

Sir, TGFBI-linked corneal dystrophies treated using superficial lamellar keratectomy combined with phototherapeutic keratectomy

Corneal dystrophy may impair vision and cause recurrent corneal erosion. Phototherapeutic keratectomy (PTK) has been reported to treat patients with *TGFBI*-linked corneal dystrophies. ^{1–5} However, PTK could not provide a specimen for histopathological examination. We reported the clinical and histopathological characteristics of three patients with *TGFBI*-linked corneal dystrophies treated using superficial lamellar keratectomy and PTK.

Case report

Patient 1 had Thiel-Behnke corneal dystrophy (TBCD)-like reticular corneal opacities (Figure 1a). Patient 2 had a lattice corneal dystrophy (LCD) type 1-like corneal haze with translucent branching lines (Figure 1b). Patient 3 had atypical LCD-like whitish polymorphic opacities with filamentous gray lines (Figure 1c). Superficial lamellar keratectomy was performed for five eyes of the three patients by using No. 64 Beaver blade and excimer laser ablation.

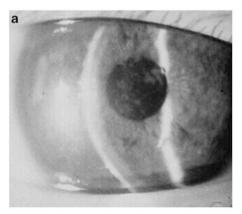
Photophobia and ocular irritation decreased postoperatively. Corneal topography showed improvement in the surface regularity index and surface asymmetric index of all five eyes after surgery (Table 1). Visual acuity improved in patients 1 and 2 but remained unchanged for patient 3. Therefore, penetrating keratoplasty (PK) was performed 1 year later for patient 3.

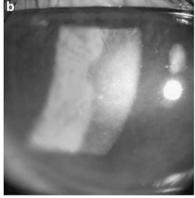
Histological examination showed vacuolization of the basal epithelium and undulating fibrous tissue interposed between the irregular epithelium for patient 1 (Figure 2a), corresponding to TBCD; irregularly thick epithelium and subepithelial amyloid deposits for patient 2 (Figures 2b and c); some focal subepithelial

Table 1 Results of eyes of corneal dystrophy patients treated with superficial lamellar keratectomy and phototherapeutic keratectomy

Eyes	Pre-treatment				Post-treatment			
	VA	AveK	SRI	SAI	VA	AveK	SRI	SAI
Patient 1 (OD)	20/40	44.85	4.38	1.96	20/25	42.95	2.42	0.72
Patient 1 (OS)	20/40	44.60	4.34	1.0	20/25	43.55	1.08	0.91
Patient 2 (OD)	20/60	40.10	5.8	1.89	20/20	38.68	0.8	0.8
Patient 2 (OS)	20/50	39.60	6.23	3.85	20/20	39.04	0.79	1.6
Patient 3 (OS)	20/100	44.30	1.19	3.15	20/100	43.12	0.89	0.85

Abbreviations: AveK, average keratometry; SAI, surface asymmetric index; SRI, surface regularity index.





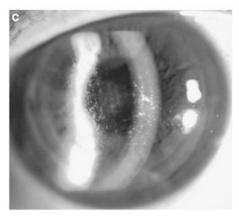


Figure 1 Slit-lamp photography showed homogenous, fine honeycomb-shaped opacities in the Bowman's layer area and superficial stroma of patient 1 (a). Numerous linear, branching, and ropey opacities were observed in the central superficial cornea of patient 2 (b). Numerous polymorphic whitish opacities with chipped-ice appearance extended deep into the stroma in patient 3 (c).

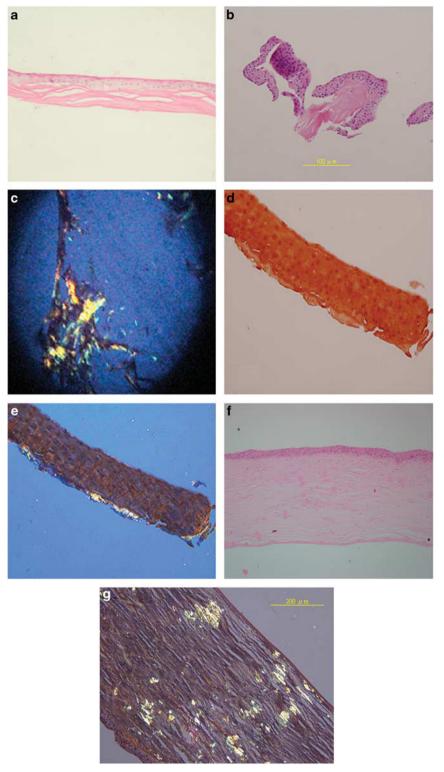


Figure 2 Superficial lamellar keratectomy specimen from patient 1 showed numerous vacuolated basal cells and sawtooth-like fibrous tissue interposed between irregular epithelium (a, periodic acid-Schiff staining, \times 400). Corneal specimen from patient 2 showed an irregular corneal epithelium with variable thickness and subepithelial deposition of eosinophilic and amorphous materials (b, hematoxylin and eosin staining, \times 200), which exhibited apple-green dichroism on Congo red staining under polarized microscopy (c, Congo red staining, \times 400). Lamellar keratectomy corneal specimen from patient 3 showed subepithelial amyloid deposition (d, Congo red staining, \times 400), which exhibited apple-green birefringence on Congo red staining under polarized microscopy (e, Congo red staining, \times 400). Corneal specimen obtained from patient 3 by penetrating keratoplasty showed amyloid opacities extending into the entire stromal layer (f, hematoxylin and eosin staining, \times 200; g, Congo red staining, \times 400).



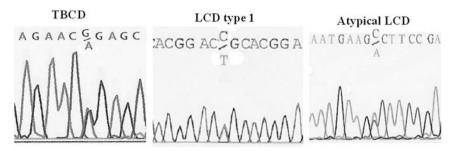


Figure 3 Sequencing analysis of *TGFBI*. Analysis of the sequence mutations showed a heterozygous arginine-to-glutamine substitution at nucleotide 1711 of codon 555 in exon 12 of patient 1; a heterozygous cytosine-to-thymine substitution at nucleotide 417 in exon 4 of patient 2; and a heterozygous cytosine-to-adenosine substitution at nucleotide 1637 in exon 12 of patient 3.

amyloid in keratectomy specimen (Figures 2d and e); and numerous amyloid deposits throughout the stroma in the PK specimen (Figures 2f and g) for patient 3. Genetic analysis showed Arg555Gln mutation in patient 1, Arg124Cys mutation in patient 2, and Ala546Asp mutation in patient 3 (Figure 3).

Comment

Lamellar keratectomy combined with PTK can obtain specimen for histological examination and smoothen corneal surface to reduce irregular astigmatism. The correlation of the histological finding with clinical features and genetic study could help in accurate diagnosis of *TGFBI*-linked corneal dystrophies.

Conflict of interest

The authors declare no conflict of interest.

References

- 1 Dinh R, Rapuano CJ, Cohen EJ, Laibson PR. Recurrence of corneal dystrophy after excimer laser phototherapeutic keratectomy. Ophthalmology 1999; 106: 1490–1497.
- 2 Inoue T, Watanabe H, Yamamoto S, Maeda N, Inoue Y, Shimomura Y et al. Recurrence of corneal dystrophy resulting from an R124H Big-h3 mutation after phototherapeutic keratectomy. Cornea 2002; 21: 570–573.
- 3 Ellies P, Bejjani RA, Bourges JL, Boelle PY, Renard G, Dighiero PL. Phototherapeutic keratectomy for BIGH3-linked corneal dystrophy recurring after penetrating keratoplasty. *Ophthalmology* 2003; **110**: 1119–1125.
- 4 Gruenauer-Kloevekorn C, Braeutigam S, Froster UG, Duncker GI. Surgical outcome after phototherapeutic keratectomy in patients with TGFBI-linked corneal dystrophies in relation to molecular genetic findings. *Graefes Arch Clin Exp Ophthalmol* 2009; 247: 93–99.
- 5 Szentmáry N, Seitz B, Langenbucher A, Schlötzer-Schrehardt U, Hofmann-Rummelt C, Naumann GO. Histologic and ultrastructural changes in corneas with granular and macular dystrophy after excimer laser phototherapeutic keratectomy. Cornea 2006; 25: 257–263.

Y-C Hou¹, C-H Hsiao², K-T Kuo², I-J Wang¹ and F-R Hu¹

¹Department of Ophthalmology, National Taiwan University Hospital, College of Medicine, National Taiwan University, Taipei, Taiwan ²Department of Pathology, National Taiwan University Hospital, College of Medicine, National Taiwan University, Taipei, Taiwan E-mail: fungronghu@ntu.edu.tw

Eye (2012) **26**, 170–172; doi:10.1038/eye.2011.267; published online 4 November 2011

Sir

A day at the races: ocular injuries in extreme motorcycle racing spectators

The risk of injury in extreme motorcycle racing is well established. ^{1,2} We report two cases of ocular injuries to spectators without eye protection.

Case report

A 14-year-old boy (patient A) sustained blunt trauma to the left eye, caused by a small stone sprayed by a passing racer while at a motocross race. Visual acuity (VA) was 1.0 logMAR in the left eye. Examination showed anterior dislocation of the lens with mild vitreous haemorrhage. A vitreolensectomy was performed and the patient is awaiting a secondary anterior chamber lens implant. Fundoscopy revealed a choroidal tear temporal to the optic disc with pigment stippling at the macula (Figure 1). Corrected VA in the left eye is currently 0.6 logMAR.

A 50-year-old man (patient B) sustained blunt trauma to the left eye by a mechanism similar to the above case. VA at presentation was 0.5 logMAR in the left eye. He had a hyphaema and superior iridodialysis. Fundoscopy revealed a traumatic retinal dialysis with an associated nasal (macular attached) retinal detachment (Figure 2). He underwent a left pars plana vitrectomy, encirclement, 360° endolaser, and gas tamponade with 20% C2F6. Post-operatively he developed an epiretinal membrane with macular pucker and cataract. Present vision is 0.9 logMAR in the left eye and he is awaiting cataract surgery. The patient was a heavy goods vehicle (HGV) driver and the injury and visual loss have resulted in the removal of his HGV licence.

Comment

Motocross takes place on an outdoor track containing natural terrain with human-made obstacles. Injuries among riders in the sport are high. Even paramedical staffs attending to the injured racers on track are advised to wear