Occurrence of pseudoexfoliation following penetrating keratoplasty for keratoconus

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Abstract

The occurrence of pseudoexfoliation (PSX) following penetrating keratoplasty for keratoconus is reported in three eyes of two patients. The patients' ages at keratoplasty were 37, 40, and 31 years, and PSX was first observed 5, 4, and 6 years following keratoplasty. The donor age was 76, 81, and 81 years. Possible explanations for the occurrence of PSX after keratoplasty include mere coincidence, some sort of non-specific reaction to surgery or to postoperative medication, an immunogenic reaction to transplanted foreign corneal tissue, and the hypothetical speculation that PSX might be a transmissible disease.

Pseudoexfoliation (PSX) syndrome is clinically characterised by typical morphological changes of the anterior ocular segment. Despite numerous investigations its exact aetiology is still unknown.

We report three eyes of two relatively young patients with keratoconus in which PSX developed 4 to 6 years after corneal transplantation from donors over 75 years old.

Case reports

CASE 1

A 37-year-old white female was referred for treatment of bilateral keratoconus in 1981. Her family history and general history were uncontributory. Ocular examination of both eyes did not reveal any abnormality, except for bilateral keratoconus, especially no signs of PSX. Both eyes underwent 7.8/7.5 mm penetrating keratoplasty

(right eye on 29 October 1981, left eye on 4 October 1984). The donors were a 76-year-old male and an 81-year-old male, respectively who had both died of myocardial infarction. The donor eyes were not specifically screened for PSX and were not available for histological examination. Postoperative medications were local corticosteroids (prednisolone acetate 1%) for 1 year and scopolamine eyedrops for 6 weeks. The postoperative courses were unremarkable in both eyes, and corrected visual acuity was 6/6 right and 6/7 left after suture removal. Both eyes underwent regular slit-lamp examination every 3 to 6 months by one of us (GOHN). In November 1986, 5 years after keratoplasty, the right eye showed signs of PSX for the first time (Fig 1). In November 1988, 4 years after keratoplasty, the left eye had also developed signs of PSX after having been normal 5 months earlier. On last examination in October 1990 both eyes showed abundant PSX material at the pupillary margin and on the anterior lens capsule, but neither eye developed increased intraocular pressure or any signs of glaucomatous damage.

CASE 2

In 1989 a 46-year-old white male was referred for penetrating repeat keratoplasty of the right eye and keratoplasty for keratoconus of the left eye. The patient's son had bilateral keratoconus. In 1974 the patient had undergone a penetrating keratoplasty for keratoconus of the right eye elsewhere. The donor had been an 81-year-old female. The donor eye was not available for histological examination. Postoperative medication had consisted of local corticosteroids. During follow up, development of PSX of the

Figure 1 Right eye of patient 1. (A) Right eye in September 1984, 3 years after penetrating keratoplasty, without any signs of pseudoexfoliation. (B) Right eye in November 1986 displaying pseudoexfoliation material on the anterior lens capsule (arrows).

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Figure 2 A, B Right eye of patient 2 in January 1991, 17 years after the first and 18 months after the second penetrating keratoplasty, with typical pseudoexfoliation material on the anterior lens capsule (arrowheads).

right eye was observed for the first time in 1980 – 6 years after keratoplasty. The right eye had also developed a corneal graft rejection. On slit-lamp examination the right eye showed abundant PSX material at the pupillary margin and on the anterior lens capsule. The left eye was completely normal except for keratoconus. Both eyes underwent 7.5/7.8 mm penetrating keratoplasty (right eye on 18 October 1989, left eye on 3 June 1990). The postoperative course was unremarkable in both eyes. On last examination in January 1991 corrected visual acuity was 6/7 in both eyes. The right eye showed the typical picture of PSX (Fig 2) whereas in the left eye there were no signs of PSX. Neither eye developed increased intraocular pressure or glaucomatous changes.

Discussion

PSX is of considerable epidemiological and socioeconomical importance because of its high prevalence in the aged population, its frequent association with open-angle glaucoma, and the increased rate of complications in PSX eyes during cataract surgery.¹

Although the clinical picture, histological appearance, and ultrastructural characteristics of PSX have been thoroughly investigated in numerous studies the exact composition of PSX material as well as its aetiology are still obscure.

Recent ultrastructural studies²⁻⁴ have demonstrated PSX material in various extraocular locations thus indicating that PSX is not confined to intraocular structures but may represent a more widespread disease.

Several authors⁵⁻¹⁰ observed familial occurrence of PSX and assumed an autosomal dominant mode of transmission with incomplete penetrance. Other investigators^{11,12} stressed the importance of environmental factors in the development of PSX. Klouman¹³ and Ringvold *et al*¹⁴ conducted population based studies on PSX. Both authors found an unexpectedly high incidence of PSX in both partners of married couples, a finding which was supposed to indicate an environmental influence on the distribution of PSX.

Our observation of the development of PSX after penetrating keratoplasty could be interpreted in several ways. (1) The occurrence could be purely coincidental. However our patients' ages at the development of PSX (42, 44, and 37 years) was exceptionally young. PSX is usually seen in older patients and only occasionally occurs in patients under age 50 although the youngest patient with PSX reported in the literature was only 31 years.¹⁰ In our patients there was also a striking constancy of the interval between keratoplasty and development of PSX which was 4 to 6 years. (2) The development of PSX could represent a non-specific reaction to intraocular surgery or to local medication thus leading to the manifestation of PSX several years after keratoplasty. (3) The development of PSX could be the result of an immunogenic interaction between the host and the transplanted donor corneal tissue. The rather constant interval between keratoplasty and development of PSX could fit in with this hypothesis. (4) PSX could be a somewhat transmissible disease that in our patients was transmitted from the donors to the recipients by means of the corneal buttons. Unfortunately we do not know whether the donor eyes showed signs of PSX but as the donors were between 76 and 81 years of age and the prevalence of PSX is high in this age group it is possible that the donor eyes were affected by PSX. This speculation would again go well with the typical interval between keratoplasty and development of PSX in our patients and would also give some sort of explanation of the frequent occurrence of PSX in married couples seen by other investigators.^{13,14}

Although the exact meaning of our observations is uncertain at present similar findings by other investigators may give important clues of the aetiology and pathogenesis of PSX in the future.

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