Letter to the Editor

A Rare Corneal Scenario: Concurrent Diagnosis of Epithelial Basement Membrane Dystrophy and Crocodile Shagreen

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Dear Editor,

Epithelial basement membrane dystrophy (EBMD), also known as "map-dot-fingerprint dystrophy", is the most common form of corneal dystrophy and is frequently encountered in clinical practice. Although asymptomatic in many patients, recurrent epithelial erosions occur in approximately 10% of cases.[1] On clinical examination, it is characterized by subepithelial map-like patterns, fingerprint-like lines, and microcystic opacities on the epithelial surface. It usually appears between the ages of 20 and 40 years.[1]

Corneal degenerations are common clinical manifestations in ophthalmology and can be classified as age-related changes, deposits, and marginal degenerations.[2] Crocodile shagreen is an asymptomatic corneal degeneration characterized by bilateral grayish-white, polygonal corneal stromal opacities reminiscent of crocodile skin, seen especially in older patients.[2,3,4] There are two subtypes: anterior and posterior. In the anterior form (shagreen of Vogt), mosaic patterns and calcium deposits are seen at the level of Bowman's layer, while the posterior form crocodile shagreen is characterized by a "sawtooth" configuration of the stromal collagen lamellae.[2,4] In most cases, it does not affect visual acuity, is detected incidentally, and requires no treatment.[2,3]

In this case report, we discuss the clinical, anterior segment optical coherence tomography (AS-OCT), and *in vivo* confocal microscopy (IVCM) findings of a patient who presented with low vision in his left eye and was diagnosed with EBMD and crocodile shagreen, a previously unreported co-occurrence.

A 52-year-old man presented with a 6-year history of declining vision in the left eye. He had a history of phacoemulsification and intraocular lens (IOL) implantation surgery in the right eye 13 years earlier. Best corrected visual acuity was 0.8 Snellen decimal in the right eye and hand movements in the left eye. On biomicroscopic examination, both corneas were hazy and exhibited map-like structures consistent with EBMD in the epithelium and mosaic opacities characteristic of crocodile shagreen in the stroma (Figure 1A, B). The right eye was pseudophakic and the left eye had posterior polar cataract. On fundus examination, the optic disc and visible retinal areas in the right eye appeared normal, while the retina of the left eye could not be examined due to lens opacity. The retina appeared attached on ultrasound examination. Intraocular pressures were measured as 15 mmHg and 13 mmHg in the right and left eye, respectively. The patient had no systemic disease, regular medication use, or family history of similar ocular disease.

On AS-OCT (DRI OCT Triton, Topcon, Tokyo, Japan), central corneal thickness was 505 µm on the right and 511 µm on the left. Basement membrane irregularities,

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hyperreflective epithelial incursions, and increased reflectivity in the anterior stroma were observed. Epithelial thickness was measured as 71 µm on the right and 73 µm on the left (Figure 2A, B). IVCM (Heidelberg Retina Tomograph II–Rostock Cornea Module, Heidelberg Engineering GmbH, Heidelberg, Germany) imaging revealed hyperreflective lines in the epithelium, an irregular, thickened basement membrane with sporadic lace-like appearance, and hyperreflective spots (Figure 3A, B). In addition, there were numerous dendritic cells situated around the basement membrane and penetrating into the epithelium (Figure 3B). A mosaic pattern of hyperreflective polygonal areas 50-200 µm in diameter, separated by black streaks, was observed in the stroma (Figure 3C). There were scattered guttae and a single hyperreflective opacity in the endothelium (Figure 3D). Endothelial cell counts were 2014 and 2107 cells/mm^2 in the right and left eyes, respectively. Phacoemulsification and posterior chamber IOL implantation were performed in the left eye. The surgery was uncomplicated, and visual acuity increased to 0.8 postoperatively.

IVCM enables histological examination of the cornea and is widely used in the diagnosis of corneal infections,

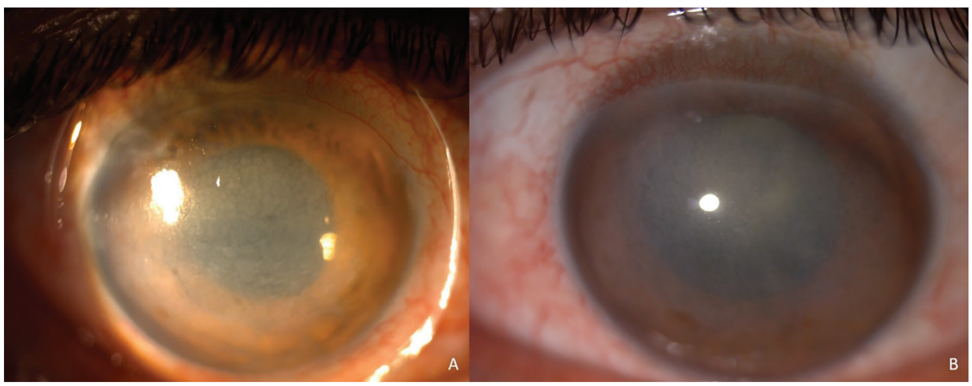

Figure 1. Anterior segment images of a patient with epithelial basement membrane dystrophy and crocodile shagreen. A) A therapeutic contact lens was applied to the right eye because of corneal haze and recurrent epithelial erosions; B) Corneal haze and the characteristic polygonal pattern of crocodile shagreen are seen in the left eye (×16 magnification)

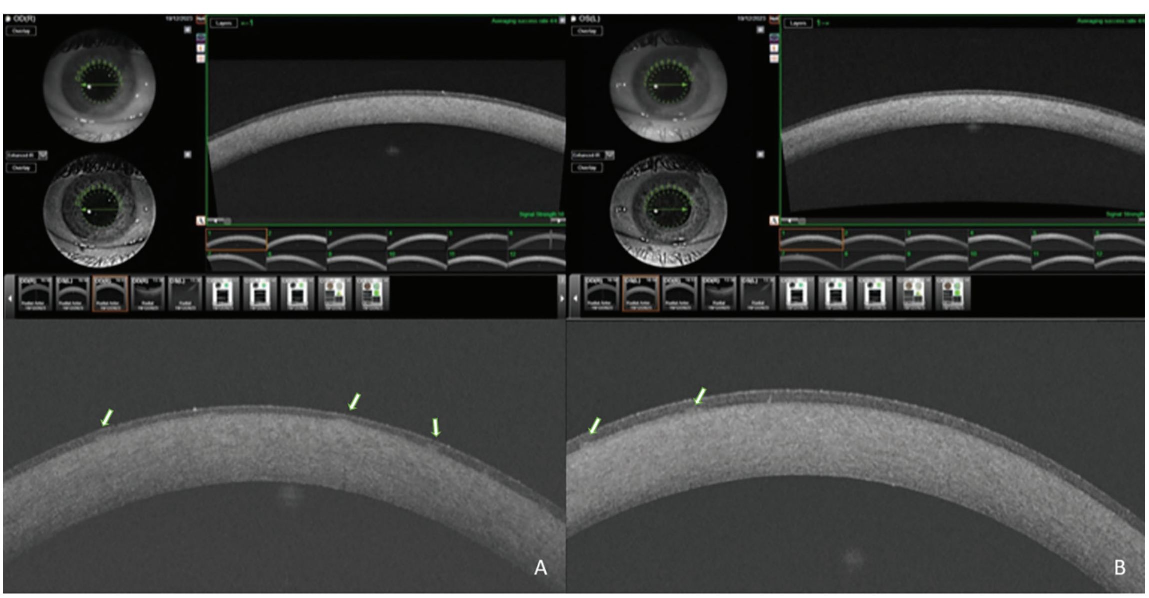

Figure 2. Anterior segment optical coherence tomography images (AS-OCT, DRI OCT Triton, Topcon, Tokyo, Japan). The right (A) and left (B) eyes show irregularities in the basement membrane, hyperreflective protrusions into the epithelium (arrows), and increased reflectivity in the anterior stroma

degeneration, and dystrophies.[5] Consistent with the literature, IVCM imaging in our patient demonstrated an irregular basement membrane with a thickened, lace-like appearance penetrating into the epithelium, round hyperreflective deposits in the epithelium, and irregular epithelial cells and structural defects in these regions, which are characteristic findings of EBMD.[6,7] In a study including 3 patients with crocodile shagreen and polymorphic amyloid degeneration, Woodward et al.[8] observed areas of acellular, hyperreflective opacity separated by linear black areas in the stroma on IVCM imaging in a patient with isolated posterior crocodile shagreen. In the patient with combined polymorphic amyloid degeneration and posterior crocodile shagreen, hyperreflective punctate lesions were observed in the stroma in addition to these findings. Similarly, our patient exhibited a mosaic pattern in the stroma consisting of hyperreflective polygonal areas with a diameter of 50-200 µm, separated by black streaks.

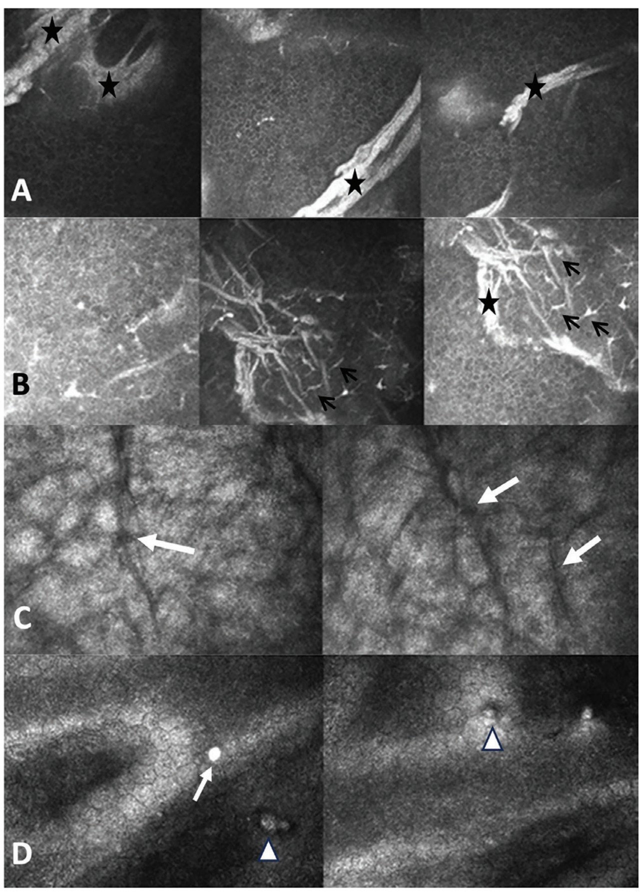

Figure 3. *In vivo* confocal microscopy images (IVCM, Heidelberg Retinal Tomograph 3/Rostock Cornea Module, Heidelberg Engineering). A, B) Hyperreflective lines in the epithelium, a sporadically lace-like, irregular, thickened basement membrane penetrating into the epithelium (black stars), and hyperreflective dots. B) Numerous dendritic cells (black arrows) around the thickened basement membrane. C) A mosaic pattern of hyperreflective polygonal areas 50-200 µm in diameter separated by black streaks (white arrows) in the stroma. D) Scattered guttae (white triangles) and a hyperreflective opacity (white arrow) in the endothelium

We found no AS-OCT study on crocodile shagreen in the literature. In this case report, images obtained with swept-source AS-OCT reveal irregularities in the epithelial basement membrane at high magnification, consistent with EBMD, with protrusion of the basement membrane into the epithelium. Although not very pronounced, there was increased reflectivity in the anterior stroma.

Belliveau et al.[4] used electron microscopy to examine corneal specimens from 3 patients with crocodile shagreen who underwent keratoplasty and demonstrated that the stromal collagen lamellae lost their parallel arrangement assuming a sawtooth-like irregular, wavy appearance. They also detected vacuoles in the stroma, particularly in the region adjacent to the banded anterior part of Descemet's membrane. The fibrillogranular electron-dense material in these vacuoles is thought to be caused by degenerative collagen products and mucopolysaccharide accumulation in the stroma. As vacuoles detected by electron microscopy are probably very small in diameter, they could not be detected on IVCM imaging in our study. Krachmer et al.[9] examined the cornea of a 75-year-old patient with posterior crocodile shagreen and polymorphic amyloid degeneration by postmortem transmission electron microscopy and showed the presence of sawtooth-like stromal collagen lamellae corresponding to central fuzzy opacities seen on clinical examination. They also confirmed the amyloid nature of the punctate and filamentous hyperreflective structures seen on clinical examination by histochemical staining and electron microscopy.

This case makes an important contribution to the literature as the first detailed documentation of EBMD and crocodile shagreen co-occurrence using both IVCM and AS-OCT. EBMD and crocodile shagreen are two separate clinical entities with pathophysiologically distinct etiologies and affect the epithelial and stromal layers of the cornea, respectively. While EBMD mostly causes structural disorders in the epithelial basement membrane, crocodile shagreen is characterized by degenerative changes in stromal collagen organization. These differences suggest that their coexistence may be a coincidental association.

Ethics

Informed Consent: Written informed consent was obtained from the patient to publish their clinical data and images for scientific purposes.

Declarations

Authorship Contributions

Surgical and Medical Practices: B.B., Ş.G., Concept: S.Ü., B.B., Ş.G., A.B.O., Design: S.Ü., B.B., Ş.G., A.B.O., Data Collection or Processing: S.Ü., B.B., Ş.G., A.B.O., Analysis or Interpretation: B.B., Ş.G., A.B.O., Literature Search: S.Ü., B.B., Ş.G., A.B.O., Writing: S.Ü., B.B., Ş.G., A.B.O.

Conflict of Interest: Banu Bozkurt, MD, is an Editor-in-Chief of the Turkish Journal of Ophthalmology. She was not involved in the peer review of this article and had no access to information regarding its peer review. The other authors has no disclosures.

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