CASE REPORT

Fuchs' superficial marginal keratitis managed with circumferential marginal corneoscleral lamellar patch graft

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Purpose: *To report a case of Fuchs' superficial marginal keratitis managed with circumferential marginal corneoscleral lamellar patch graft.*
Methods: *Interventional case report.*
Results: *A 34-year-old man presented with several years' history of ill-defined symptoms of binocular ocular irritation associated with vision loss, mostly in the left eye. A superior marginal corneal thinning was found at biomicroscopy of the left eye, with 2 finely vascularized descemetoceles, and a gray epithelial demarcation line without lipid infiltrates. The right eye was clinically normal. Anterior segment optical coherence tomography demonstrated an important corneal thinning from the 7 o'clock to 3 o'clock positions, without scleral involvement. A circumferential marginal corneoscleral lamellar patch graft was done involving 3 mm of sclera and 3 mm of cornea.*
Conclusions: *Fuchs' superficial marginal keratitis is a rare entity, mostly affecting young adults. It should be considered part of a spectrum of corneal thinning disorders, together with Terrien's marginal degeneration. Definitive treatment with a marginal corneoscleral lamellar patch graft with or without conjunctival autograft is suggested.*

Keywords: *Corneoscleral patch graft, Descemetocele, Fuchs', Terrien's*

Accepted: August 2, 2013

INTRODUCTION

Fuchs' superficial marginal keratitis is a rare entity, first described by Ernst Fuchs in 1889 (1). It usually affects young adults, bilaterally but asymmetrically. It is characterized by progressive marginal corneal thinning, recurrent red eye episodes, and visual acuity reduction secondary to irregular astigmatism. Spontaneous ocular perforation seldom occurs (2).

CASE REPORT

A 34-year-old Hispanic man presented with several years' history of recurrent ill-defined red eye episodes, mainly in the left eye, visual acuity loss secondary to myopic astigmatism, and poor tolerance to soft contact lenses and later to rigid gas-permeable lenses. The patient denied previous ocular surgery or trauma.

He was referred to the cornea service, where his corrected visual acuity was found to be 20/20 OD (–2.00 –3.00 × 5) and 20/70 OS (–1.75 –8.75 × 145). The right eye was clinically normal. The left eye had healthy eyelids and conjunctiva; there was a clockwise corneal marginal thinning between 7 and 4 o'clock (Fig. 1) with variable depth and radial extension that included 2 superior marginal descemetoceles at the 10 and 12 o'clock positions; these descemetoceles were approximately 3 mm in diameter, had a superficial fine vascularization, and had a subtle gray epithelial anterior demarcation line. There were no

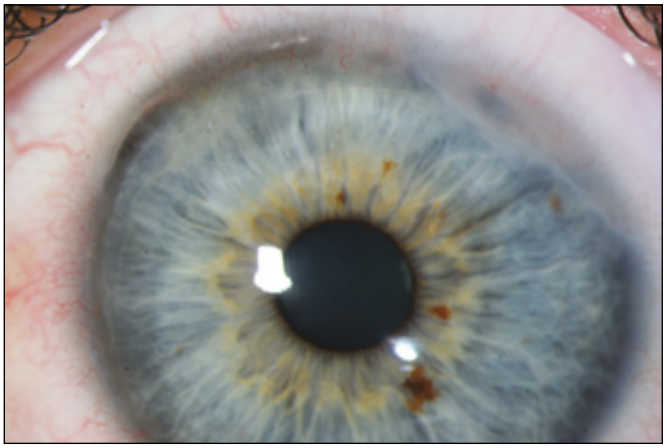

Fig. 1 - *Left eye. Note peripheral corneal involvement form 7 o'clock to 4 o'clock (clockwise), descemetoceles at the 10 and 12 o'clock positions, superficial vascularization between 12 and 3 o'clock, and gray epithelial line between 1 and 3 o'clock.*

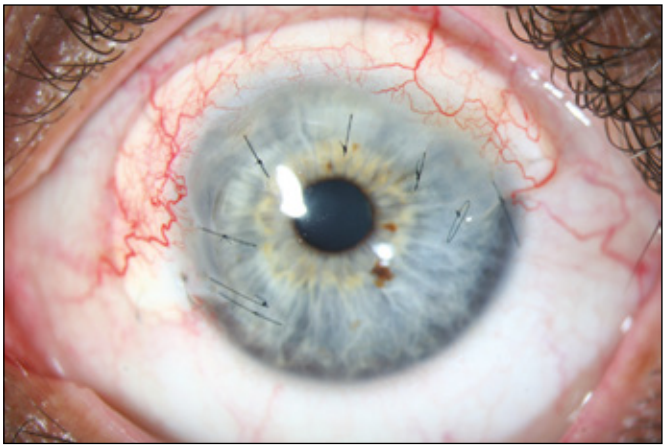

Fig. 2 - *Sixth postoperative month.*

lipid deposits. Intraocular pressure was 12 mm Hg OU. The remaining ocular examination was normal.

Anterior segment optical coherence tomography demonstrated an extensive and variable peripheral corneal thinning between 7 and 3 o'clock positions, with normal scleral thickness.

Corneal elevation topography of the left eye showed an oblique and irregular astigmatism, normal central pachymetry, and extensive peripheral superior thinning. There was no pathologic anterior or posterior elevation.

A complete systemic workup to rule out underlying inflammatory diseases was done and was normal.

A diagnosis of Fuchs' superficial marginal keratitis was made. Given the imminence of a corneal perforation, a 270-degree allogenic circumferential marginal corneoscleral lamellar patch graft was performed without HLA or ABO matching, comprising 3 mm of sclera and 3 mm of cornea, at a 50% depth in sclera and at the pre-Descemet plane in cornea extending 3 mm from the limbus towards both sides. The cornea donor was a 55-year-old man; death to preservation time was 10 hours, and preservation media was MK. The donor graft was obtained with the same dimensions from a whole ocular globe and sutured with radial interrupted nylon 10-0 stitches, after which the eye was occluded for 24 hours; no contact lens was used at this time or afterwards. Moxifloxacin chlorhydrate 0.5% and dexamethasone phosphate 0.1% eyedrops (Vigadexa, Alcon Laboratories, Fort Worth, Texas, USA) 5 times a day was prescribed during the first postoperative month, after which prednisolone acetate 1% (Pred-F, Allergan, Irvine, California, USA) was prescribed for the following 6 postoperative months (Fig. 2). No oral/systemic medications were prescribed. The patient was followed up monthly for the first 6 months, then bimonthly up to the first postoperative year, and every 6 months thereafter. Postoperatively, the eye was quiet at all times, without intraocular inflammation or any sign of graft rejection or recurring thinning. At the last follow-up (18th postoperative month) (Fig. 3) keratometry, corneal topography and refraction was still variable, so no refractive correction (glasses, contact lenses) has been offered.

DISCUSSION

Fuchs' superficial marginal keratitis is a rare entity characterized by marginal corneal thinning, usually bilateral, asymmetric, and progressive. It most commonly affects young adults between the 2nd and 4th decades of life. It often has a chronic course with recurrent bouts of red eye, pain, and tearing (2), associated with a marginal stromal thinning, which typically is irregular in its depth and axial extension, delimited by a fine intraepithelial gray line on its advancing edge and without any accompanying lipid deposits (2, 3). The marginal thinning does not have a preferred limbal location and is frequently associated with a pseudo-pterygium (3, 4). Visual acuity can be compromised in advanced cases secondary to irregular astigmatism.

The main differential diagnosis of Fuchs' superficial marginal keratitis is Terrien's marginal degeneration (3). Both

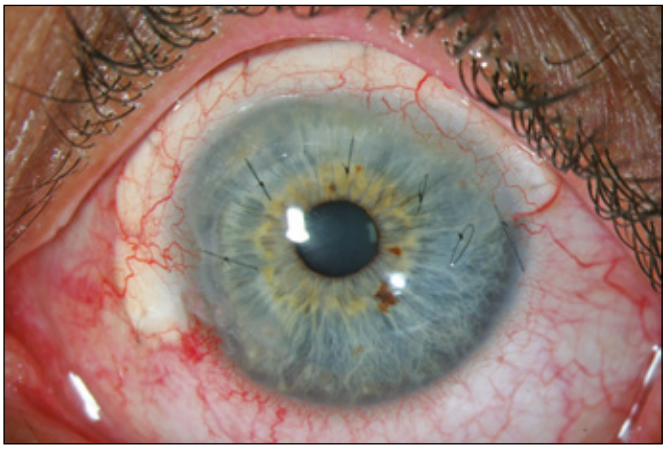

Fig. 3 - *The 18th postoperative month. Only 2 sutures have been removed, because of loosening.*

entities affect young adults; exhibit a paralimbal stromal thinning, usually bilateral, asymmetric, and progressive (3); a pseudo-pterygium occurs in either disease, although more classically in Fuchs' superficial marginal keratitis (3); and either one can lead to a spontaneous or traumatic corneal perforation (5). Because of these similarities, some authors had suggested that both disorders are manifestations of the same disease (3).

Fuchs' superficial marginal keratitis differentiates from Terrien's marginal degeneration in that it affects the limbus at any localization (no preference for superior cornea), presents with accompanying epithelial defects, has an epithelial delimitation gray line, and does not have lipid deposition (2, 3).

Fuchs' superficial marginal keratitis treatment depends on its stage. Symptoms improvement with the use of lubricants (4), topical steroids (3, 4), oral doxycycline (3), and vitamin C (3) in the acute phase has been reported. Cyanoacrylate has transient usefulness in corneal microperforations.

There are a few case reports treated with penetrating keratoplasty (2), lamellar keratoplasty (6), amniotic membrane reconstruction (4), superficial keratectomy with conjunctival autograft (3, 4), and corneoscleral lamellar patch graft. We decided to perform a 270-degree circumferential marginal corneoscleral lamellar patch graft in this patient, aiming to provide a complete tectonic solution for the peripheral cornea. Because the thinning involved the cornea up to the limbus, a corneoscleral graft was chosen in order to provide a good fixing plane for the donor tissue. We are not aware of any published report using a 270-degree corneoscleral lamellar patch graft to manage this entity.

Fuchs' superficial marginal keratitis is a rare disorder that shares several characteristics with Terrien's marginal degeneration, to the extent that they can be considered a different manifestation of the same degenerative marginal corneal disease whose etiology is unknown. Definitive treatment with a circumferential corneoscleral lamellar patch graft provides good anatomical and functional reconstruction in advanced cases.

Financial Support: No financial support was received for this submission.

Conflict of Interest Statement: None of the authors has conflict of interest with this submission.

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