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Case Report

Simultaneous Vitreoretinal Surgery and Sclerokeratoplasty for Keratoglobus with Intraocular Hemorrhage and Extensive Corneal Rupture

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Keywords
25G pars plana vitrectomy · Sclerokeratoplasty · Simultaneous surgery · Keratoglobus · Corneal rupture

Abstract
We reported a case of simultaneous vitrectomy and sclerokeratoplasty (SKP) performed for keratoglobus with extensive corneal rupture and intraocular hemorrhage caused by trauma. A 73-year-old woman was treated for keratoglobus and glaucoma. She was punched in both eyes, her right eye showed corneal rupture and the left eye showed prolapse of the ocular contents due to eyeball rupture. She immediately underwent corneal sutures in the right eye and resection of the prolapsed ocular contents in the left eye at a nearby ophthalmological clinic. Three days after the injury, the patient was referred to our clinic for vision recovery. The best corrected visual acuity of the right eye was measured by counting fingers. Her right eye presented severe corneal edema with a sutured corneal wound in the upper periphery, which was positive in the Seidel test. B-mode ultrasound revealed choroidal detachment and subchoroidal hemorrhage. Fourteen days after injury, simultaneous corneal suture and posterior sclerotomy were performed in the right eye, but corneal fragility and corneal opacity were prominent, and B-mode examination revealed prolonged vitreous hemorrhage and retinal detachment. Twenty-one days after injury, we performed simultaneous SKP and 25-G pars plana vitrectomy (PPV). In this procedure, we initially performed SKP followed by 25-G PPV without a keratoprosthesis or endoscope. The visibility of the fundus through the corneoscleral

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graft was good during vitrectomy. Three months after surgery, her corrected visual acuity improved to 10/1,000. Although there was mild corneal stromal edema and khodadoust line, there were no obvious fundus complications. Simultaneous SKP and PPV for keratoglobus with extensive corneal rupture and vitreous diseases may be a good option.

Introduction

Corneal opacities often interfere with the diagnosis and management of posterior segment pathology [1]. In patients with coexisting corneal and vitreoretinal disorder, the use of a temporary keratoprosthesis or endoscopy is necessary to allow intraoperative fundus visualization when performing vitrectomy [2]. However, these techniques have intraoperative complications and surgical limitations, and endoscopic surgery has problems with postoperative fundus management with severe corneal opacity [3, 4]. As a safer and less invasive method, simultaneous vitrectomy with deep anterior lamellar keratoplasty [5] or Descemet Stripping Automated Endothelial Keratoplasty [6] has been reported. We have previously reported simultaneous 25G vitrectomy with PKP (Penetrating Keratoplasty) for the patient with vitreoretinal disease and severe corneal opacity [7].

Keratoglobus is a rare noninflammatory corneal disorder characterized by generalized thinning and globular protrusion of the cornea [8]. The advanced corneal thinning causes the cornea to bulge forward and even the slightest trauma poses a risk of perforation [9–11]. Surgical closure in these cases is difficult because of the extreme thinness of the cornea that leads to cut through of the sutures and inability for wound closure [8]. Various surgical techniques for keratoglobus with perforation have been proposed, but there is no consensus on an optimal method [12].

Sclerokeratoplasty (SKP) is an anterior segment reconstruction surgery that uses a large corneoscleral graft [13]. Corneal perforation caused by severe whole corneal infection and autolysis adjacent to the sclera are indications for SKP [14–16]. SKP can be required in cases where infected tissue is present in the cornea limbus and allows for reliable reconstruction of the anterior segment [17]. In keratoglobus, SKP is generally performed to avoid corneal perforation and improve visual acuity [18].

With the development of wide-angle vision systems and chandelier endoillumination, it has become possible to fully visualize the retina during vitreoretinal surgery, making the procedure safer and more effective, even in eyes with small pupils and corneal opacity [19, 20]. Chandelier illumination improves anterior chamber visualization in cataract surgery with severe corneal opacity or vitreous hemorrhage [21]. The development of vitreoretinal surgical instruments has expanded the indications for microincisional vitrectomy for serious retinal diseases, such as proliferative diabetic retinopathy and proliferative vitreoretinopathy [7]. Here, we report a case of simultaneous SKP and 25-gauge pars plana vitrectomy (25G PPV) for keratoglobus with extensive corneal rupture and vitreous hemorrhage caused by trauma.

Case Report

A 73-year-old woman being treated for keratoglobus and glaucoma was punched in both eyes, and experienced corneal rupture in her right eye and prolapse of the ocular contents of her left eye. She immediately underwent corneal suturing in the right eye and resection of the

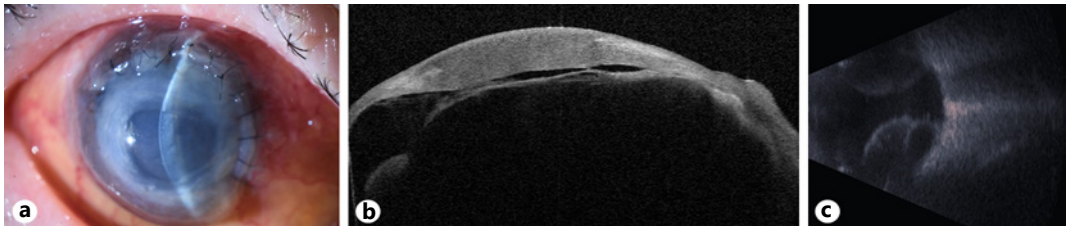

Fig. 1. Slit-lamp microscopy, anterior segment optical coherence tomography, and B-mode ultrasound examination for the right eye before the surgery. **a** Corneal stromal edema and sutured corneal wound in the upper periphery were noted. **b** The anterior chamber was extremely narrow (sagittal view). **c** Abnormal findings of choroidal detachment and subchoroidal hemorrhage were revealed (sagittal view).

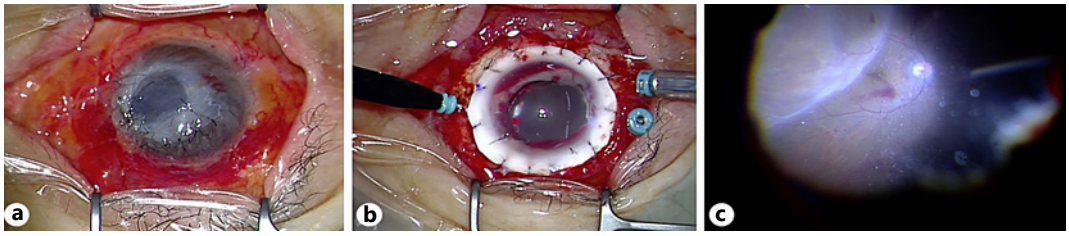

Fig. 2. Intraoperative images of the anterior segment and fundus. **a** Severe corneal edema and sutured corneal wound in the upper periphery. **b** After corneoscleral transplantation, we performed 4-channel 25G PPV with chandelier illumination. **c** Choroidal detachment was observed in the temporal mid-peripapillary region. The fundus visualization through the graft was good.

prolapsed ocular contents in the left eye at a nearby ophthalmological clinic. After 3 days, she was referred to our department because of insufficient corneal wound closure and subchoroidal hemorrhage in her right eye.

On her first visit, the best corrected visual acuity (BCVA) was at the level of counting fingers in her right eye and no light sensation in her left eye. Intraocular pressure (IOP) was 2 mm Hg in the right eye. Images obtained from the patient are shown in Figure 1. Slit-lamp microscopy showed severe corneal edema and the sutured corneal wound in the upper periphery, which was positive for the Seidel test. Fundus examination could not be performed because of the corneal opacity and intraocular hemorrhage. B-mode ultrasound revealed abnormal findings suggestive of choroidal detachment and subchoroidal hemorrhage. Initially, we started treatment with antimicrobial therapy and expected spontaneous wound closure and hemolysis of the subchoroidal hemorrhage. The subchoroidal hemorrhage gradually hemolyzed; however, the corneal wound did not close. Fourteen days after the injury, posterior sclerotomy was performed in the right eye to improve subchoroidal hemorrhage. Intraoperatively, corneal resuturing was performed because of leakage from the corneal wound. B-mode ultrasound revealed suspected retinal detachment and choroidal detachment 3 days after surgery. We required both vitrectomy and anterior segment reconstruction that could tolerate vitrectomy. Twenty-one days after injury, we performed simultaneous SKP and 25G PPV.

Surgery was performed under general anesthesia. We initially performed SKP followed by 25G PPV (Fig. 2). The host cornea was incised along the keratolimbus at 360°. The donor cornea was incised in the sclera, approximately 1 mm beyond the keratolimbus. The graft was sutured with 16 interrupted 10-0 nylon sutures. After corneoscleral transplantation, we performed 4-channel 25G PPV with chandelier illumination. We used a Constellation 25G system (Alcon, Hünenberg, Switzerland) and a wide-angle viewing system (Resight noncontact

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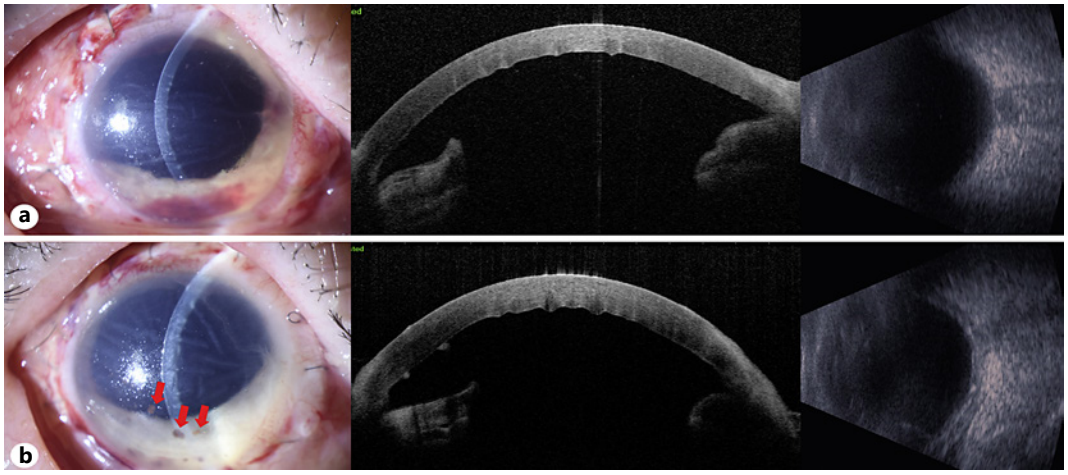

Fig. 3. Slit-lamp microscopy, anterior segment optical coherence tomography, and B-mode ultrasound examination for the right eye after the surgery. **a** Seven days after surgery. Corneal stromal edema was observed, the anterior segment of the eye was reconstructed, and the anterior chamber was sufficiently deep. There were no abnormal fundus findings (sagittal view). **b** Three months after surgery. A khodadoust line and corneal stromal edema were found in the graft. There were no abnormal fundus findings (sagittal view).

wide-angle lenses; Carl Zeiss Meditec, Jena, Germany) to observe the fundus. The visibility of the fundus during vitrectomy was improved without using temporary keratoprosthesis or endoscopy. After core vitrectomy and peripheral vitrectomy, the clot attached to the retina was removed using intralimiting membrane forceps (Alcon). Choroidal detachment was observed in the temporal mid-peripapillary region, but no obvious retinal tears or retinal detachments were observed. At the end of surgery, the graft was sutured with an additional two interrupted 10-0 nylon sutures because of an aqueous humor leakage between the graft and sclera.

Images obtained from the patient 7 days and 3 months after the surgery are shown in Figure 3. The BCVA of her right eye improved to 20/1,000, and IOP was elevated to 16 mm Hg. The shape of the anterior eye was reconstructed. Due to mild corneal stromal edema, the number of corneal endothelial cells could not be measured and the fundus was obscured. B-mode ultrasound revealed no abnormal findings. As a postoperative management for SKP, we used intravenous 0.4% betamethasone for 4 days postoperatively. Eye drops of 0.1% betamethasone sodium phosphate (Betamethasone Sodium Phosphate PF Ophthalmic and Otorhinologic Solution 0.1%; Nitten Pharmaceutical, Nagoya, Japan) and 0.3% gatifloxacin hydrate (GATIFLO OPHTHALMIC SOLUTION 0.3%, Senju Pharmaceutical, Osaka, Japan) were used four times daily. At 3 months postoperatively, the BCVA of her right eye was 10/1,000, and IOP was 2 mm Hg. A khodadoust line was found in the graft, we used 2% cyclosporin eye drops four times daily. The number of corneal endothelial cells could not be measured. B-mode ultrasound revealed no abnormal findings in the fundus. Currently, although there is mild corneal stromal edema, there are no obvious vision loss or fundus complications.

Discussion

In this case, we performed simultaneous SKP and 25G PPV for keratoglobus with extensive corneal rupture and intraocular hemorrhage. To achieve postoperative visual function by improving severe corneal opacity and performing vitrectomy in a closed state, we performed

simultaneous SKP and PPV surgery. Intraoperative intraocular translucency through the corneoscleral graft was good. The BCVA of her right eye improved to 10/1,000, and the ocular shape was maintained after surgery. Currently, there are no obvious complications associated with the fundus. Our results suggest that simultaneous SKP and 25G PPV may be an option for the treatment of keratoglobus with extensive corneal rupture and intraocular hemorrhage caused by trauma to achieve rapid visual rehabilitation.

In the case of coexisting vitreoretinal disease and severe corneal opacity, some solutions are needed to visualize the fundus during operation [22]. The use of an ophthalmic endoscope [2] and temporary keratoprosthesis have been previously reported. Depending on the location of the corneal opacity, simultaneous vitrectomy with PKP [4] or deep anterior lamellar keratoplasty [5] and Descemet Stripping Automated Endothelial Keratoplasty [6] has been reported. Simultaneous vitreous and corneal surgery should be considered when fundus visualization is sufficient to allow vitrectomy [23]. We have previously reported simultaneous PKP and PPV without the use of endoscopes and keratoprosthesis in a patient with severe corneal opacity and retinal disease [7] Intraoperative fundus visualization was good, the postoperative corneal graft remained transparent, and there was no recurrence of retinal disease [7]. In this case, intraoperative fundus visualization with a wide-angle viewing system through the corneoscleral graft was sufficient and did not interfere at all with the performance of vitreoretinal surgery.

Various surgical techniques have been proposed for the management of keratoglobus, including epikeratoplasty [8], central lamellar keratoplasty with peripheral intralamellar tuck [24], limbal stem cell-sparing lamellar keratoplasty [12], use of a corneoscleral rim over the thinned corneal periphery [25], conventional penetrating keratoplasty [26], SKP [27], a 2-step tectonic lamellar keratoplasty followed by secondary penetrating keratoplasty [28], and limbal stem cell-sparing corneoscleroplasty with peripheral intralamellar tuck [11]. When corneal perforation occurs in keratoglobus, the perforation wounds are commonly severe due to the fragility of the thinned cornea [29]. If extensive perforation occurs in the peripheral cornea, as in our case, SKP may be preferred because suturing the graft to an intact sclera avoids cutting through the sutures. This allows for reliable anterior segment reconstruction.

SKP is an effective treatment for diseases of the entire cornea, but postoperative immunologic rejection occurs frequently and is a challenge [13]. Rejection after corneoscleral transplantation occurs in approximately 70% of cases, mostly between 2 weeks and 1 month after surgery [14]. This procedure is associated with a high risk of immunologic rejection due to antigen-presenting cells and vascular endothelial cells in the donor limbus [30]. Topical treatments for immunologic rejection include 1% prednisolone acetate, 0.1% fluorometholone acetate, 0.1% dexamethasone, and cyclosporin A, and systemic treatment is rarely used [31]. We use 0.1% betamethasone acetate eye drops, 2% cyclosporin eye drops, and cyclosporin oral drops as routine treatment after SKP in our hospital. However, in this case, only 0.1% betamethasone acetate eye drops were used four times daily postoperatively. At 3 months postoperatively a khodadoust line was found, 2% cyclosporin eye drops were used for four times daily. In the event of rejection, prompt diagnosis and initiation of treatment are important.

Simultaneous SKP and PPV for keratoglobus with extensive corneal rupture and intraocular hemorrhage may be good options because of reliable anterior segment reconstruction and good intraoperative fundus transparency. Simultaneous surgery allows for early vision rehabilitation with minimal invasion.

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Statement of Ethics

Written informed consent was obtained from the patient for publication of this case report and any accompanying images. This study protocol was exempted from the need for approval by the Institutional Review Board of Yamaguchi University Hospital.

Conflict of Interest Statement

The authors report no conflicts of interest in this work.

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Author Contributions

Fumiaki Higashijima designed the study and wrote the initial draft of the manuscript. Kazuhiro Kimura was corresponding author and contributed to analysis and interpretation of data, and assisted in the preparation of the manuscript. Ren Aoki, Masanori Mikuni, Takuya Yoshimoto, Nanako Iwamoto, Manami Ohta, Tadahiko Ogata, and Naoyuki Yamada have contributed to data collection and interpretation, and critically reviewed the manuscript. All authors approved the final version of the manuscript and agreed to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.

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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