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Femtosecond laser-assisted keratoplasty combined with cataract extraction in a patient with keratoconus and oculocutaneous albinism

Dorottya Pásztor, Bence Lajos Kolozsvári, Gergely Losonczy, Mariann Fodor

In this study, we present a case of a 58-year-old male patient with oculocutaneous albinism, keratoconus, total cataract, and glaucoma originating from father-daughter incest. He underwent femtosecond laser-assisted keratoplasty with "open-sky" cataract extraction and posterior chamber intraocular lens implantation. One week after surgery his uncorrected visual acuity improved from hand motion to 20/200. Six months later corneal *K* values were 49.1 D in the flat and 50.0 D in the steep meridian. The graft had a central corneal thickness of 488 µm and was well fitted. The patient's quality of life improved substantially due to the surgery. To the best of our knowledge, this is the first report on the association of albinism with advanced keratoconus, total cataract, and glaucoma. Moreover, no previous report on femtosecond laser-assisted keratoplasty using VisuMax femtosecond laser system with "open-sky" cataract extraction is available in the literature. The VisuMax femtosecond laser-assisted keratoplasty ensures fast patient rehabilitation in such challenging cases.

Key words: Father-daughter incest, femtosecond laser assisted-keratoplasty, keratoconus, oculocutaneous albinism, "open-sky" cataract extraction

Keratoconus is a progressive distorting corneal disorder with an incidence of 1 in 500–1 in 2000. The disease is mostly sporadic; however, autosomal dominant and recessive cases have also been reported.[1]

Oculocutaneous albinism (OCA) is an autosomal recessive disorder with multiple ocular manifestations such as photophobia, refractive errors, pendular nystagmus, transilluminable iris, foveal hypoplasia with significantly reduced visual acuity (in the range of 20/60–20/400) and abnormal decussation of the optic nerve fibers.[2] Corneal thickness shows correlation with skin pigmentation: Lighter skin (especially albino) pigmentation associates to the thicker cornea.[3]

Children originating from father-daughter incest have high-risk of genetic diseases. There are only two ophthalmological cases published so far in relation to incest.[4,5]

To the best of our knowledge, there has only been one case of OCA associated with keratoconus and cataract reported so far, however, without any surgical treatment.[6] Our albino patient originating from a father-daughter incest, underwent femtosecond laser-assisted keratoplasty combined with "open-sky" cataract extraction, and posterior chamber intraocular lens (PC-IOL) implantation.

Case Report

A 58-year-old hyperopic Caucasian male patient with OCA Type 1 was referred to our department with progressive binocular visual disturbance. The patient originated from a father-daughter incest. The visual acuity was hand motion on both eyes. Slit lamp examination showed extremely advanced keratoconus features and total cataract in both eyes [Fig. 1]. On palpation, the eye felt undoubtedly firm owing to elevated intraocular pressure (IOP). Steep *K* value of the right eye was 79.3 D, and the thinnest point was 193 µm (Pentacam HR, Oculus Optikgeräte GmbH, Germany). The fundus could not be visualized in either eye due to total cataract. The axial length was 20.9 mm and 21.3 mm in the right and left eye, respectively. Normal IOP was achieved with combined antiglaucoma eye drops (latanoprost, timolol-brimonidine, brinzolamide).

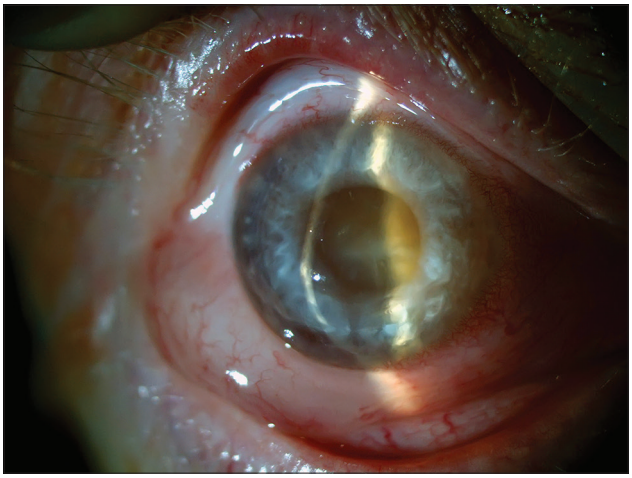

Figure 1: Slit lamp examination shows an extremely out bulging cornea among other advanced keratoconus features (thinning of the cornea, stromal scarring) and total cataract

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Because both cataract and keratoconus accounted for the decreased visual acuity, a triple procedure was performed. Visual improvement was unpredictable due to glaucoma and albinism.[7] All procedures adhered to the Declaration of Helsinki. Written consent was obtained from the patient for the publication of his case and images. The patient underwent femtosecond laser-assisted keratoplasty (VisuMax, Carl Zeiss Meditec, Germany) combined with "open-sky" cataract extraction and PC IOL implantation (+29.0 D AMO Sensar AR40e, Germany) on the right eye performed by an experienced surgeon (Mariann Fodor) [Video 1]. Intraoperative data of the femtosecond laser-assisted keratoplasty (VisuMax, Carl Zeiss, Germany) were the following: The interface type of the recipient cornea was M and the donor cornea was L; the posterior cut diameter was 6.4 mm and 6.7 mm in the recipient and the donor corneas, respectively; the cut thickness of the recipient cornea was 770 µm and the donor cornea was 820 µm; the side angle of cut was 90° both in the recipient and the donor corneas. Postoperative management consisted of the continuation of antiglaucoma therapy along with antibiotic and steroid eye drops. Postoperative the IOP could be measured by applanation tonometry. The postoperative course was uneventful. The patient had a complete ophthalmological evaluation every month during the half-year follow-up. One week after surgery the uncorrected distance visual acuity (UCDVA) improved to 20/200 and remained unchanged and uncorrectable throughout follow-up. The graft showed good fit and centration with anterior segment optical coherence tomography. Funduscopy revealed a pale optic disc and foveal hypoplasia in the right eye. Pentacam data are shown in Table 1. Both *K* and pachymetry values improved in the first postoperative month and remained unchanged thereafter. K_1 value altered from 73.8 D to 39.07 D, then to 49.13 D; K_2 value from 79.3 D to 45.03 D, then to 50.0 D; Pachy pupil from 426.00 um to 700.33 um, then to 488.33 um (pre- and post-operative are 1 week and 6 months, respectively). Indices of the Pentacam gradually decreased until the end of the 6 months follow-up.

Discussion

In this study, we report the successful management of an albino patient suffering from glaucoma, advanced keratoconus, and total cataract. Upon admission, the patient's visual acuity was hand motion on both eyes and he had a destitute social life. He underwent VisuMax femtosecond laser-assisted keratoplasty combined with "open-sky" cataract extraction and IOL implantation. With an improvement in visual acuity, he became self-sufficient and he could reintegrate into society.

The patient is an offspring of a father-daughter incest, which most likely accounts for his combined genetic disorder including OCA, cataract, and keratoconus. OCA Type 1A is associated with the highest rate of hyperopia and the poorest visual acuity among other subtypes of albinism. High hyperopia is frequent in albinism but unusual in keratoconus.[8] Corneal development is influenced by pigment-associated genes, and corneal thickness shows strong association with skin pigmentation.[3] Therefore, it is very likely that keratoconus in our patient is a rare consequence of OCA rather than an independent disease. The management of our patient required a unique approach due to the complex ophthalmic phenotype. Femtosecond laser-assisted keratoplasty has numerous benefits over traditional trephination including stable wound configuration, decreased "open-sky" time, and faster wound healing.[9] The hyperopia was corrected with a +29.0 D IOL resulting in a spherical equivalent of around zero with minimal astigmatism during the 6 months follow-up.

Angle developmental abnormalities have been considered as coincidence in albinism.[10] Our patient had an open and normally developed anterior chamber angle and a well-controlled IOP during the follow-up period.

Surgical management of low-vision patients with multiple ophthalmic comorbidities can be challenging, and the visual outcome is usually unpredictable in OCA patients.[7] Cataract operation and penetrating keratoplasty have been planned in two separate sessions in a previously reported case of OCA with keratoconus and cataract.[6] On contrary, femtosecond laser in a single setting combined operation assured low postoperative inflammation, fast wound healing and short recovery time in our case.

Visual improvement was unpredictable due to the multiplex ophthalmic phenotype including OCA, keratoconus, cataract, and glaucoma. Due to the use of femtosecond laser, corneal parameters improved very fast and remained stable thereafter. Complications did not occur during or after surgery. As far as we know, this is the first report on femtosecond laser-assisted keratoplasty using VisuMax femtosecond laser system with "open-sky" cataract extraction. UCDVA of 20/200 1 week after surgery can be considered as an excellent result. Interestingly,

Table 1: Pre- and post-operative mean values of the measured parameters over a 6 months follow-up period from the Pentacam

Days	K_1 (D)	K_2 (D)	K_{max} (D)	Astigmatism (D)	Pachy apex (µm)	Pachy pupil (µm)	Pachy minimum (µm)	Anterior chamber angle (°)	KI	CKI	IVA	IHA	ISV	IHD
0	73.80	79.30	103.70	5.55	364.00	426.00	193.00	39.80	1.88	1.26	1.88	129.90	257.00	0.51
7	39.07	45.03	51.00	5.97	711.67	700.33	644.00	12.63	1.29	1.12	1.86	129.43	189.33	0.18
23	45.70	48.60	53.90	2.80	655.00	658.00	634.00	17.30	1.25	1.11	1.00	23.80	148.00	0.14
45	48.00	48.20	53.50	0.20	525.00	518.00	501.50	28.40	1.17	1.08	0.73	53.75	128.50	0.06
112	48.40	51.20	54.93	2.80	485.67	479.33	472.00	29.67	1.04	1.05	0.47	16.90	72.33	0.04
167	49.13	50.00	53.93	0.83	496.33	488.33	481.67	27.97	1.02	1.05	0.48	12.33	67.00	0.04

K_1, K_2: Holladay equivalent keratometry values, K_{max}: *K* maximum, KI: Keratoconus-index, CKI: Center keratoconus index, IVA: Index of vertical asymmetry, IHA: Index of height asymmetry, ISV: Index of surface variance, IHD: Index of height decentration

our patient does not complain of glare or photophobia with the use of sunglasses.

The coexistence of albinism with keratoconus is unusual. Our case is the first where OCA has been associated with keratoconus, cataract, and glaucoma. Vision rehabilitation can be complicated in patients with multiple comorbidities. As a result of the combined surgery, the quality of life of the albino patient improved substantially. In summary, the VisuMax femtosecond laser-assisted triple procedure ensures fast patient rehabilitation even in challenging cases.

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Conflicts of interest

There are no conflicts of interest.

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