Granular Corneal Dystrophy Manifesting after Radial Keratotomy

Sepehr Feizi, MD; Mohammad Pakravan, MD; Alireza Baradaran-Rafii, MD; Shahin Yazdani, MD

Shaheed Beheshti Medical University, Tehran, Iran

Purpose: To report manifestation of granular corneal dystrophy after radial keratotomy (RK).

Case Report: A 32-year-old man presented with white radial lines in both corneas. He had undergone uncomplicated RK in both eyes 8 years ago. Preoperative refraction had been OD: -3.5 -0.75@180 and OS: -3.0 -0.5@175. Uncorrected visual acuity was OD: 8/10 and OS: 7/10; best corrected visual acuity was 9/10 in both eyes with OD: -0.5 -0.5@60 and OS: -0.75 -0.5@80. Slit lamp examination revealed discrete well-demarcated whitish lesions with clear intervening stroma in the central anterior cornea consistent with granular dystrophy. Similar opacities were present within the RK incisions.

Conclusion: Granular dystrophy deposits may appear within RK incisions besides other previously reported locations.

Iran J Ophthalmic Res 2002; 2 (1): 81-84.

Correspondence to: Sepehr Feizi, MD, Anterior segment fellow; Ophthalmic Research Center, Labbafinejad Medical Center, Boostan 9 St., Pasdaran Ave., Tehran, 16666, Iran; Tel: +98 21 22585952, Fax: +98 21 22590607, e-mail: sepehrfeizi@yahoo.com

INTRODUCTION

Granular corneal dystrophy is the most common hereditary stromal corneal dystrophy. It is an autosomal dominant condition,[1] characterized by deposition of opacities in the corneal stroma. Visual impairment is rarely severe until the fifth decade.[2] Visual impairment results from progressive loss of corneal transparency due to deposition of aberrantly processed kerato-epithelin.[3]

Radial keratotomy (RK) is a keratorefractive procedure used to correct myopia in which deep non-penetrating, radial corneal incisions flatten the central cornea.[4] Although several reports have indicated the occurrence of various corneal dystrophies after RK, to the best of our knowledge this report is the first describing of granular corneal dystrophy after RK.

CASE REPORT

A 32-year-old man, who had undergone uncomplicated RK in both eyes 8 years ago, was seen complaining of white lines in his eyes. Past medical history was unremarkable and he did not mention any systemic disorder. Upon presentation, uncorrected visual acuity was OD: 8/10 and OS: 7/10. Refraction was OD: -0.5 -0.5@60 and OS: -0.75 -0.5@80 and best corrected visual acuity was 9/10 in both eyes. Slitlamp examination revealed several discrete well-demarcated white opacities in the anterior corneal stroma in both eyes, typical of granular dystrophy (Fig. 1). The intervening cornea was clear except for the RK incisions which showed similar white deposits within the incisions. Corneal sensation and other ocular examinations were normal. Central corneal thickness

measured by ultrasonic pachymetry was 533 μm in the right eye and 542 μm in the left eye. Preoperative data revealed baseline refraction of OD: -3.5 -0.75@180 and OS: -3.0 -0.5@175 and normal corneal clarity in both corneas. Other ocular examinations had been unremarkable. We called on first- and second-degree relatives for ophthalmic examination and diagnosed granular dystrophy in one of his cousins.

DISCUSSION

Granular corneal dystrophy appears in the first and second decades of life.[1] It is a bilateral condition characterized by deposition of small, discrete, sharply demarcated, grayish-white opacities in the central anterior stroma. The intervening stroma remains clear and vision is usually not affected early in the course. With disease progression, lesions increase in size and number and may coalesce with extension into deeper and more peripheral stroma. However, the peripheral cornea usually remains free of deposits. Visual impairment is rarely severe until the fifth decade[2] which is due to progressive loss of corneal transparency secondary to deposition of aberrantly processed kerato-epithelin.[3] Light microscopy demonstrates eosinophilic, rod, or trapezoidal-shaped hyaline deposits in the stroma and beneath the epithelium. These deposits stain bright red with Masson's trichrome and weakly with periodic acid-Schiff.[5] The exact nature and source of the deposits remain unclear. It is also unknown whether these deposits are produced by epithelium, stromal keratocytes, or both; however, characteristic rod-shaped structures have been seen within both cell types.[6]

The histopathologic features of post-RK corneas have been described in a number of reports. Different reparative responses in RK incisions have been observed, ranging from hypertrophic scar formation to persistent non-healing incisions.[7] A striking feature of the post-RK cornea is the presence of an epithelial plug which remains several years after surgery.[8] Additional abnormalities include aberrant epithelial basal lamina synthesis including basal lamina duplication and absence of basal lamina over areas of extensive fibrosis,[9] Bowman layer discontinuity, extracellular matrix formation around the incisions, inability to synthesize normal type IV collagen around epithelial plugs, increased number of stromal cells, and abnormalities in underlying endothelial cells such as endothelial permeability and cell loss.[10] Subepithelial fibrosis after radial keratotomy was reported by Gieser and Sugar.[11] A review of the literature indicates that in most cases, post-RK corneal scarring occurs along the radial incisions.[12] Other variations in wound healing are formation of epithelial retention cysts, incomplete wound healing, persistent wound gape and hypertrophic scar formation.[7]

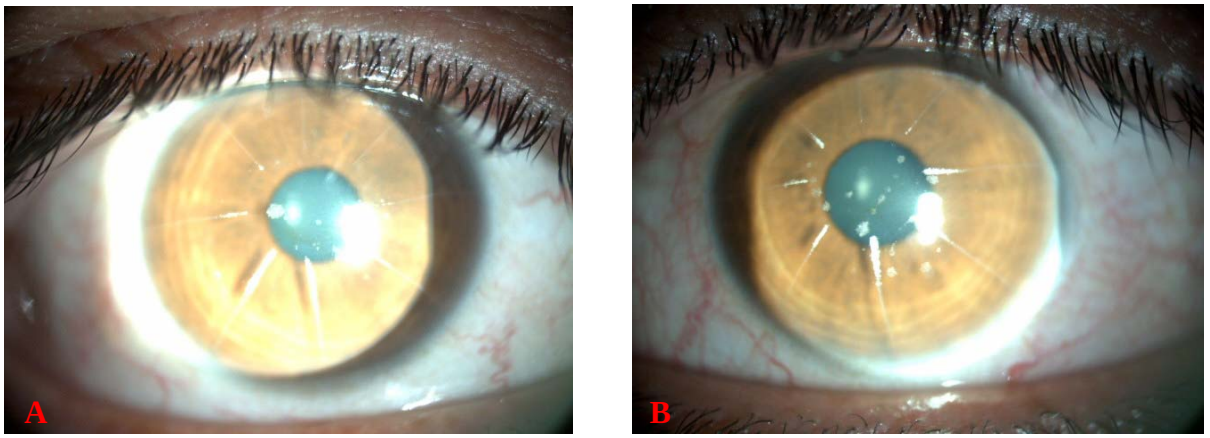

Figure 1 Slitlamp photograph of the right (A) and left (B) eyes reveals deposits compatible with granular dystrophy in the central cornea and within radial keratotomy incisions.

Deposition of abnormal material in corneas with epithelial (map-dot-fingerprint) dystrophies after RK[13] or stromal dystrophies after other keratorefractive procedures has been reported.[14-16] Production of such abnormal material can be due to activation of keratocytes following LASIK or epithelial cells after PRK. The first hypothesis which may explain the deposition of dystrophic material in such eyes is that severing corneal collagen fibers during RK induces attachment of large amounts of abnormal material (hyaline) to the collagen in the incisions. Roh et al[16] reported three cases with Avellino corneal dystrophy who underwent LASIK. Abnormal deposition of extracellular granular material was noted in the interface where collagen bundles were cut by the microkeratome or damaged by laser. This material was not observed in deeper collagen fibers in the same cornea which was not damaged during the operation.

Another hypothesis is stromal migration of epithelial cells into the incisions. Frising et al[17] suggested the epithelial origin of granular dystrophy deposits by observing deep stromal deposits in suture tracts and at the host-donor interface. Some suggest that since epithelial findings are more prominent in recurrent granular dystrophy after penetrating keratoplasty, the condition may have an epithelial origin.[18-20] Electron microscopic studies have found these deposits to be surrounded by cytoplasmic extensions of epithelial cells or even within the corneal epithelium.[18,19] According to this hypothesis, abnormal kerato-epithelin material may accumulate within RK incisions because of activation and proliferation of epithelial cells to fill the incisions.[10] We presume that proliferating and migrating corneal epithelial cells from the margin of the RK incisions might be altered both morphologically and functionally with a tendency to over-express abnormal kerato-epithelin.

This case report may provide some understanding about the predilection of deposits of recurrent granular dystrophy for certain locations, such as within suture tracts and at the host-donor interface after penetrating keratoplasty, beneath the corneal flap in LASIK, at the periphery of the ablation zone in PRK and within RK incisions as described by this report.

REFERENCES

1. Waardenburg PJ, Jonkers GH. A specific type of dominant progressive dystrophy of the cornea, developing after birth. *Acta Ophthalmol(Copenh)* 1961;39:919-923.
2. Waring GO 3rd, Rodrigues MM, Laibson PR. Corneal dystrophies. I. Dystrophies of the epithelium, Bowman's layer and stroma. *Surv Ophthalmol* 1978;23:71-122.
3. Klintworth GK, Valnickova Z, Enghild JJ. Accumulation of big-h3 gene product in corneas with granular dystrophy. *Am J Pathol* 1998;152:743-748.
4. Waring GO, Lynn M, McDonnell P. Results of the prospective evaluation of radial keratotomy (PERK) study 10 years after surgery. *Arch Ophthalmol* 1994;112:1298-1308.
5. Jones ST, Zimmerman LE. Histopathologic differentiation of granular, macular, and lattice dystrophies of the cornea. *Am J Ophthalmol* 1961;51:394-410.
6. Wittebol-Post D, van der Want JJ, van Bijsterveld OP. Granular dystrophy of the cornea (Groenouw's type I): is the keratocyte the primary source after all? Ophthalmologica 1987;195:169-177.
7. Jester JV, Villasenor RA, Schanzlin DJ, Cavanagh HD. Variations in corneal wound healing after radial keratotomy: possible insights into mechanisms of clinical complications and refractive effects. Cornea 1992;11:191-199.
8. Khoroshilova-Maslova IP, Andreeva LD, Ilatovskaia LV, Kuznetsova IA. Clinical and histopathologic examination of enucleated eyes with contusion ruptures of cornea after radial keratotomy. Vestn Oftalmol 1998;4:3-8.
9. Jester JV, Villasenor RA, Schanzlin DJ, Cavanagh HD. Variations in corneal wound healing after radial keratotomy: possible insights into mechanisms of clinical complications and refractive effects. Cornea 1992;11:191-199.
10. Bergmanson J, Farmer E, Goosey J. Epithelial plugs in radial keratotomy: the origin of incisional keratitis? Cornea 2001;20:866-872.
11. Gieser J, Sugar A. Radial keratotomy and corneal scarring [letter]. Arch Ophthalmol 1992;110:1527-1528.
12. Majmudar PA, Forstot SL, Dennis RF, Nirankari VS, Damiano RE, Brenart R, et al. Topical mitomycin-C for subepithelial fibrosis after refractive corneal surgery. Ophthalmology 2000;107:89-94.

13. Nelson JD, Williams P, Lindstrom RL, Doughman DJ. Map-fingerprint-dot changes in the corneal epithelial basement membrane following radial keratotomy. Ophthalmology 1985;92:199-205.
14. Wan XH, Lee HC, Stultingv RD. Exacerbation of Avellino corneal dystrophy after laser in situ keratomileusis. Cornea 2002;21:223-226.
15. Jun RM, Tchah H, Kim TI, Stulting RD, Jung SE, Seo KY, et al. Avellino corneal dystrophy after LASIK. Ophthalmology 2004;111:463-468.
16. Roh MI, Grossniklaus HE, Chung SH, Kang SJ, Kim WC, Kim EK. Avellino corneal dystrophy exacerbated after LASIK: scanning electron microscopic findings. Cornea 2006;25:306-311.
17. Frising M, Wildhardt G, Frisch L, Pitz S. Recurrent granular dystrophy of the cornea: An unusual case. Cornea 2006;25:614-617.
18. Johnson BL., Brown SI, Zaidman GW. A light and electron microscopic study of recurrent granular dystrophy of the cornea. Am J Ophthalmol 1981;92:49-58.
19. Witschel H, Sundmacher R. Bilateral recurrence of granular corneal dystrophy in the grafts. A clinico-pathologic study. Graefes Arch Clin Exp Ophthalmol 1979;209:179-188.
20. Lyons CJ, McCartney AC, Kirkness CM, Ficker LA, Steele AD, Rice NS. Granular corneal dystrophy. Visual results and pattern of recurrence after lamellar or penetrating keratoplasty. Ophthalmology 1994;101(11):1812-1817.