Case report

Femtosecond laser-assisted cataract surgery in anterior lenticonus due to Alport syndrome

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A R T I C L E   I N F O

Article history:
Received 11 August 2016
Received in revised form 13 January 2017
Available online 14 March 2017

Keywords:
Anterior lenticonus
Alport syndrome
Femtosecond laser
Cataract surgery

A B S T R A C T

Purpose: We describe a case of bilateral anterior lenticonus in a patient with Alport syndrome treated with femtosecond laser-assisted cataract surgery (FLACS).

Observations: FLACS was performed without complication, and a desirable postoperative visual acuity was achieved.

Conclusions and importance: Femtosecond laser-assisted cataract surgery is an effective approach for managing patients with anterior lenticonus secondary to Alport syndrome.

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1. Introduction

Alport syndrome is a rare, hereditary basement membrane disorder associated with renal failure, sensorineural deafness, and ocular defects. Typical ocular manifestations include dot-and-fleck retinopathy, anterior lenticonus, and posterior polymorphous corneal dystrophy.1–3 Anterior lenticonus occurs in approximately 50% of men with X-linked Alport syndrome and can cause significant visual limitations.4,5 Visual acuity and quality deficits resulting from anterior lenticonus can be managed surgically through removal of the natural lens, though modern techniques, particularly the creation of a manual capsulorhexis, are complicated by abnormal lens capsule structure in these patients.3,4,6–7

The role of femtosecond laser assisted cataract surgery (FLACS) in the treatment of anterior lenticonus in patients with Alport syndrome has not been frequently described. To our knowledge, there has been only one publication of two cases by Ecsedy et al.8 reporting on the use of FLACS in this patient population. We describe another case of bilateral anterior lenticonus secondary to Alport syndrome treated with FLACS.

2. Case report

A 53-year-old Caucasian male with a history of high myopia corrected with rigid gas permeable lenses and Alport syndrome was found to have progressive decreased distance visual acuity in both eyes. The patient had a history of early-onset hearing loss at age nine and end-stage renal disease at age 25 requiring renal transplant. His three brothers were all noted to have end-stage renal disease secondary to Alport syndrome.

On presentation, best-corrected visual acuity was 20/50 in both eyes. The refractive error was −9.00 + 0.75 × 125 in the right eye and −6.00 + 2.75 × 155 in the left eye. Corneal topography (ATLAS, Carl Zeiss Meditec AG, Jena, Germany) showed 0.63 diopters of regular astigmatism with a steep keratometry reading of 43.56 diopters at the 159-degree axis and a flattest keratometry reading of 42.93 diopters at the 69-degree axis. Topography of the left eye showed 1.45 diopters of regular astigmatism with a steep keratometry value of 42.93 diopters at the 163-degree axis and a flattest keratometry value of 43.08 diopters at the 73-degree axis.

Intraocular pressure was 15 mm Hg in both eyes. Slit lamp examination revealed anterior lenticonus and nuclear sclerosis of both eyes. The left eye had evidence of central posterior polymorphous corneal dystrophy, which historically had limited the patient’s best-corrected vision to 20/40. Fundus examination of both eyes was normal. Biometric measurements were obtained (IOL Master, Carl Zeiss Meditec AG, Jena, Germany) and repeated two months after discontinuing rigid gas permeable contact lenses to assess stability.

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http://dx.doi.org/10.1016/j.ajoc.2017.03.004
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The repeat measurements yielded an axial length was 23.60 mm in the right eye and 22.77 mm in the left eye. Keratometry readings were 43.72 diopters at 43° and 44.23 at 133° in the right eye. The left eye keratometry values were 43.66 at 64° and 44.82 at 154°. The decision was made to proceed with femtosecond assisted cataract surgery in both eyes to correct for astigmatism using the LenSx™ laser system (Alcon Laboratories) and the SoftFit™ interface (Alcon Laboratories). The plan was for monovision with the right eye corrected for distance and the left eye corrected for 1.5 diopters of myopia.

Anterior lenticonus was confirmed with anterior segment OCT through the LenSx interface (Fig. 1). The femtosecond laser then created a 5.2 mm capsulorhexis along with lenticular division in the right eye. The first limbal relaxing incision was placed at the 156-degree axis for a total length of 20°. A second limbal relaxing incision was placed at the 336-degree axis for a total length of 20°. Following laser pre-treatment, the surgery proceeded with creation of the primary incision using a manual keratome and standard removal of lens nucleus and cortex with phacoemulsification. A one-piece acrylic posterior chamber lens was placed in the capsular bag. Post-operative week one uncorrected vision was 20/20. A centered posterior chamber intraocular lens within the capsular bag. Post-operative week one uncorrected vision at distance was 20/60 at distance and Jaeger 2 at near. The posterior chamber intraocular lens was again well-centered in the capsular bag.

3. Discussion

Anterior lenticonus, even without the coexistence of a cataract, can lead to progressive visual acuity decline due to lenticular myopia and may require surgical intervention to restore full visual potential. Cataract surgery (or clear lens extraction) with phacoemulsification has proven to be an effective means of treating this patient population. However, more cases are necessary to fully determine the role of FLACS in managing these patients.

FLACS has been shown to provide a more regularly shaped capsulorhexis with better centration and lens/capsule overlap than manual capsulorhexis. Early experimental studies suggested that femtosecond laser-produced capsulotomies may also be stronger. Later findings from a large prospective cohort study of human patients seemed to contradict this notion, reporting a significantly increased rate of anterior capsular tears in the FLACS group attributed to the femtosecond laser’s postage-stamp pattern of perforation and aberrant pulses resulting from suspected eye movements. However, a more recent meta-analysis concluded that there were no significant differences in rates of anterior capsular tear between femtosecond laser and manual techniques.

The manual capsulorhexis in patients with anterior lenticonus can be technically challenging due to increased fragility or elasticity of the lens capsule. Further, an atypical cogwheel-like tearing pattern during manual capsulorhexis could predispose to capsular runoff or rupture. FLACS obviates this difficulty by providing an automated, well-centered capsulorhexis uniform in shape and size.

Ecsedy et al. described two patients (three eyes) treated with FLACS that had superb visual outcomes with good centration of the capsulotomy and intraocular lens position. They experienced one anterior capsular tear caused by injury to the capsular rim by the phaco tip during phacoemulsification. The case described herein represents the third such patient with anterior lenticonus secondary to Alport syndrome treated with FLACS. Similarly, we found FLACS to be a reliable technique for treating this population. Both eyes in this case had excellent visual outcomes with no intraoperative complications. It is worth noting that while the operations reported by Ecsedy et al. were performed under general anesthesia, our procedures were conducted using monitored anesthesia care without a retrobulbar block. This distinction is notable considering prior concerns regarding increased risk of anterior capsular tear with eye movements during FLACS. This case establishes that FLACS can safely treat this population without the need for general anesthesia.

Given the paucity of FLACS-treated anterior lenticonus cases in the scientific literature, conclusions regarding its superiority over traditional phacoemulsification techniques employing manual capsulotomy cannot yet be made. However, this case demonstrates that FLACS can achieve desirable outcomes in these surgically challenging patients. The uniform, centered femtosecond laser anterior capsulotomy appears particularly well-suited for this population considering the difficulties historically encountered with manual capsulorrhesis creation.
4. Conclusions

Femtosecond laser assisted cataract surgery appears to be a safe and effective approach to treating anterior lenticonus in Alport syndrome.

Patient consent

The patient consented to publication of the case orally.

Financial disclosures

None.

Acknowledgements and disclosures

Funding

No funding or grant support.

Conflict of interest

The following authors have no financial disclosures: ACB, ASR.

Authorship

All authors attest that they meet the current ICMJE criteria for Authorship.

Acknowledgements

None.

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