Bilateral Keratectasia 34 Years after Corneal Transplant

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Key Words

Bilateral keratectasia · Penetrating keratoplasty · Bilateral keratoconus

Abstract

We report the clinical findings of a patient with severe bilateral keratectasia 34 years after a penetrating keratoplasty (PK) in both eyes. An otherwise healthy 67-year-old man complained of deterioration of the eyesight in both eyes over the last 6 months. The patient was diagnosed with bilateral keratoconus at the age of 32 years, and he underwent a bilateral PK. At presentation, visual acuity was 20/200 in the right eye and light perception in the left eye. A Pentacam pachymetric map revealed a central pachymetry of 720 μm in the right eye and of 710 μm in the left eye, as well as an average paracentral pachymetry of 436 and 270 μm in the 9-mm zone in the right and the left eye, respectively. Corneal topography revealed bilateral irregular and asymmetric bowing with generalized steepening and high corneal power. We describe a case of bilateral keratectasia 34 years after PK in a patient who was originally diagnosed with bilateral keratoconus.

Introduction

Keratoglobus is a non-progressive or minimally progressive, non-inflammatory corneal degeneration defined by a globular protrusion of the cornea with or without enlargement of the corneal diameter and a generalized thinning mainly in the periphery, without the presence of neovascularization [1]. The cornea is usually transparent, but acute corneal edema due to rupture of Descemet's membrane and perforation, even from minimal trauma, is the most frequent complication [2]. This rare condition is usually bilateral and has been associated with several eye conditions, such as blue sclera syndrome [3], Leber's congenital amaurosis [4], pellucid marginal degeneration [5], inflammatory

orbital pseudotumor, chronic marginal blepharitis, and chronic eye rubbing, and it can also develop following penetrating keratoplasty (PK) [1]. In addition, it has been related to joint hypermobility and consanguineous parents [6]. Acquired keratoglobus has also been described in association with vernal keratoconjunctivitis [1], hyperthyroidism [7], advanced keratopathy (keratoconus), trauma and exophthalmos [8]. The pathogenesis and the molecular events responsible for the disease expression remain unclear [3], and there are limited reports of the genetic background of keratoglobus [4].

Case Report

An otherwise healthy 67-year-old man presented with progressive deterioration of vision in both eyes over the last 6 months. Best-corrected visual acuity (BCVA), using contact lens combined with spectacle correction, was 20/200 in the right eye (–18.50 dpt sph, –1.50 dpt cyl) and light perception (–4.00 dpt sph) in the left eye.

The patient was diagnosed with bilateral keratoconus at the age of 32 years, with no associated connective tissue disorders or other systemic diseases. At that time, BCVA was 20/200 in the right eye and 20/70 in the left eye, and refraction was –14.00 dpt in both eyes. One year after the keratoconus diagnosis, the patient underwent a PK in the left eye for a progressive deterioration of the eyesight and thinning of the cornea. Six years later, another PK was performed in the right eye.

A 6.5-mm diameter keratoplasty was performed in the right eye and a 7.00-mm diameter keratoplasty in the left eye to include the abnormal cornea. BCVA recovered rapidly to 20/40 in the left eye and 20/50 in the right eye after the surgeries, and it remained stable until six months before the patient attended our clinic.

At presentation, slit-lamp examination showed clear corneal buttons (fig. 1), globular protrusion of the cornea with ectatic profile and midperipheral thinning (fig. 2). Bilateral full-thickness corneal scars, resulting from the previous PK, were also noted. Vertical and horizontal corneal diameters were 11.0 and 11.6 mm, respectively, in the right eye, and 11.0 and 11.8 mm, respectively, in the left eye. Intraocular pressure (IOP) was 12 mm Hg in the right eye and 14 mm Hg in the left eye. Anterior chamber depth was 4.63 mm in the right eye and 5.71 mm in the left eye, and axial length was 33.04 and 35.40 mm, respectively (CineScan 'S'; Quantel Medical, Bozeman, Mont., USA). No other findings apart from moderate nuclear lens opacity in both eyes were found in the anterior segment. Fundus examination revealed myopic changes, including posterior staphyloma and mild chorioretinal atrophy around the optic disc. After the complete ophthalmological examination, no other cause but the corneal ectasia could be found to explain the severe vision loss.

Pachymetric maps, using the Pentacam, a Scheimpflug-based imaging system (Oculus Optikgeräte GmbH, Wetzlar, Germany), revealed a central pachymetry of 720 μ m in the right and of 710 μ m in the left eye, as well as an average paracentral pachymetry of 436 and 270 μ m in the 9-mm zone, respectively. The thinnest areas on the temporal side were 290 μ m in the right eye and 81 μ m in the left eye. These findings were confirmed by ultrasonic corneal pachymetry (Corneo-Gage™ Plus; Sonogage Inc., Cleveland, Ohio, USA). Corneal topography (EyeMap version 6.2; CSO, Firenze, Italy) revealed irregular and asymmetric bowing in both eyes. In addition, generalized bilateral steepening was observed with high corneal power (fig. 3). The patient underwent a second larger bilateral PK – 9 mm this time – in an attempt to improve visual acuity. Six months after this second keratoplasty, BCVA was 20/100 in both eyes.

Discussion

We present a case of a patient diagnosed with bilateral keratoconus and treated for this condition with a PK in both eyes. Thirty-four years later, he presented with decreased vision and bilateral keratectasia.

The initial diagnosis of bilateral keratoconus 34 years before is to be discussed. If we believe the diagnosis was right, late keratoconus recurrence on the corneal graft could be an explanation for the current situation. Increasing astigmatism should have been

expected as a sign of late progression of the keratoconus in the host cornea [9], but only a high spherical refractive error was found, with no astigmatic shift. Moreover, pachymetry of the corneal button was not reduced (715 and 709 μm), with no central thinning as would be expected in a keratoconus recurrence. Corneal thinning was observed outside the graft (436 and 270 μm in the 9-mm zone). Thus, if the patient did not have a recurrence but a keratoconus, the actual corneal profile could also be secondary to a severe PK wound thinning, developing many years after a graft. Nevertheless, the thinning usually observed in this situation is not so severe and not for 360°.

However, a misdiagnosed previous keratoglobus with a secondary keratectasia generated by a peripheral thinning of the cornea could also be a possible explanation for the actual corneal ectasia, which is more plausible for us. The diameter of the transplant would have been sufficient for a keratoconus, but was not large enough to stabilize the graft-host junction in a keratoglobus. The small-diameter PK significantly improved the patient's visual acuity, probably secondary to the flattening effect of the graft rather than to the resolution of his underlying condition. The mechanism of production of this globular corneal contour is not totally clear, but wound weakness in the area of the graft-host junction could be a possible cause. This idea was already stated by Cameron [1], who reported a case of limbus-to-limbus thinning and globular contour in a patient who underwent two PK in the same eye. He suggested that in most cases of keratoglobus, the marked mid-peripheral thinning could represent the relative weakness of this area and be the main predisposing factor for developing such a contour. In the patient described by Cameron, though, the thinning involved both the central and peripheral cornea and the IOP was 40 mm Hg, which could have contributed to the global ectasia. In our case, the IOP was normal and the mechanical stress of the grafts was not especially high.

In summary, we present a case with an end-stage evolution of a corneal ectasia and severe bilateral visual impairment, many years after a PK in both eyes. Whether this situation was originally caused by a keratoconus or by a misdiagnosed keratoglobus with secondary keratectasia cannot be cleared at the present time. Nevertheless, this new condition was satisfactorily treated with larger PK in both eyes.

Fig. 1. Slit-lamp photograph showing clear corneal button in the right eye, taken when the patient was admitted with complaints of decreased visual acuity.

Fig. 2. Picture showing the left (a) and right (b) eye cornea profile with severe corneal ectasia. Vertical and horizontal diameters were 11.0 and 11.6 mm in the right eye, and 11.0 and 11.8 mm in the left eye, respectively.

Fig. 3. Pentacam anterior corneal curvature map for the right eye (**a**) and the left eye (**b**), revealing irregular bowing with generalized steepening and high corneal power.

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