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# Fuchs Endothelial Corneal Dystrophy and Keratoconus: A Very Rare Coincidence

## *Fuchs Endoteliyal Korneal Distrofi ve Keratokonus: Çok Nadir bir Tesadüf*

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**Abstract**

It was aimed to represent a case with concurrent Fuchs endothelial corneal dystrophy (FECD) and keratoconus (KC) as a rare entity. A 35-year-old woman had a best-corrected visual acuity was 20/20 in the right eye and 20/60 in the left eye (Snellen). Biomicroscopy revealed bilateral cornea guttata and Fleischer ring in the left eye. Corneal topography demonstrated early KC in the right eye and advanced KC in the left eye. Maximum keratometry (Kmax) and pachymetry at the thinnest location were 46.2 diopters (D) in the right eye and 56.3 D in the left eye, and 530 and 495 microns, respectively. Corneal thinning in KC and subclinical corneal thickening in FECD might lead delay in disease diagnosis.

**Keywords:** Corneal dystrophy; endothelium; fuchs; keratoconus; topography.

**Özet**

Nadir bir antite olarak Fuchs endotel korneal distrofisi (FECD) ve keratokonus (KC) birlikteliği olan bir olgu sunmak amaçlanmıştır. Otuz beş yaşında bir kadın, sağ göz görme keskinliği 20/20 sol göz 20/60 (Snellen) olarak başvurdu. Biyomikroskopide bilateral kornea guttata ve sol gözde Fleischer halkası görüldü. Kornea topografisi sağ gözde erken KC ve sol gözde ileri KC gösterdi. Korneanın en ince yerinde maksimum keratometri (Kmax) ve pakimetri sağ ve sol gözde sırasıyla 46.2 diyoptri (D) ve 56.3 D ve 530 ve 495 mikron idi. KC'de kornea incelmesi ve FECD'de subklinik kornea kalınlaşması hastalıların tanısında gecikmeye neden olabilir.

**Anahtar Kelimeler:** Korneal distrofi; endotelyum; fuchs,; keratokonus; topografi.

## Introduction

Keratoconus (KC) is an asymmetric corneal ectasia characterized by progressive myopia, irregular astigmatism and stromal thinning (1,2). Although clinical examination is enough for diagnosing moderate and advanced KC cases, advanced imaging techniques such as corneal topography or tomography and biomechanical assessment are necessary in early KC. Fuchs endothelial corneal dystrophy (FECD) is a bilateral slowly progressing dystrophy of corneal endothelium and the most common cause of corneal transplantation (3). Guttae formation, which are focal posterior excrescences of Descemet's membrane (DM), and accelerated endothelial cell loss are the hallmarks of the FECD (3,4). Clinical findings range from asymptomatic corneal thickening to significant visual deterioration with painful corneal edema that shows diurnal variation (3–5). The coincidence of FECD and KC is very rare and total number is too low worldwide based on the current literature (6). This case-report aims to represent clinical and topographical features of a young patient with concurrent FECD and KC as a very rare entity, as well to emphasize clinical importance of this condition.

## Case

A 35-year-old woman presented with low vision in her left eye. Uncorrected distance visual acuity (UDVA) was 0.8 OD / 0.05 OS (Snellen equivalent); and best spectacle-corrected distance visual acuity (CDVA) was 1.0 (-0.50/-0.25x180) OD and 0.3 (-4.0/-4.0x60) OS (Snellen equivalent). Biomicroscopic examination revealed bilateral cornea guttata without clinical corneal edema (Figure 1), and Fleischer ring OS. Crystalline lens was clear in both eyes. Intraocular pressure (IOP) measurements (Goldman applanation tonometer) and fundus examination were within normal limits.

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**Figure 1**

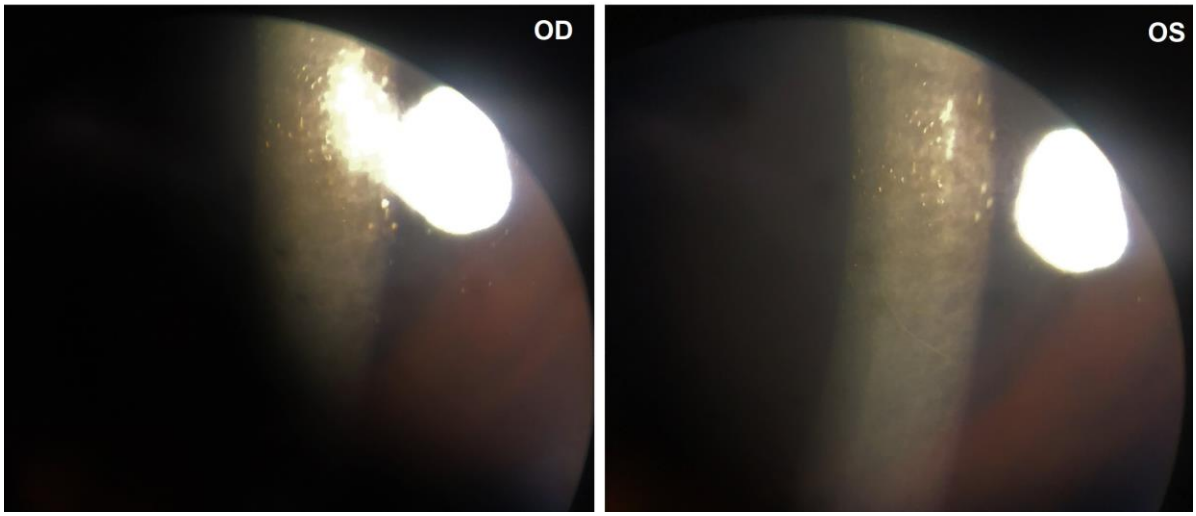


**Figure 1.** Slit-lamp biomicroscopic images of the patient showing bilateral *cornea guttata* indicative of Fuchs endothelial corneal dystrophy.

**Figure 2**

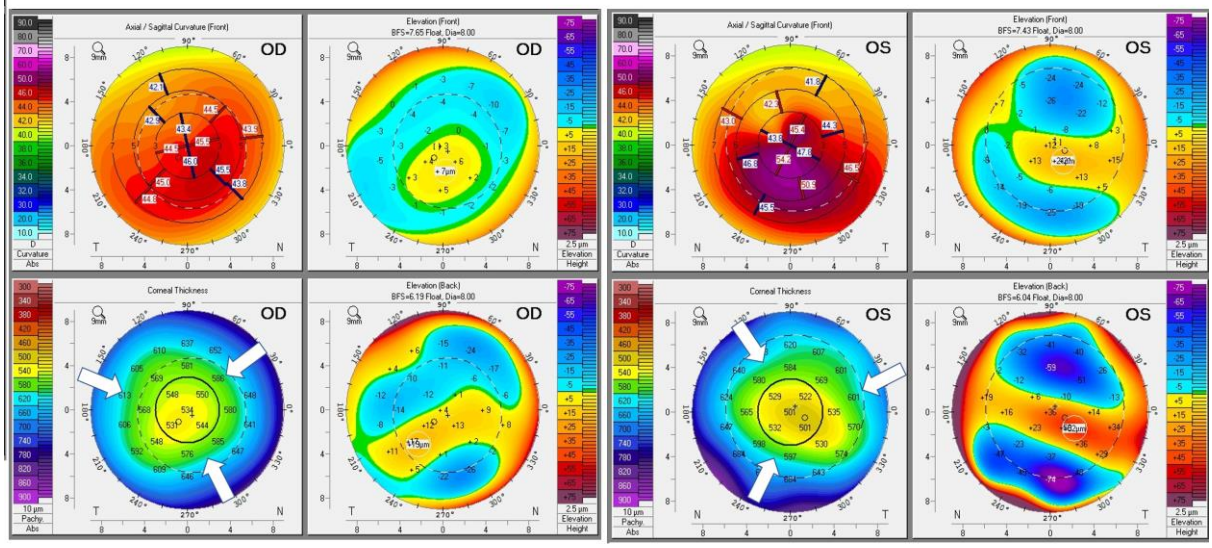


**Figure 2.** Corneal topographic maps (axial/sagittal curvature, front elevation, corneal thickness and back elevation) (Pentacam HR (Oculus Optikgerate, Wetzlar, Germany) demonstrated early keratoconus (KC) in the right eye and advanced KC in the left eye, with mild loss of regular isopachs in the corneal thickness map (outlined by the white arrows).

Specular microscopy of corneal endothelium failed to give a reliable result in both eyes. Corneal topography (*Pentacam HR, Oculus Optikgerate, Wetzlar, Germany*) revealed early KC in the right eye and advanced KC in the left eye (Figure 2). Maximum keratometry (K*max*) and back elevation values for OD/OS were 46.2/56.3 diopters and 21/52 microns, respectively. Thinnest pachymetry was 530 microns OD and 495 microns OS. Moreover, mild loss of regular isopachs was observed in corneal thickness maps bilaterally (Figure 2). The patient was diagnosed as concurrent FECD and KC. She was informed about rigid gas-permeable contact lenses for visual rehabilitation (for the left eye); however, she preferred spectacle correction. The patient was unaware of family history regarding these diseases and she is being followed up for progression of KC and FECD. The patient signed an informed consent to participate in this case report.

## Discussion

This report presented clinical and topographical features of a patient with concurrent FECD and KC to draw attention to its clinical importance. In the current literature, 69 similar cases were reported up to date by 15 publications based on a recent review (6). In this case, in addition to cornea guttata and typical findings of KC, mild loss of regular isopachs was noted in the pachymetric map. This finding was previously demonstrated by Sun et al (4) and Patel et al (5) in early FECD with subclinical corneal edema. Furthermore, they also showed nasalization of the thinnest corneal point and focal posterior corneal surface depression in this group of patients (4,5). In the current case-report, deviation in the thinnest corneal point was not so prominent to reach the nasal sector. On the other hand, posterior surface depression seen in FECD might have reduced the amount of posterior elevation in our case, since back elevation values in the right eye were in borderline. The left eye had advanced KC. Hence, potential depressive effect of FECD on posterior corneal surface appears to be eliminated by significant back elevation. The coincidence of FECD and KC might have clinical consequences; i.e. subclinical corneal edema in eyes with FECD might mask topographical findings of KC, particularly in early KC as in this report and likewise stromal thinning in KC might lead underestimation of corneal edema in FECD (6,7). Moreover, fluctuations in corneal thickness measurements might also affect surgical planning of the surgeon. For instance, 400 microns of corneal thickness (after removal of epithelium) is the safety limit to perform a standard CXL procedure in eyes with progressive KC and diurnal variation in corneal thickness in FECD might mislead the surgeon regarding corneal thickness assessment (8,9). This condition might result in unexpected postoperative outcomes and serious complications (3–5). A previous study by Jurkunas and Azar (7), suggested that disease severity could be underestimated in patients with concurrent FECD and KC; and the surgeon might be faced with more frequent complications in the area of cataract and refractive surgery. Similarly, Mylona et al (6) reviewed 69 cases with concurrent FECD and KC reported by 15 studies in the literature. They concluded that the coincidence of FECD and KC was very low and treatment planning should involve preoperative topography and specular microscopy to avoid unexpected outcomes and complications in these patients as also stated by Jurkunas and Azar (6,7).

## Conclusion

Although FECD and KC coincidence is uncommon, clinical importance of this combination should be kept in the mind and an all layer-corneal assessment with ancillary tests appears to be necessary.

**Informed consent:** The patient signed an informed consent to participate in this case report.
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