Corneal Biomechanical Properties and Densitometry in a Case with Iridocorneal Endothelial Syndrome

İridokorneal Endotelyal Sendromlu Bir Olguda Korneanın Biyomekanik Özellikleri ve Korneal Dansitometri

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ABSTRACT Iridocorneal endothelial (ICE) syndrome is a rare disorder which includes iris nevus syndrome, Chandler syndrome and essential iris atrophy; and represents a spectrum of the similar disorder process predominantly affecting the corneal endothelium, anterior chamber angle and iris. ICE syndrome is often under-diagnosed especially in early stages when there is no apparent corneal edema and glaucoma. The accurate diagnosis is mainly based on clinical examination and in-vivo confocal microscopy by showing epithelium-like transformation of the corneal endothelium with irregularly shaped cells and hyperreflective nuclei. We hereby report corneal biomechanical properties and densitometry in a 49-year-old patient with ICE syndrome and discuss role of these parameters for the diagnosis and follow-up of ICE syndrome.

Key Words: Iridocorneal endothelial syndrome; corneal topography

ÖZET İridokorneal endotelyal sendrom temel olarak kornea endoteli, ön kamara açısı ve irisin etkilendiği; iris nevüs sendromu, Chandler sendromu ve esansiyel iris atrofisi gibi benzer bir hastalığın farklı spektrumundaki hastalıkları içeren nadir bir hastalıktır. Kornea ödemi ve glokomun henüz ortaya çıkmadığı erken hastalık evrelerinde tanı koymak güçtür. Kesin tanısında klinik bulgularla birlikte in-vivo konfokal mikroskopik bulgular önemli rol oynar. Konfokal mikroskop ile kornea endotelinde hiperreflektif nükleuslu düzensiz şekilli hücrelerden oluşan epitel benzeri bir dönüşüm izlenir. Bu yazıda iridokorneal endotelyal sendromu tanısı alan 49 yaşındaki bir olguda korneanın biyomekanik özellikleri ve korneal dansitometri ölçümleri sunularak bu değişkenlerin hastalığın tanı ve takibindeki rolü tartışılmıştır.

Anahtar Kelimeler: Iridokorneal endotelyal sendrom; kornea topografisi

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Iridocorneal endothelial (ICE) syndrome which includes iris nevus syndrome, Chandler syndrome and essential iris atrophy represents a spectrum of the similar disorder process predominantly affecting the corneal endothelium, anterior chamber angle and iris.[1-3] The accurate diagnosis is mainly based on confocal microscopy by showing epithelium-like transformation of the corneal endothelium with irregularly shaped cells and hyperreflective nuclei.[4,5] The aim of our report is to present corneal densitometry and biomechanical properties in a patient with ICE syndrome and discuss role of these parameters for the diagnosis and follow-up of ICE syndrome.

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CASE REPORT

A 49-year-old woman was presented with a chief complaint of distorted pupillary shape in her left eye. The patient reported no systemic disease and no previous history of an ophthalmic disorder or ocular trauma. Ophthalmological examination of the right eye was unremarkable (Figure 1A). Visual acuity was 20/20 on her left eye and slit-lamp examination revealed an iris atrophy at 3 o'clock position and an infero-nasally displaced pupil toward an area of anterior synechia with an ectropion uvea at 8 o'clock position (Figure 1B). No clinically prominent corneal edema was present. The intraocular pressure was 15 mmHg and dilated fundus examination was normal at her left eye. Central corneal thickness measured by ultrasonic pachymetry was 570 and 590 μm in the right and left eyes, respectively. ICE syndrome was suspected and confocal microscopy (Confoscan 3.0, Nidek, Vigonza, Italy), corneal topography (Pentacam HR, Oculus, Inc., Wetzler, Germany) and ocular response analyzer (ORA) (Reichert Ophthalmic Instruments, Depew, NY, USA) was used for further evaluation of the patient.

In-vivo confocal microscopy revealed a normal hexagonal cell morphology of corneal endothelium at her right eye (Figure 2A) and an epithelium-like transformation of the corneal endothelium with irregularly shaped cells and hyperreflective nuclei in the left eye (Figure 2B). Corneal biomechanical properties were altered in the affected eye (Table 1). The average cornea densitometry of the left eye were higher when compared to right eye (18.6 vs 15.7 grayscale units-GSU). The corneal densitometry in all concentric radial zones and anteroposterior layers were higher in the affected eye when compared to fellow eye (Table 2).

DISCUSSION

ICE syndrome is often under-diagnosed especially in early stages when there is no apparent corneal edema and glaucoma. Visualisation of the cornea endothelium is essential for differential diagnosis of this disorder. This first report on corneal densitometry and biomechanical properties of a patient with ICE syndome was designed to discuss the role of these parameters on the diagnosis and follow-up of this rare condition. Decreased corneal hysteresis (CH) and resistance factor (CRF) and increased corneal densitometry in the affected eye of the patient can facilitate the early and accurate diagnosis, may reveal complementary information about the disease process and can be used as an objective measure in order to monitor disease progression. Even though no significant corneal change was present in slit-lamp biomicroscopy,

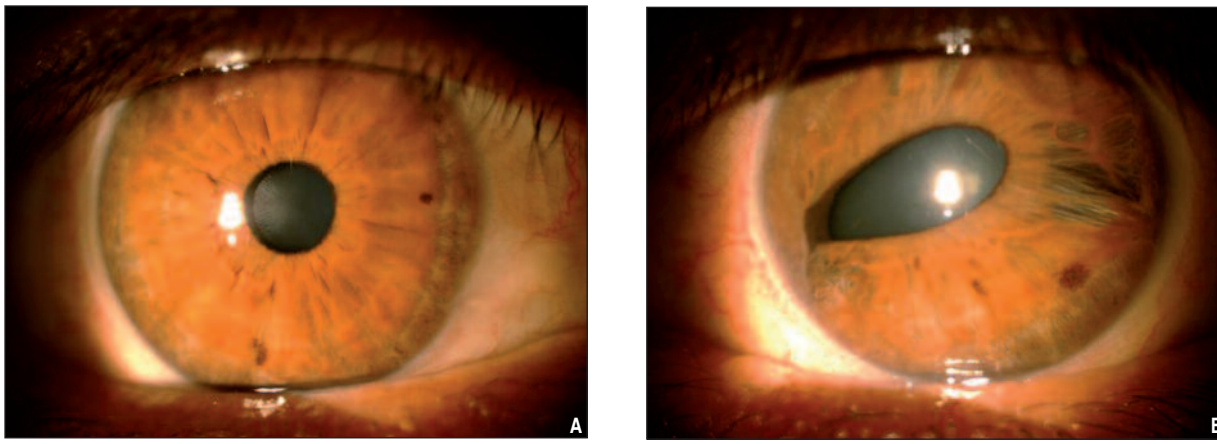

FIGURE 1A,B: Slit-lamp photograph of a 49-year-old woman with iridocorneal endothelial syndrome. **A,** Normal appearing right eye. **B,** Left eye, pupil drawn towards area of anterior synechia and ectropion uvea at 8 o'clock with an iris atrophy at 3 o'clock.

(See color figure at http://www.turkiyeklinikleri.com/journal/oftalmoloji-ozel-dergisi/1308-111X/)

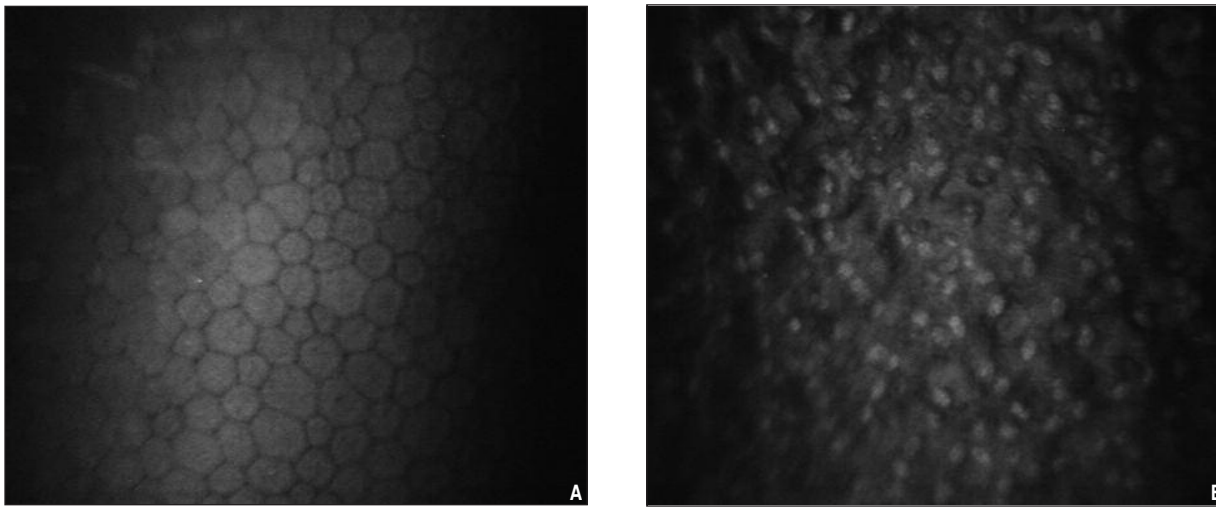

FIGURE 2A,B: In-vivo confocal microscopy. **A,** Normal corneal endothelium of the right eye. **B,** Left eye, epithelium-like transformation of the corneal endothelium with irregularly shaped cells and hyperreflective nuclei.

corneal densitometry and biomechanical properties were found to be affected.

Pentacam® HR is a noninvasive optical system and has been designed to assess the anterior segment of the eye to provide corneal pachymetry, corneal topography, anterior chamber analysis and densitometry of the cornea and lens. Densitometry of the corneal tissue provides information about the clarity of the cornea.[6] The densitometry output is expressed in GSU and ranged from 0 (no clouding, maximum transparency) to 100 (completely opaque cornea, no transparency). Mean corneal densitometry in healthy adults has been recently reported as 12.3±2.4 and 19.7±3.9 GSU in two different studies.[6,7] The right and left corneal densitometry values were also found to be highly correlated in a previous study.[7] However, the corneal densitometry of our patient was 18.6 GSU in the affected eye and 15.7 GSU in the fellow eye.

For densitometry analysis, the 12-mm diameter area is subdivided into four concentric radial zones (First zone is 2 mm in diameter and centered on the apex; Second zone is an annulus extending from 2 to a 6-mm-diameter circle; Third zone annulus extends from 6 to 10 mm; Fourth zone extending from 10 to a 12- mm-diameter circle). The output can also be subdivided based on corneal depth into anterior, central, and posterior layers.

TABLE 1: Corneal biomechanical properties obtained with ocular response analyzer.

Corneal Hysteresis (mmHg)	10.9	8.7
Corneal Resistance Factor (mmHg)	10.1	8.6
IOPcc (mmHg)	13.3	16.6
IOPg (mmHg)	13.0	14.1

OD: Right Eye; OS: Left Eye; IOPcc: Cornea compensated intraocular pressure; IOPg: Goldmann correlated intraocular pressure.

TABLE 2: Cornea densitometry (annulus and layer averages) of the patient.

	0-2 mm	2-6 mm	6-10 mm	10-12 mm	Total
Anterior (120 µm)					
OD	21.0	19.0	20.2	28.7	21.4
OS	25.6	23.6	24.7	33.2	25.9
Center					
OD	13.8	12.4	14.4	17.9	14.2
OS	14.3	13.3	15.5	19.8	15.3
Posterior (60 µm)					
OD	10.6	9.9	12.3	14.2	11.5
OS	13.2	12.1	16.5	16.3	14.5
Total					
OD	15.1	13.8	15.6	20.3	15.7
OS	17.7	16.4	18.9	23.1	18.6

OD: Right Eye; OS: Left Eye.

Anterior layer corresponds to the anterior 120 µm, and the posterior layer to the most posterior 60 µm of the cornea. The corneal densitometry in all con-

centric radial zones and anteroposterior layers were higher in the affected eye of our patient when compared to fellow eye. Using corneal densitometry for ICE syndrome can provide an objective and reproducible method of measuring corneal haze, therefore will provide a more reliable monitoring of disease progression.

ORA provides valuable information about biomechanical properties of the cornea. Of these, corneal hysteresis and resistance factor characterize the viscoelastic properties of the cornea, especially those of the ground substance.[8] In our case, CH of the right and left eyes were 10.9 and 8.7 mmHg and CRF were 10.1 and 8.6 mmHg, respectively; means that corneal rigidity is lower in the affected eye. This decrease may be similar to the principle in postoperative corneal edema, bullous keratopathy and Fuch's corneal dystrophy in which corneal thickness increase and CH-CRF decreases.[8] The ORA values of CH and CRF measures viscosity of the corneal ground substance, which, in turn provides an information related to corneal biomechanical state. Using ORA for ICE syndrome can provide an objective and reproducible method of measuring corneal biomechanics which will be possibly affected from disease progression. Also we should be careful about altered corneal biomechanics in ICE syndrome that may lead to inaccurate IOP measurements.

In conclusion, in-vivo confocal microscopy is a crucial tool for the accurate diagnosis of ICE syndrome. However other non-invasive imaging modalities to assess corneal biomechanics and corneal densitometry can facilitate the diagnosis of this rare, complex and variable spectrum of disorder. Corneal densitometry can be used as an objective measure of corneal clarity in order to monitor disease progression. Further studies are necessary to establish relationship of corneal densitometry and biomechanical properties with ICE syndrome.

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