anterior segment optical coherence tomography image obtained at a visit to our hospital. **c** Severe corneal thinning and central corneal edema were noted 4 weeks after the treatment. **d** Anterior segment optical coherence tomography image obtained 4 weeks after the treatment. There was no perforation of the cornea, but the nasal peripheral cornea had exposed Descemet's membrane.

The patient underwent surgery of combined DALK and peripheral LK including the corneal limbus. We made a 2/3 layer deep host corneal incision using a 9.5-mm Beaver visitec's manual corneal trephine blade, shifting from the center to the temporal side, where the cornea was mildly thinned. A deep peripheral corneal ulcer was found in the range of 120° around the entire cornea, so the incision with the trephine was made in the range of 240°. Then, a layered incision was made to expose Descemet's membrane. The corneal stroma containing the corneal ulcer was resected in a layer of the same depth from the temporal side to the nasal, and the Descemet's membrane was exposed all around the cornea. The adjacent necrotic anterior sclera was also resected from the nasal side to the upper part of the corneal limbus. Katena's BARRON vacuum donor corneal punch 9.5 mm was used to prepare the donor cornea. A round 9.5-mm donor cornea with the corneal endothelium peeled off prepared with trepan was sutured end-to-end with 10-0 nylon at 240° with the corneal limbus on the temporal side aligned with the margin. For the remaining crescent-shaped recipient corneal defect and necrotizing anterior sclera, a freehand crescent-shaped graft was prepared from the remaining donor graft according to the size of the defect. A circular 9.5-mm

Fig. 2. Surgical findings. **a** We made a 2/3 layer deep host corneal incision using a 9.5-mm corneal trephine blade, shifting from the center to the ear. **b** Layered incision was made to expose Descemet's membrane. **c** Donor corneal punch 9.5 mm was used to prepare the donor cornea. **d** A round 9.5-mm donor cornea was sutured end-to-end with 10-0 nylon at 240° with the corneal limbus on the ear side aligned with the margin. **e, f** A freehand crescent-shaped graft was prepared from the remaining donor graft according to the size of the defect. **g–i** A round 9.5-mm donor cornea, a freehand crescent-shaped graft, and host sclera were sewn together at 120° using 10-0 nylon thread (**a–i**).

donor cornea, a freehand crescent-shaped graft, and host sclera were sewn together at 120° using 10-0 nylon thread. The combined suture was carried out a total of 10 threads. We confirmed the junction of the lamellar crescent-shaped graft and the DALK graft under a normal operating microscope because there was no microscope equipped with anterior segment OCT. At the end of surgery, air was injected into the anterior chamber to prevent double anterior chamber. The patient was fitted with Bausch & Lomb Inc.'s Medilist therapeutic contact lenses, and the operation was completed (Fig. 2a–d).

From the day after the operation, topical prednisolone acetate 1% eye drops 4 times daily, ofloxacin eye ointment once daily, oral 30 mg steroid daily, and cyclosporine 100 mg daily were started while monitoring liver dysfunction. The edema of the donor cornea was dense in the early postoperative period, but it gradually decreased. As the edema reduced, the sutures became loose, and there was a slight gap between the adhesive parts of both transplanted corneas. Two weeks after the surgery, a double anterior chamber appeared, but it disappeared spontaneously. One year and 6 months after the surgery, the inflammation had not recurred, the cornea remained transparent, and the thickness of the cornea had been maintained (Fig. 3a, b). Her visual acuity improved to 20/200 at the time of 1 year and 6 months after surgery, and IOP was controlled without glaucoma eye drops in her right eye.

Discussion

Peripheral corneal ulcers are usually associated with systemic autoimmune diseases, including rheumatoid arthritis [1, 7]. Rheumatic corneal ulcer is caused by tissue destruction of the immune complex to the corneal limbus and conjunctiva due to collagen disease [1].

Fig. 3. After surgical treatment. **a, b** A decentered 9.5-mm DALK and LK including the corneal limbus were performed in her left eye. Slit-lamp photograph and anterior segment optical coherence tomography image were taken the day after she was operated on. **c, d** One year and 6 months after surgical treatment. The inflammation did not recur, and the cornea remained transparent. The thickness of the cornea was maintained.

Idiopathic peripheral corneal ulcer is not associated with collagen disease. It develops due to a specific autoimmune reaction in the ocular region [7]. In both cases, an arcuate corneal ulcer develops in the peripheral part of the cornea. Tissue destruction occurs in the surrounding corneal limbus and sclera, resulting in their thinning.

Various surgical modifications have been described for peripheral corneal ulcers. The design of the transplanted cornea varies depending on the range and size of the perforation site and the thinning site [6]. The most common surgical technique is crescentic LK. However, in cases with extensive thinning of the corneal ulcer, melting necrosis of the sclera, and marked edematous opacity in the center of the cornea, resection of the perilimbal conjunctiva, lamellar grafting, and amniotic membrane transplantation are not appropriate for the treatment. Penetrating keratoplasty may be required in severe cases. When the transplanted tissue in penetrating keratoplasty is large, there is a high risk of postoperative rejection and IOP elevation due to gonio angle obstruction [8, 9].

In the present case, tissue destruction was observed not only in the peripheral cornea but also in the corneal limbus and the sclera at the first visit to our hospital. Therefore, conjunctival resection was considered difficult, and medical treatment was selected. Topical and systemic steroid therapy temporarily reduced the area of the lesion. However, decreased systemic steroid therapy exacerbated the lesion. We considered using immunosuppressants from the beginning of treatment, but immunosuppressants were not available early in treatment because she was being tested for unexplained liver dysfunction.

The tissue destruction including the cornea and sclera was irregular. So, freehand preparation of one large corneal graft was considered likely to cause postoperative poor adhesion. We therefore felt that the best way to fill the defective region of the recipient cornea and sclera with a good-sized implant was by dividing the graft into 2 parts.

We chose DALK to maintain transparency of the central cornea after surgery. A decentered 9.5-mm DALK was performed along the corneal limbus of the temporal side. To reconstruct the remaining corneal defect and necrotizing anterior sclera, LK including the corneal limbus was performed. Although vascular invasion was observed in the surface layer of the lamellar graft after surgery, no postoperative complications were observed with regard to the DALK graft. There have been few reports of DALK during the active phase of keratitis. Fungal keratitis that was poorly responsive to medical treatment was reported to be treated by this operation [10, 11]. DALK reportedly has a high degree of difficulty in surgical procedures. There have been issues such as Descemet's membrane perforation occurring during surgery and postoperative double anterior chamber. However, compared to penetrating keratoplasty, there are also major benefits with this approach, such as a reduction in rejection and the suppression of intraocular pressure elevation [12, 13]. In penetrating keratoplasty of large grafts, adhesion of the peripheral iris is reportedly more likely to cause angle obstruction and increase the intraocular pressure. Although the present case included transplantation of a large graft, we performed the DALK procedure, so the angle was maintained, and the IOP did not increase after the surgery.

In this case, a marked edematous change was observed in the center of the cornea, and we were wondering whether this was due to corneal endothelial dysfunction or inflammation associated with PUK. Measurement of corneal endothelial cell density was difficult due to corneal edema. We have clinically experienced cases of strong edema in the center of the cornea in PUK, which are found in cases with extensive lesions in the sclera and cornea. For corneal edema, the transparency of the cornea improves again when the treatment for PUK is successful. The pathophysiology of PUK is thought to be related to the deposition of immune complexes in the corneal limbus and vasculitis. We attributed corneal edema to inflammation of the stroma, not to corneal endothelium damage in this case. Therefore, anterior lamellar keratoplasty was performed to preserve the corneal endothelium of the host.

The major problem that occurred in the present case was the development of double anterior chamber 2 weeks after the surgery. The small gap between the two grafts in the early postoperative period may have contributed to the onset of this phenomenon. Stromal edema of the transplanted cornea is seen early after transplantation. However, when the edema disappears over time, loosening of the suture appears. The accompanying temporary gap formation may have contributed to the formation of the double anterior chamber. The regeneration of the corneal epithelium likely repaired the gap, and the double anterior chamber disappeared.

In conclusion, the present case was successfully treated for severe PUK with surgery combining DALK and peripheral LK. This operation may be a surgical option for managing melting nonperforated extensive peripheral corneal ulcer with marked edema in the center of the cornea.

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Statement of Ethics

The study adhered to the tenets of the Declaration of Helsinki for research involving human subjects. Written informed consent was obtained from the patient for the publication of this case report and any accompanying images. This study protocol was approved by the Institutional Review Board of Oita University Hospital, Approval No. 2199.

Conflict of Interest Statement

The authors have no conflicts of interest to declare.

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Author Contributions

This work was carried out in collaboration among all authors. K.Y. and R.N. drafted the manuscript. K.Y., R.N., and T.O. collected the data. K.Y. and R.N. reviewed the literature. K.Y. and K.K. interpreted the data and critically reviewed the manuscript. T.K. critically reviewed the final version of the manuscript. All authors read and approved the final manuscript.

Data Availability Statement

All data generated or analyzed during this study are included in this article. Further enquiries can be directed to the corresponding author.

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