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SHORT COMMUNICATION

Unilateral corneal endothelial dystrophy and anterior keratoconus

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PURPOSE. *To describe a case of keratoconus and Fuchs' corneal endothelial dystrophy in the left eye with no corneal disease in the right eye.*
METHODS. *A 64-year-old woman presented with visual impairment in her left eye; computer-assisted topographic analysis and specular microscopy were performed in both eyes and left cornea was histopathologically examined.*
RESULTS. *Keratoconus was diagnosed by slit-lamp examination, keratometry, and computer-assisted topographic analysis. Corneal endothelial dystrophy was diagnosed on the basis of clinical examination and specular microscopy. Histopathologic examination revealed a stromal degeneration typical of keratoconus and a non-guttae form of endothelial dystrophy.*
CONCLUSIONS. *This is a rare case of unilateral corneal endothelial dystrophy and keratoconus. (Eur J Ophthalmol 2007; 17: 430-2)*

KEY WORDS. *Keratoconus, Corneal endothelial dystrophy, Histopathology*

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INTRODUCTION

Keratoconus is a noninflammatory corneal ectasia that results in thinning and protrusion of the cornea. Bilateral involvement occurs in as many as 90% of cases (1). Keratoconus has been reported in association with corneal dystrophies (2).
Fuchs' corneal endothelial dystrophy is characterized by a gradual dysfunction of the endothelium leading to stromal and endothelial edema (3). Histopathologically it consists of endothelial cell loss, thickening of Descemet membrane, and focal endothelial excrescences (guttae). However, a non-guttae form of Fuchs' dystrophy has been described as a variant form of the same disease (4).
Herein we describe a patient with unilateral keratoconus and the non-guttae variant of Fuchs' corneal endothelial dystrophy.

Case report

A 64-year-old woman was referred because of visual impairment in her left eye. Her past ocular history was positive for unilateral keratoconus in her left eye diagnosed 10 years earlier.
Neither history of ocular inflammation and trauma nor significant general medical history was present. Family history of ocular disease was negative.
Best-corrected visual acuity was 20/25 in her right eye and 20/100 in her left eye.
Left eye slit lamp examination showed inferior corneal thinning and protrusion associated with a diffuse beaten-metal endothelial appearance. No corneal abnormalities were detected in the right eye. A mild cataract was present in both eyes.
Posterior segment and intraocular pressure were normal

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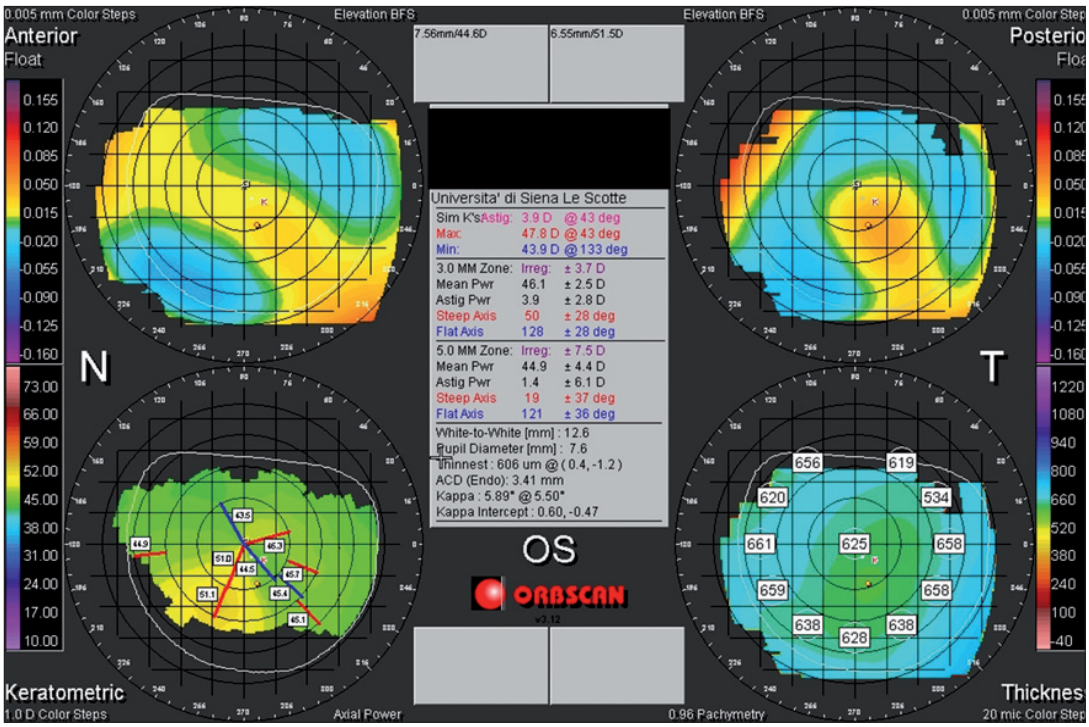

Fig. 1 - *Orbscan analysis reveals an inferior corneal steepening with a mild increase of thickness.*

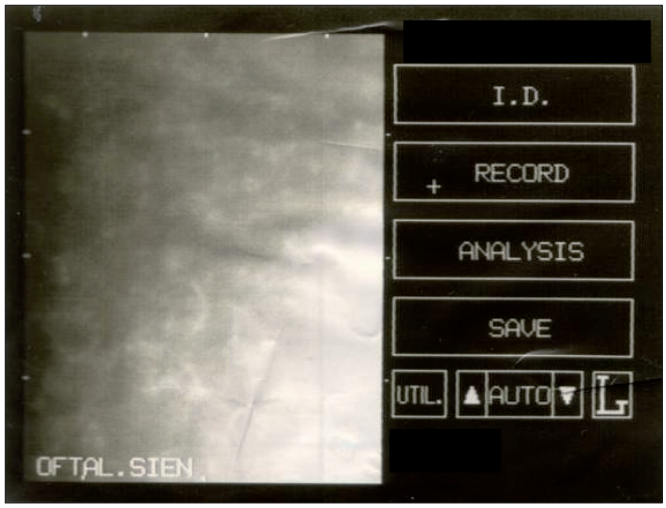

Fig. 2 - *Specular microscopy with a completely disorganized pattern of the endothelium.*

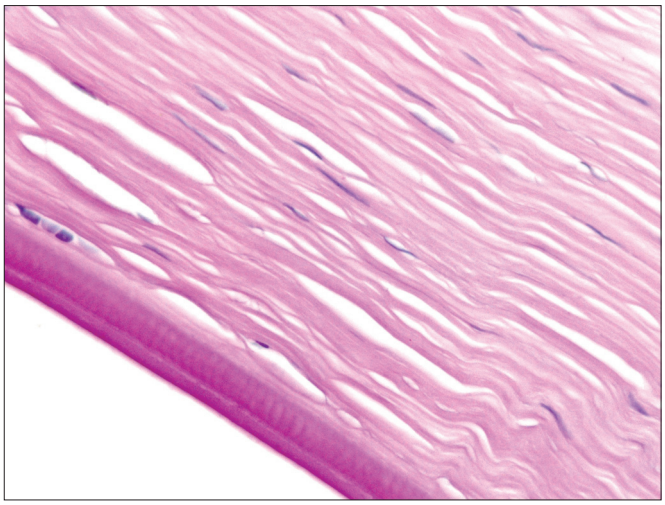

Fig. 3 - *Hematoxylin-eosin: Uniform increased thickness of Descemet membrane without guttae (original magnification x200).*

in both eyes.

Orbscan (Bausch & Lomb) analysis showed an inferior steepening (52.3 D) with a corneal thickness of 636 µm in the left eye (Fig. 1).

Specular microscopy of the left eye showed a completely disorganized endothelial pattern (Fig. 2). Orbscan and specular microscopy of the right eye were within normal limits.

The patient underwent corneal transplantation in her left eye.

Histopathologic analysis of the excised button revealed marked central stromal ectasia with folds and focal dehiscences in Bowman's membrane that confirmed the previous clinical and topographic picture suggestive of keratoconus. In addition, a decreased number of endothelial

cells with a thickened Descemet membrane without prominence of guttate structures were present, supporting the clinical diagnosis of a non-guttae form of Fuchs' corneal endothelial dystrophy (Fig. 3).

DISCUSSION

Lipman et al (5) described coexistent bilateral Fuchs' dystrophy and keratoconus in a 44-year-old patient; the examination of the patient's family revealed keratoconus in the patient's son and central guttata and abnormal endothelial cells in the patient's mother and daughter (5). Orlin et al (6) described coexistent bilateral Fuchs' dystrophy and keratoconus in five patients. Jurkunas and Azar described the complications of cataract and refractive surgery in eight patients with bilateral Fuchs' endothelial dystrophy and keratoconus (7).

Fuchs' dystrophy is characterized by a gradual dysfunction of endothelium with a diffuse beaten-metal aspect. It consists of endothelial cell loss and thickening of Descemet membrane. Focal endothelial excrescences (guttae) are characteristic, but, as found by Stocker (8), not visible by light microscopy in 20% of cases. This supports the view that both guttae and non-guttae Fuchs' corneal endothelial dystrophy are morphologic variants of the same disease (8).

Herein, we illustrate a patient with the non-guttae variant of Fuchs' endothelial dystrophy, diagnosed by specular microscopy and confirmed by the finding of mild increase of corneal thickness in a keratoconus cornea (corneal apex = 636 μm).

Interestingly, unilateral endothelial dystrophy was associated with keratoconus only in the left eye, while no corneal disease was present in the right eye.

Although this association could be coincidental, we hypothesize that it might be explained either by a currently unknown common genetic transmission or by secondary damage of the posterior cornea caused by keratoconus. In fact, as suggested by Matsuda et al, the endothelium in corneas affected by keratoconus presents an unstable state which could predispose it to damage (9).

None of the authors has proprietary or financial interest in any material or device mentioned.

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