Combination of phototherapeutic keratectomy and wavefront-guided photorefractive keratectomy for the treatment of Thiel–Behnke corneal dystrophy

Chia-Chieh Hsiao, Yu-Chih Hou

Phototherapeutic keratectomy (PTK) has been used to treat anterior corneal dystrophy but may cause hyperopic shift or corneal high-order aberration. We describe the successful treatment of a case of a 43-year-old female with Thiel–Behnke corneal dystrophy, using PTK followed by wavefront-guided photorefractive keratectomy (PRK). The patient had high corneal aberration and hyperopic shift after PTK in both eyes. The corneal surface regularity index and surface asymmetric index increased after PTK and decreased after wavefront-guided PRK. Vision improved from preoperative 20/400 and 20/60 to postoperative 20/20 and 20/25 in the right eye and the left eye, respectively. Additional wavefront-guided PRK may enhance the effects of PTK by reducing irregular corneal surfaces and by correcting PTK-induced hyperopic shift.

Key words: Corneal dystrophy, phototherapeutic keratectomy, wavefront-guided photorefractive keratectomy

Corneal dystrophies are bilateral, inherited, noninflammatory corneal opacities. The mutation of the transforming growth factor β-induced (*TGFBI*) gene may cause Thiel–Behnke corneal dystrophies (TBCDs).[1] Stage-related management of corneal dystrophies is advocated.[2] The management includes the use of lubricants, therapeutic soft contact lenses, phototherapeutic keratectomy (PTK), superficial lamellar keratectomy (LK), or penetrating keratoplasty (PK).[3,4] However, some patients may develop hyperopic shift or irregular corneal surfaces after PTK. This study reports the use of wavefront-guided photorefractive keratectomy (PRK) for the treatment of high-order aberrations and hyperopic shift after PTK in a TBCD patient. This procedure was rarely reported previously.

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Case Report

A 43-year-old female had a history of progressive blurred vision and recurrent corneal erosions since childhood. She presented with superficial honeycomb opacities in both eyes in May 2012 [Fig. 1a and b]. She and her two daughters had an Arg555Gln mutation in the *TGFBI* gene. The best-corrected visual acuities (BCVAs) were 20/200 with a refraction of +1.5 −0.75 × 30° in the right eye and 20/60 with a refraction of +0.25 −2.5 × 30° in the left eye. Central corneal thickness was 570 μm in the right eye and 600 μm in the left eye. Anterior segment-optical coherence tomography (AS-OCT) showed the highest opacity at the anterior cornea, about 150 μm in both eyes. Corneal topography (TMS-4, Tomey Corporation, Nagoya, Japan) showed surface regularity index (SRI) values of 1.18 and 1.34 and surface asymmetric index (SAI) values of 2.67 and 1.97 in the right and left eyes, respectively [Fig. 2a and b]. In August 2012, superficial LK and PTK were performed to remove the most superficial opacities in the right eye. Surgical techniques of LK employed 64 Beaver blade to remove epithelial and subepithelial opacities under topical 0.5% proparacaine anesthesia.[4] To remove the residual superficial stroma opacities and smooth corneal surface, the following PTK was performed with the excimer laser machine (Technolas 217Z, Bausch and Lomb, Rochester, NY, USA). The laser was applied with an ablation diameter of 7.0 mm and an ablation depth of 80 μm. During the procedures, hypromellose (0.32%) was applied to smooth corneal surface irregularities. Mitomycin-C (0.02%) solution was applied for 10 s at the end of the surgery. Postoperatively, topical fluorometholone (0.1%), levofloxacin ophthalmic solution (0.5%), and hypromellose (0.32%) eye drops were administered four times daily. The histology of the removed corneal specimen showed a misalignment of epithelial cells and subepithelial deposition of fibrous tissue; these observations were consistent with the histological findings in TBCD. Postoperatively, most opacities of the right eye were removed, and the vision improved to 20/40 with a refraction of +4.00 −2.24 × 30° [Fig. 1c]. However, the patient experienced a mild glare, and the SRI and SAI increased to 1.52 and 3.68, respectively [Fig. 2c]. Ocular aberrometry was performed with an iDesign aberrometer (Advanced WaveScan aberrometer; Abbott Medical Optics, Santa Ana, CA, USA), and wavefront-guided PRK was performed using the VISX Star S4 IR excimer laser platform (Abbott Medical Optics, Santa Ana, CA, USA) with a 10 s application of 0.02% mitomycin-C solution.

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The laser treatment profile used the aberrometric correction of +4.45 −2.38 × 30° with an ablation of 6.0 mm for the optical zone and 1.5 mm for the transition zone. At postoperative month 4, the vision improved to 20/25 with a refraction of −0.50 −1.50 × 50° without glare. The postoperative SRI and SAI of the right eye decreased to 1.02 and 1.18, respectively. As the vision and glare of the right eye showed improvement, wavefront-guided PRK was repeated 5 months later. The postoperative vision improved to 20/20 with a refraction of −1.0, and corneal opacity decreased [Fig. 1e]. The SAI and SRI further reduced to 0.61 and 0.91, respectively [Fig. 2e].

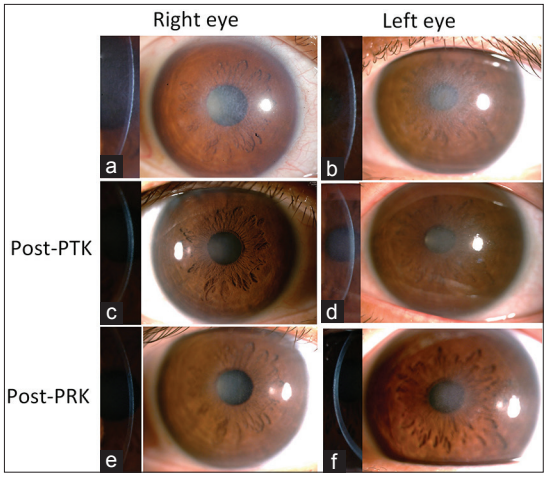

Figure 1: (a and b) Slit-beam photography and the insets demonstrate superficial honeycomb opacities in both eyes. (c and d) Corneal opacities much decreased after phototherapeutic keratectomy. (e and f) Less opacity was left after subsequent wavefront-guided photorefractive keratectomy

In April 2015, superficial LK and PTK were performed in the left eye with an ablation depth of 120 µm and an ablation zone of 6.0 mm. The left cornea became clearer; the BCVA was 20/50 with a refraction of +7.75 −0.50 × 10° postoperatively [Fig. 1d]. The post-PTK SRI and SAI increased to 1.35 and 3.12, respectively [Fig. 2d]. Due to a high hyperopic shift, wavefront-guided PRK was performed at 5 and 12 months postoperatively. Corneal opacity decreased, especially in the mid-peripheral area, and vision improved to 20/25 with a refraction of +0.75 −1.0 × 80° [Fig. 1f]. The SAI and SRI decreased to 1.07 and 1.89, respectively [Fig. 2f]. AS-OCT showed that most superficial opacities were removed in both eyes; the central corneal thicknesses were 436 and 431 µm in the right and left eyes, respectively. To date, no complication was found in our case after PTK and wavefront-guided PRK except the recurrence of faint superficial opacity in the right eye. The vision was still 20/20 in the right eye and 20/25 in the left eye. Since the visual acuity of the patient was still satisfactory, further treatment can be deferred. Similar PTK and wavefront-guided PRK settings could be repeated in the future if needed.

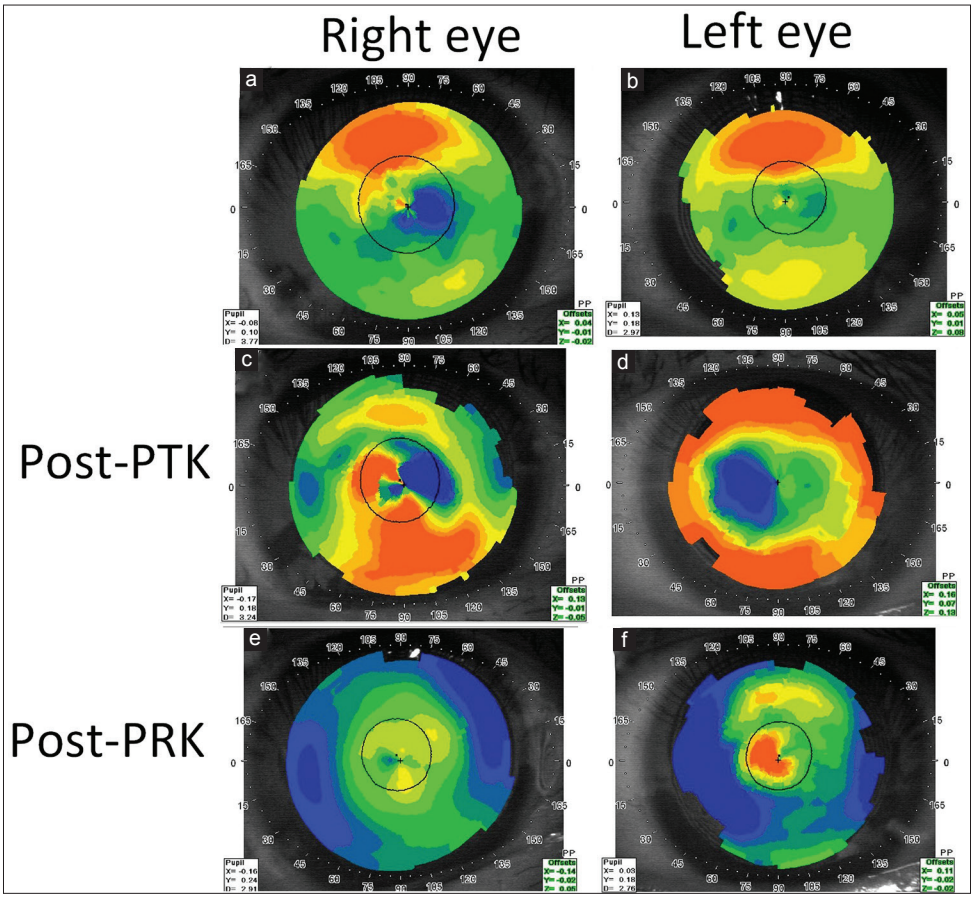

Figure 2: Corneal topography. (a and b) Mild irregular surface in both corneas. (c and d) Irregularity of corneal surface increased after phototherapeutic keratectomy. (e and f) Corneal surface became smooth and regular after wavefront-guided photorefractive keratectomy

Discussion

Corneal opacity in TBCD presents as a diffuse honeycomb shape and is located in the Bowman layer and anterior stroma. TBCD is usually caused by an Arg555Gln mutation in the *TGFBI* gene.[5] Histopathology shows an undulating fibrous tissue in the subepithelium and focal disruption of the Bowman's membrane. The genetic study and histologic findings in this patient were indicative of TBCD. PTK is a less-aggressive and relatively safer procedure with shorter recovery time as compared to LK and PK. The results of PTK for the treatment of anterior corneal dystrophies are promising.[6,7] Hieda *et al*. showed good outcomes of PTK surgery for TBCD in the mean follow-up period of 60 months.[3] PTK combined with superficial LK may achieve a similar effect of PTK while providing histological specimens for clinical diagnosis clarification.[4] AS-OCT demonstrates the depth and extent of corneal opacities, which helps to determine the proper ablation depth and diameter for PTK in corneal dystrophies.[8]

However, PTK may induce hyperopic shift and affect visual outcomes, especially after deep ablation. These complications may lead to high-order aberration and result in vision with glare or halo. Wavefront-guided ablation has been used to treat patients with high-order aberrations in refractive surgery.[9] We used wavefront-guided PRK to reduce surgically induced high-order aberrations and correct hyperopic shifts after PTK; vision, refraction, and corneal surface regularity showed significant improvement in this TBCD case. Simultaneous PRK and PTK have been used to treat high astigmatism alongside radial keratotomy and keratoplasty.[10] In our procedure, the accuracy of the wavefront examination may be enhanced after the removal of superficial corneal opacities by the initial PTK; by following up with wavefront-guided PRK, favorable results may be achieved. This procedure may take a long recovery time and needs a further study to evaluate its efficacy.

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Conflicts of interest

There are no conflicts of interest.

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