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**SHORT COMMUNICATION**

# Corneal graft rejection after penetrating keratoplasty for keratoconus in Turner's syndrome

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PURPOSE. *To report a patient with Turner's syndrome who developed graft rejection after penetrating keratoplasty (PK) for keratoconus and to review the ophthalmic literature on the association between keratoconus and Turner's syndrome.*
METHODS. *A woman with bilateral keratoconus and Turner's syndrome (45,XO) was referred for progressive visual loss in the right eye. Best-corrected visual acuity was 20/400 in the right eye. Slit-lamp examination revealed corneal thinning with ectatic protrusion of the central cornea and Vogt's striae in the right eye. The patient underwent PK in the right eye in January 2001. She developed graft rejection in April 2003 and visual acuity dropped to hand motion. After treatment with topical and systemic steroids and systemic cyclosporine A, visual acuity recovered to 20/80 in July 2003.*
RESULTS. *The authors know of only three other reported patients (six eyes) with keratoconus in Turner's syndrome. Five eyes underwent PK with good visual rehabilitation, but one developed immunologic graft rejection 7 years after surgery. On the whole, considering the current report and the other cases described in the literature, graft rejection occurred in 2 out of 6 eyes (33.3%). The graft survival rate was 80% after 2 years and 40% after 7 years.*
CONCLUSIONS. *The results suggest that grafts for keratoconus in patients with Turner's syndrome might have an increased risk of immunologic rejection. Corneal grafts in Turner's syndrome need to be monitored closely. Early detection of graft rejection and aggressive treatment with topical and systemic steroids and systemic cyclosporine A can save the graft and restore useful vision. (Eur J Ophthalmol 2005; 15: 271-3)*

KEY WORDS. *Graft rejection, Keratoconus, Turner's syndrome*

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## INTRODUCTION

Turner's syndrome is a chromosomal anomaly in which phenotypic females have a missing or abnormal X chromosome (45,XO). The estimated incidence is 1:3000 live female births (1).

Ovarian dysgenesis, primary amenorrhea, undeveloped breasts, infantile genitalia, scanty pubic hair, short stature, web neck, shield chest, and cubitus valgus are cardinal features of the syndrome. Aorta coarctation, cardiac anomalies, multiple pigmented nevi, hypertension, urinary tract anomalies, and congenital lymphedema occur often.

The ocular findings include a wide spectrum of abnormalities (Tab. I) (2). Keratoconus is exceedingly rare in patients with Turner's syndrome.

We report a patient with Turner's syndrome who developed graft rejection after penetrating keratoplasty (PK) for keratoconus.

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## Case report

A 35-year-old woman with bilateral keratoconus and Turner's syndrome was referred for progressive visual loss in the right eye. She had used gas permeable contact lenses for years. There was no history of eye rubbing or atopy, or a family history of keratoconus. She was of average intelligence and had successfully completed high school. Best-corrected visual acuity was 20/400 in the right eye and 20/40 in the left. Central keratometry showed distorted mires and disclosed irregular astigmatism of 6 D in the right eye and 4 D in the left. Slit-lamp examination revealed bilateral corneal thinning with ectatic protrusion of the central cornea and Vogt's striae in the right cornea. The lenses were clear and the intraocular pressures normal. Fundus examination was unremarkable. The patient underwent uneventful PK in the right eye in January 2001. An 8.0 mm diameter corneal button was sutured into a 7.5 mm wound with a double continuous 10-0 nylon suture. Postoperative therapy consisted of topical tobramycin (0.3%) four times daily for 10 days and topical dexamethasone (0.1%), initially given four times daily for 10 days and then gradually tapered in 3 months. She had no short-term postoperative complications and best-corrected visual acuity was 20/40 in the right eye in December 2001. Although there were no factors that could have increased the risk of graft rejection, such as vascularization of the host rim/graft, suture related problems, or noncompliance, the patient developed endothelial rejection in April 2003 and visual acuity dropped to hand motion. After treatment with intensive topical dexamethasone (0.1%), intravenous methylprednisolone, and oral cyclosporine A (200 mg daily), visual acuity recovered to 20/80 in July 2003.

## DISCUSSION

Keratoconus has been reported to occur frequently in patients with Down syndrome, in patients with atopy, or in patients who rub their eyes (3-6). The microtrauma associated with eye rubbing is generally thought to be the etiologic link between keratoconus and associated systemic and ocular diseases. Conversely, keratoconus is very rarely seen in patients with Turner's syndrome. Its etiology is unknown; however, Nucci et al (7) postulated that corneal thinning in Turner's syndrome might be an expression of a mesodermal defect, because mesodermal structures are sometimes affected in Turner's syndrome. In the case presented here, keratoconus appeared to be idiopathic. Indeed, there was no history of eye rubbing or atopy, or family history of keratoconus.

We know of only three other reported cases of keratoconus in Turner's syndrome, all with bilateral involvement (2, 7). Five eyes underwent PK with good visual rehabilitation, but one of them developed immunologic graft rejection 7 years after surgery (2). Graft rejection was successfully treated with topical steroids. On the whole, considering our report and the other patients described in the literature, graft rejection occurred in 2 out of 6 eyes (33.3%). The graft survival rate determined using the Kaplan-Meier method (8) was 80% after 2 years and 40% after 7 years.

Although our sample includes a limited number of eyes and the number of cases reported in the literature is not representative of the entire population, results suggest that grafts for keratoconus in patients with Turner's syndrome might have a higher risk of immunologic rejection than grafts for keratoconus in patients with normal karyotype, which have 5-year and 10-year survival rates of 97% and 92% (9). Possibly, the donor cornea keratocytes, which have normal 46,XY or XX karyotype, might

**TABLE I -** OCULAR FEATURES ASSOCIATED WITH TURNER'S SYNDROME

| | | | |
|---|---|---|---|
| Bilateral epicanthus | Amblyopia | Hypermetropia | Strabismus |
| Congenital cataract | Ptosis | Myopia | Nystagmus |
| Color blindness | Hypertelorism | Blue sclera | Iris coloboma |
| Congenital glaucoma | Corneal nebulae | Brushfield spot | Congenital lymphangiectasia |
| Choroidal coloboma | Microcornea | Microphthalmos | Persistent pupillary membrane |
| Eccentric pupils | Lid hemangioma | Abducens palsy | Absence of the caruncle |
| Retinitis pigmentosa | Oval cornea | Duane's syndrome | Lachrymal gland hypoplasia |
| Anti-mongoloid slant | Keratoconus | Diminished retinal pigmentation | |

act as potential triggering factors for immunologic graft rejection in patients with 45,XO karyotype.

Corneal grafts in Turner's syndrome should be monitored closely for immunologic rejection. It is also important to educate these patients about the symptoms of rejection and emphasize the need for prompt medical care to save the graft. Early detection of graft rejection and aggressive treatment with topical and systemic steroids and systemic cyclosporine A can save the graft and restore useful vision.

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