Granular corneal dystrophy: treatment with soft contact lenses

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Abstract Perforating keratoplasty was performed in both eyes of two siblings (sister and brother) with typical granular corneal dystrophy. Three of the operated eyes showed a recurrence of the underlying disease. In one eye, a recurrence could not be detected 14 years after the operation. This eye had been fitted with a contact lens 16 months after keratoplasty because of aphakia. Thus a contact lens may have a therapeutic effect in this primary epithelial disease.

Introduction

In granular corneal dystrophy which has been present for a long period and which has also affected the stroma, perforating keratoplasty is performed in order to improve vision. If the opacity is only situated superficially, as in the initial stage of the disease and in recurrences, abrasion [14, 15], superficial keratectomy without covering [12, 16], phototherapeutic keratectomy [1, 5, 6, 9] and lamellar keratoplasty [13] are recommended.

Recurrences can be observed irrespective of treatment method [2, 3, 7, 8, 13, 18, 21–24]. According to Weidle [23] recurrences are always found if the patients are observed for long enough. Only one out of 12 patients he followed up had not shown a recurrent opacity after 6 years.

There is no method of preventing recurrences. For this reason, we report the cases of two patients (siblings) who underwent perforating keratoplasty on both sides. One of the four operated eyes did not show a recurrence after 14 years' observation.

Case reports

Patient 1

This female patient was 41 years old on initial presentation at the Department of Ophthalmology, University of Cologne. Both eyes displayed the following features in 1972: Typical clinical picture of granular corneal dystrophy with opacities extending down into the deep stroma (Fig. 1). In 1972, perforating keratoplasty on the right eye was performed. The same procedure was carried out on the left eye in 1973. Histological examination confirmed the clinical assumption of granular corneal dystrophy. No follow-up was carried out at the University of Cologne in the following years. Incipient visual defects were found 10 years after the operations. The right eye deteriorated severely. In 1994 there was pronounced recurrence on both sides (Fig. 2): visual acuity of right eye, +1.5 sph. comb. –2.0 cyl A 10°=0.2; left eye, +2.5 sph. comb. –3.0 cyl A 170°=0.5 in part.

Rekeratoplasty of the right eye was carried out in 1994 with extracapsular lens removal and lens implantation. No complications ensued. Histological findings are shown in Fig. 3. The last check-up was 13 months after the operation: Visual acuity of right eye, +0.5 sph. comb. –1.0 cyl A 65°=0.6, clear transplant; reduction of vision due to opacity of posterior lens capsule. Visual acuity of left eye, +2.25 sph. comb. –1.25 cyl A 140°=0.5.

Patient 2

In this male patient, the brother of patient 1, the initial findings were the typical clinical picture of bilateral granular corneal dystrophy. The left eye was treated by perforating keratoplasty in 1982. The histological findings confirmed the clinical diagnosis. Sixteen months later, intracapsular lens extraction was carried out and reductive correction was achieved with a soft contact lens. This lens (8.6 mm, +13.0 dptr, 14.0 mm, 70% H2O) was worn for 12–14 h a day and cleaned with a system based on H2O2. The lens was renewed every 8–9 months. Retinal detachment occurred 9 years after cataract surgery. The retina was successfully surgically re-attached. The corneal transplant remained clear up to the last follow-up 14

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years after keratoplasty. The visual acuity with soft contact lens was 0.5 in part. There was no indication of recurrence of the underlying disease (Fig. 4).

The right eye was subjected to perforating keratoplasty in 1987. The histological findings corresponded to those of a granular corneal dystrophy. Six months later, extracapsular cataract extraction was performed with lens implantation. Four years after the keratoplasty, there were incipient whitish deposits in the suture channels. The findings could not be unequivocally evaluated as a recurrence.

When the patient presented again 7 years after keratoplasty, recurrence was clearly discernible (Fig. 5).

Discussion

After surgical treatment of a granular corneal dystrophy, recurrences occur which are initially superficial, but which can later penetrate into the stroma. According to Weidle [23], these opacities almost always occur eventually. Only one of his six patients did not show any recurrence after 6 years. Other reports also indicate that superficial recurrences can occur relatively quickly (2 years [10, 18, 19]; 4 years [13]; up to 5.5 years [24]. The opacities can remain superficial for years. The process can still be purely superficial 3 years after the first observation of the recurrence [20]. Only after longer periods do opacities also occur in the stroma.

In patient 1 (female), rekeratoplasty was performed 21 years after the first operation because of recurrence of the granular corneal dystrophy. According to the patient, slight visual deficiencies occurred about 10 years after the first keratoplasty.

In patient 2 (male), perforating keratoplasty was performed on the left eye in 1982. Sixteen months later, lens extraction was carried out. Refractive correction was accomplished by a soft contact lens. Retinal detachment occurred 9 years after the intracapsular aphakia. This was successfully treated by surgery. No signs of recurrence were found in this eye at the last follow-up examination 14 years after keratoplasty.

Perforating keratoplasty was performed on the right eye in 1987 because of the corneal dystrophy. Nine months later extracapsular lens extraction with lens implantation was carried out. Four years after the keratoplasty, the first signs of a superficial recurrence of the underlying disease could be detected.