DOI: 10.14744/eer.2022.18209
Eur Eye Res 2023;3(1):32–35

EUROPEAN EYE RESEARCH

CASE REPORT

Long-term management of gelatinous droplet dystrophy with phototherapeutic keratectomy and toric soft contact lenses

Zeynep Ozbek,[1] Betul Akbulut Yagci,[1] Bora Yuksel,[2] Ismet Durak[1]

[1]Department of Ophthalmology, Dokuz Eylul University Faculty of Medicine, Izmir, Türkiye
[2]Department of Ophthalmology, Bozyaka Training and Research Hospital, Izmir, Türkiye

Abstract

The aim of the study was to report the results of phototherapeutic keratectomy (PTK) and toric contact lens fitting for a young man with recurrent gelatinous droplet dystrophy (GDD) after penetrating keratoplasty (PK). A 21-year-old man was referred for pain, photophobia, and decreased vision. The patient who experienced decreasing vision for 15 years had undergone PK 2 years ago due to GDD. He was having frequent recurrent epithelial erosions lately. Visual acuity (VA) was counting fingers at 3 m in the right eye and 0.8 in the left eye. Biomicroscopic examiantion revealed nodular dystrophic lesions on the nasal side of the graft in the right eye. Keratometric values were K1: 54.5, K2: 52.5 in the right eye and K1: 41.2, K2: 39.7 in the left eye. PTK was performed twice in the right eye and once in the left eye in 3 years. Final VA was 0.5 and 0.8 in the right and left eyes, respectively (with glasses and toric contact lenses) during 10 years of follow-up. A superficial corneal scar was noted on the right graft and the left cornea. No recurrence of dystrophy was observed. PTK decreases photophobia and provides visual improvement in patients with GDD and may help defer PK in case of recurrent GDD.

Keywords: Gelatinous droplet dystrophy; phototherapeutic keratectomy; toric contact lens.

Gelatinous droplet dystrophy (GDD) is an initially insidious autosomal recessive disease first described by Nakaizumi in 1914; characterized by corneal subepithelial and stromal accumulation of gelatinous amyloid material.[1–3] It is a rare amyloid expression disorder with an incidence of 1:300,000 in the Japanese population. The majority of cases manifest as severe photophobia, foreign-body sensation, lacrimation, and blepharospasm between the first and second decades of life.[2] The characteristics of GDD include gelatinous mass deposition in the anterior corneal stroma, the superficial elevated nodules (called mulberry-type nodules) and the progressive corneal opacity causing blurred vision. Initial treatment is predominantly conservative by means of artificial tears and soft contact lenses.

Over time, the corneal surface gradually becomes irregular and deep corneal involvement with vascularization causes progressive visual loss. Surgical treatment options include phototherapeutic keratectomy (PTK), superficial kerate-

Cite this article as: Ozbek Z, Akbulut Yagci B, Yuksel B, Durak I. Long-term management of gelatinous droplet dystrophy with phototherapeutic keratectomy and toric soft contact lenses. Eur Eye Res 2022;3(1):32-35.

Correspondence: Zeynep Ozbek, M.D. Department of Ophthalmology, Dokuz Eylul University Faculty of Medicine, Izmir, Türkiye
Phone: +90 232 390 37 88 **E-mail:** zeynep_ozbek@hotmail.com
Submitted Date: 14.07.2022 **Revised Date:** 06.09.2022 **Accepted Date:** 13.09.2022

ctomy, lamellar keratoplasty, or penetrating keratoplasty (PK). High recurrence rate of GDD may necessitate multiple surgical treatments. In cases of recurrence, repeat grafts may be performed and ultimately limbal stem cell transplantation or keratoprosthesis may be required as last resort.[4–6]

PTK, approved by the Food and Drug Administration in 1995, is widely used in the treatment of anterior corneal dystrophies.[7] The aim of PTK is to eliminate opacities affecting visual acuity (VA) and to provide a smooth and stable corneal surface. Today, PTK is widely used to treat stromal dystrophies in the early stages and may be an important alternative to delay the need for PK.[8,9]

We hereby describe a patient who was experiencing decreased VA and photophobia due to recurrent GDD on graft. A PTK was performed since recurrence was superficial and the most of the graft was clear.

Case Report

A 21-year-old man was referred for pain, photofobia, and decreased vision in the right eye in August 2008. The patient had decreased vision since he was 4 years old and had undergone right eye PK 2 years ago due to GDD. We noticed recurrent GDD on graft in the right eye with an 3×3 mm epithelial defect and a central GDD lesion on the left cornea. VA with Snellen chart was counting fingers at 3 m with extreme photophobia in the right eye and 0.8 in the left eye. At the slit-lamp nodular dystrophic lesions were seen on the nasal side of the graft in the right eye. Intraocular pressures were 16 and 17 mmHg in the right and left eyes, respectively, with Goldmann Applanation Tonometry. Fundus examination was normal.

Orbscan readings were silip mean power: 54.5, in the right eye and Sim K's astigmatism was 20.9×114°, central corneal thickness (CCT) was 613 microns. Parameters for the left eye were mean power: 41.2, and Sim K's astigmatism: 3.9×79°, CCT: 546 microns. After obtaining informed consent, the patient was planned PTK for the right eye in September 2008. VA was 0.6 (−3.00−4.00 × 10°) at the 1st week postoperatively and remained stable during 1 year of follow-up. The patient presented with bilateral decreased vision in 2010. VA was 0.1 and 0.3 in the right and left eyes, respectively. Biomicroscopy revealed new similar nodular lesions in the right graft and an increase in the GDD lesion on the left cornea (Fig. 1). He was planned for bilateral PTK in June 2010.

Post-operative Orbscan topographic parameters were: mean power: 47.6, Sim K's Astigmatism: 7.5×99°, CCT: 509 microns in the right eye; mean power: 39.4, Sim K's Astigmatism: 4.4×60°, CCT: 574 microns in the left eye.

VA increased up to 0.5 with the refraction: (−3.0−5.50 × 180) in the right eye and 0.7 with the refraction: (−1.0 −4.00 × 160) in the left eye. Rigid gas permeable contact lens (CL) fitting was suggested and applied. However, the patient did not agree to use rigid gas permeable CL and stated that he would rather wear soft contacts. After soft toric CL fitting (−3.00 −2.25 × 180 OD/−1.00 −2.25 × 160 OS), his VA was still 0.5 OD and 0.7 OS. Since high cylinder soft toric CL (XR torics) was not available at the time and VA improved 2 more lines with an overrefraction of (0.0 −3.00 × 180) over the soft toric CL for the right eye, (0–1.25 × 160°) over the soft toric CL for the left eye, he stated that he could wear glasses over his toric soft lenses. Therefore, these cylinder values were given as spectacles. Final VA increased to 0.8 in the right eye and 0.9 in the left eye with both toric soft contact lenses stated above and glasses (0.0–3.00 × 180/0–1.25 × 160°) over soft lenses. A central scar on the graft and a paracentral nasal scar on the left cornea was observed at the slit-lamp (Fig. 2).

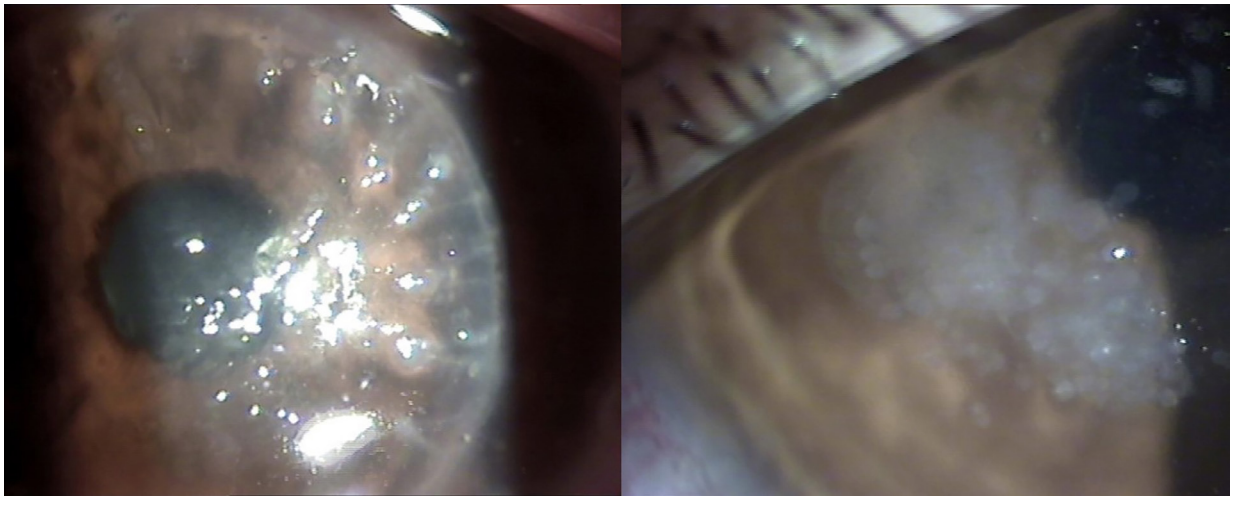

Fig. 1. Gelatinous droplet dystrophy in the right eye and left eye at presentation.

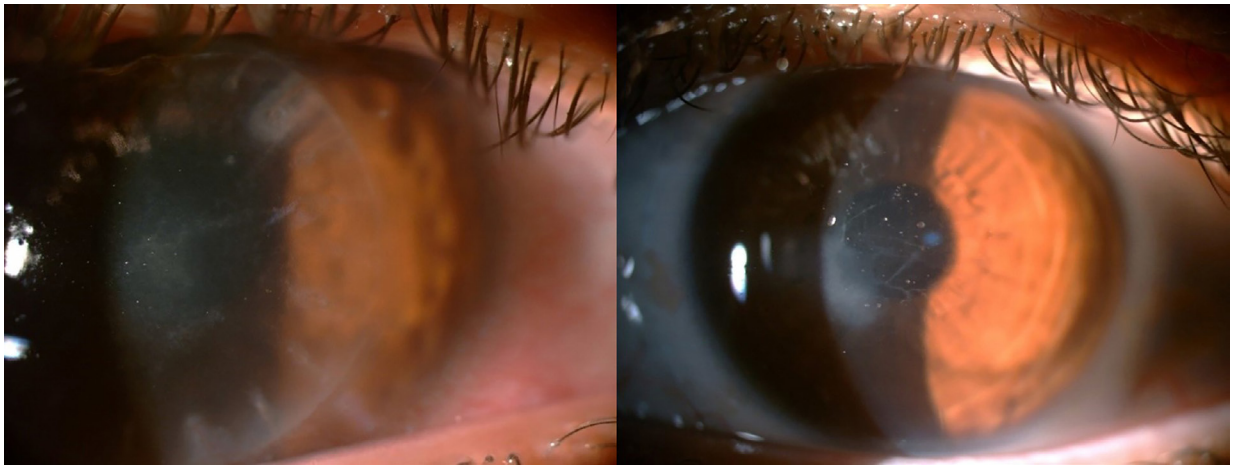

Fig. 2. Central scar in the right graft and paracentral nasal scar in the left cornea 2 years after bilateral PTK.

Fig. 3. Central scar in the right graft and paracentral nasal scar in the left cornea 10 years after bilateral PTK.

At the last examination of the patient, 10 years after the first examination, VA was 0.5 in the right eye and 0.8 in the left eye (with his toric contact lenses and glasses). There was no recurrence but a slight corneal scar in both eyes (Fig. 3) Keratometric values were: K1: 42.4, K2: 47.7, Km: 44.9, 9 and, K1: 40.2, K2: 45.7, in the right and left eyes, respectively, with Pentacam®. The patient had no complaints.

Discussion

GDD is an autosomal recessive disease in which the majority of cases manifest with severe photophobia, foreign-body sensation, lacrimation, and blepharospasm between the first and second decades of life. Initially, treatment is predominantly conservative by means of artificial tears, soft contact lenses, and, where appropriate, topical immunomodulatory eye drops. The standard treatment is keratoplasty; however, recurrences are reported and repeated keratoplasty has possible secondary complications.[3,9]

PTK is a safe and effective procedure for the treatment of corneal opacities located in the anterior one-third of the cornea.[7–13] The main goals of performing PTK are to remove or decrease the opacities affecting the VA, to smooth the corneal surface, to improve the adherence of the corneal epithelium, or a combination thereof to help with painful symptoms.[8] PTK has been widely used for corneal dystrophies, mostly for granular dystrophy.[10–12] Although corneal dystrophies can recur after PTK as well, PTK can be repeated more than once if necessary depending on the amount of stromal ablation of each treatment and performed in grafted or non-grafted corneas.[7,8]

Dinh et al.[7] and Chen et al.[9] performed PTK on eyes with different corneal dystrophies with or without corneal grafts.[8] Dinh et al. reported a mean time to significant recurrence of 3.4 years and Reddy et al.[8] at 3.6 years. Chen et al. reported a mean time to significant recurrence of 2 years in 15 eyes. Variations in the PTK technique may account for differences in reported recurrence rates. In our case, the right eye had a recurrence 20 months after PTK.

Corneal subepithelial and stromal accumulation of gelatinous amyloid material in GDD was suspected to originate

from corneal basal cells; however, it is now suspected that it comes from the tear film, migrates underneath the epithelium due to impaired epithelial barrier function, and aggregates to form a drop-like pattern.[14]

Maeno et al.[13] investigated if therapeutic soft CL usage has positive effects in GDD management on a retrospective observational case series. They saw that continuous soft CL usage lowered rate of progression on nodular lesions and reduces the need for surgical interventions. They found a greatly prolonged surgery-free interval as well as a lower progression of corneal lesions in the eyes treated with soft CL and postulated that soft CL usage might influence the invasion of excessive proteins. They discussed that continuous wear soft CL usage might decrease the turnover and fall-off of surface epithelial cells; decrease the turnover of tear fluid around the corneal surface; protect from mechanical stimulation stress, such as blinking or capture lactoferrin, factors that are all considered to decrease the permeation of the tear fluid into the corneal tissue; and contribute to reduced subepithelial precipitation in GDD.

Also the same year, Hieda et al.[15] reported clinical outcome and time to recurrence of PTK on 714 eyes of 477 consecutive patients over a follow-up of mean 44 months. There was only one GDD eye with significant recurrence in 16 eyes with GDD. The restricted mean survival time for observing a significant recurrence for GDD was 10 years with a soft CL.[15]

Conclusion

We hereby report our case with recurrent GDD on a graft managed by PTK. Although it was not our main intention, our patient chose to wear monthly toric soft CL after the second PTK for both eyes. Interestingly, no recurrence of dystrophy was noted on the graft or the left cornea with soft toric CL use after PTK over 10 years now whereas first recurrence after PTK was within the 2nd year without CL.

Informed Consent: Written informed consent was obtained from the patient for the publication of the case report and the accompanying images.

Peer-review: Externally peer-reviewed.

Authorship Contributions: Concept: Z.O.; Design: Z.O.; Supervision: I.D.; Resource: B.Y.; Materials: B.Y.; Data Collection and/or Processing: Z.O., B.Y.; Analysis and/or Interpretation: Z.O.; Literature Search: B.A.Y.; Writing: B.A.Y.; Critical Reviews: I.D.

Conflict of Interest: None declared.

Financial Disclosure: The authors declared that this study received no financial support.

References

1. Nakaizumi G. A rare case of corneal dystrophy. Acta Soc Ophthalmol Jpn 1914;18:949–50.
2. Kaza H, Barik MR, Reddy MM, Mittal R, Das S. Gelatinous drop-like corneal dystrophy: A review. Br J Ophthalmol 2017;101:10–5. [CrossRef]
3. Quantock AJ, Nishida K, Kinoshita S. Histopathology of recurrent gelatinous drop-like corneal dystrophy. Cornea 1998;17:215–21. [CrossRef]
4. Alex AF, Eter N, Uhlig CE. Combined excimer laser photoablation and amniotic membrane overlay for relief of symptomatic discomfort in gelatinous drop-like corneal dystrophy. Cornea 2015;34:1316–7. [CrossRef]
5. Movahedan H, Anvari-Ardekani HR, Nowroozzadeh MH. Limbal stem cell transplantation for gelatinous drop-like corneal dystrophy. J Ophthalmic Vis Res 2013;8:107–12.
6. Cortina MS, Porter IW, Sugar J, De la Cruz J. Boston Type I keratoprosthesis for visual rehabilitation in a patient with gelatinous drop-like corneal dystrophy. Cornea 2012;31:844–5.
7. Dinh R, Rapuano CJ, Cohen EJ, Laibson PR. Recurrence of corneal dystrophy after excimer laser phototherapeutic keratectomy. Ophthalmology 1999;106:1490–7. [CrossRef]
8. Reddy JC, Rapuano CJ, Nagra PK, Hammersmith KM. Excimer laser phototherapeutic keratectomy in eyes with corneal stromal dystrophies with and without a corneal graft. Am J Ophthalmol 2013;155:1111–8e. [CrossRef]
9. Chen M, Xie L. Features of recurrence after excimer laser phototherapeutic keratectomy for anterior corneal pathologies in North China. Ophthalmology 2013;120:1179–85. [CrossRef]
10. Yuksel E, Cubuk MO, Eroglu HY, Bilgihan K. Excimer laser phototherapeutic keratectomy in conjunction with mitomycin C in corneal macular and granular dystrophies. Arq Bras Oftalmol 2016;79:69–72. [CrossRef]
11. Rathi VM, Taneja M, Murthy SI, Bagga B, Vaddavalli PK, Sangwan VS. Phototherapeutic keratectomy for recurrent granular dystrophy in postpenetrating keratoplasty eyes. Indian J Ophthalmol 2016;64:140–4.
12. Nakamura T, Kataoka T, Kojima T, Yoshida Y, Sugiyama Y. Refractive outcomes after phototherapeutic refractive keratectomy for granular corneal dystrophy. Cornea 2018;37:548–53.
13. Maeno S, Soma T, Tsujikawa M, Shigeta R, Kawasaki R, Oie Y, et al. Efficacy of therapeutic soft contact lens in the management of gelatinous drop-like corneal dystrophy. Br J Ophthalmol 2020;104:241–6. [CrossRef]
14. Kawasaki S, Kinoshita S. Clinical and basic aspects of gelatinous drop-like corneal dystrophy. Dev Ophthalmol 2011;48:97–115. [CrossRef]
15. Hieda O, Kawasaki S, Yamamura K, Nakatsukasa M, Kinoshita S, Sotozono C. Clinical outcomes and time to recurrence of phototherapeutic keratectomy in Japan. Medicine (Baltimore) 2019;98:e16216. [CrossRef]