Clinical Case Report

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# A CARE-compliant article: optical coherence tomography for epithelial basement membrane dystrophy

## A case report

Yi-Chun Kuo, MD[a], Yuan-Chieh Lee, MD, PhD[a,b,c,*]

**Abstract**

**Rationale:** The etiology of anterior corneal opacities and the effect of debridement cannot be determined by biomicroscopy. Optical coherence tomography (OCT) helps identify the character and depth of these lesions.

**Patient concerns:** A 45-year-old female complained of progressive blurred vision for a long time. Slit lamp biomicroscopy showed irregular, faint scar-like opacity of anterior cornea in her both eyes. Pentacam Scheimpflug camera tomography showed irregular astigmatism of anterior corneal surface. Anterior segment spectral-domain OCT revealed thickened, hyper-reflective linings, and scattered lesions, mainly in the epithelial layer.

**Diagnoses:** Epithelial basement membrane dystrophy (EBMD).

**Intervention:** Epithelial debridement and bandage lenses.

**Outcomes:** The cornea became clear and the vision improved soon after debridement. The pathology showed thickened aberrant basement membrane extending into mid-epithelial layer, with microcyst-like lesions also noted.

**Lessons:** OCT defines the depth of lesions and helps diagnosis and management of EBMD.

**Abbreviations:** EBMD = epithelial basement membrane dystrophy, OCT = optical coherence tomography.

**Keywords:** epithelial basement membrane dystrophy, epithelial debridement, map-dot-fingerprint dystrophy, spectral domain optical coherence tomography

### 1. Introduction

Epithelial basement membrane dystrophy (EBMD) affects at least 2% of the general population,[1] and is also known as anterior basement membrane dystrophy, map-dot-fingerprint dystrophy or Cogan dystrophy. It may present with minute pinpoint spots to irregular or oval shaped grayish to milk white opacities,[2] map-like changes,[3] fingerprint parallel lines,[4] and Bron rare bleb changes.[5] On facing corneal opacities, 1 important issue is the depth of these opacities, which might determine whether we can remove these opacities completely without disturbing the smooth corneal surface. Here, we report a case of EBMD in whom the depth of opacities was demonstrated by spectral domain optical coherence tomography (OCT). Written informed consent for the publication of this case and any additional related information was taken from the patient involved in the study.

### 2. Case report

A 45-year-old female with major depression disorder complained of progressive blurred vision for a long time. The examination revealed a spectacle-corrected visual acuity of 20/200 in the right eye and 20/400 in the left eye. Slit lamp examination revealed multiple irregular, scar-like opacities of bilateral anterior cornea (Fig. 1A). The anterior chambers were deep and clear, and the lenses were also clear. Ophthalmoscopic examination was normal. Pentacam showed irregular astigmatism of anterior corneal surface. Because the depth of the corneal opacity could not be ascertained by biomicroscope, spectral domain OCT examination of cornea was performed. The OCT demonstrated hyperreflective linings between epithelial layer and stroma, scattered hyperreflective lesions of variable size located in the superficial layer, middle layer, deep layer, or full thickness of the epithelium. All these lesions were confined to the epithelial layer (Fig. 1B). Under tentative diagnosis of corneal epithelial dystrophy, the patient received corneal epithelial debridement and bandage soft contact lens application. The cornea became clear and her vision improved soon after debridement. The pathological examination showed thickened aberrant basement membrane extending into mid-epithelial layer and intraepithelial microcyst, which were compatible with the diagnosis of EBMD

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*[a] Department of Ophthalmology, Buddhist Tzu Chi General Hospital, [b] Department of Ophthalmology and Visual Science, [c] Institute of Medical Science, Tzu Chi University, Hualien, Taiwan.*

*[*] Correspondence: Yuan-Chieh Lee, Department of Ophthalmology, Buddhist Tzu Chi General Hospital, No. 707, Sector 3, Chung-Yang Road, Hualien, Taiwan 970, R.O.C. (e-mail: yuanchieh.lee@gmail.com).*



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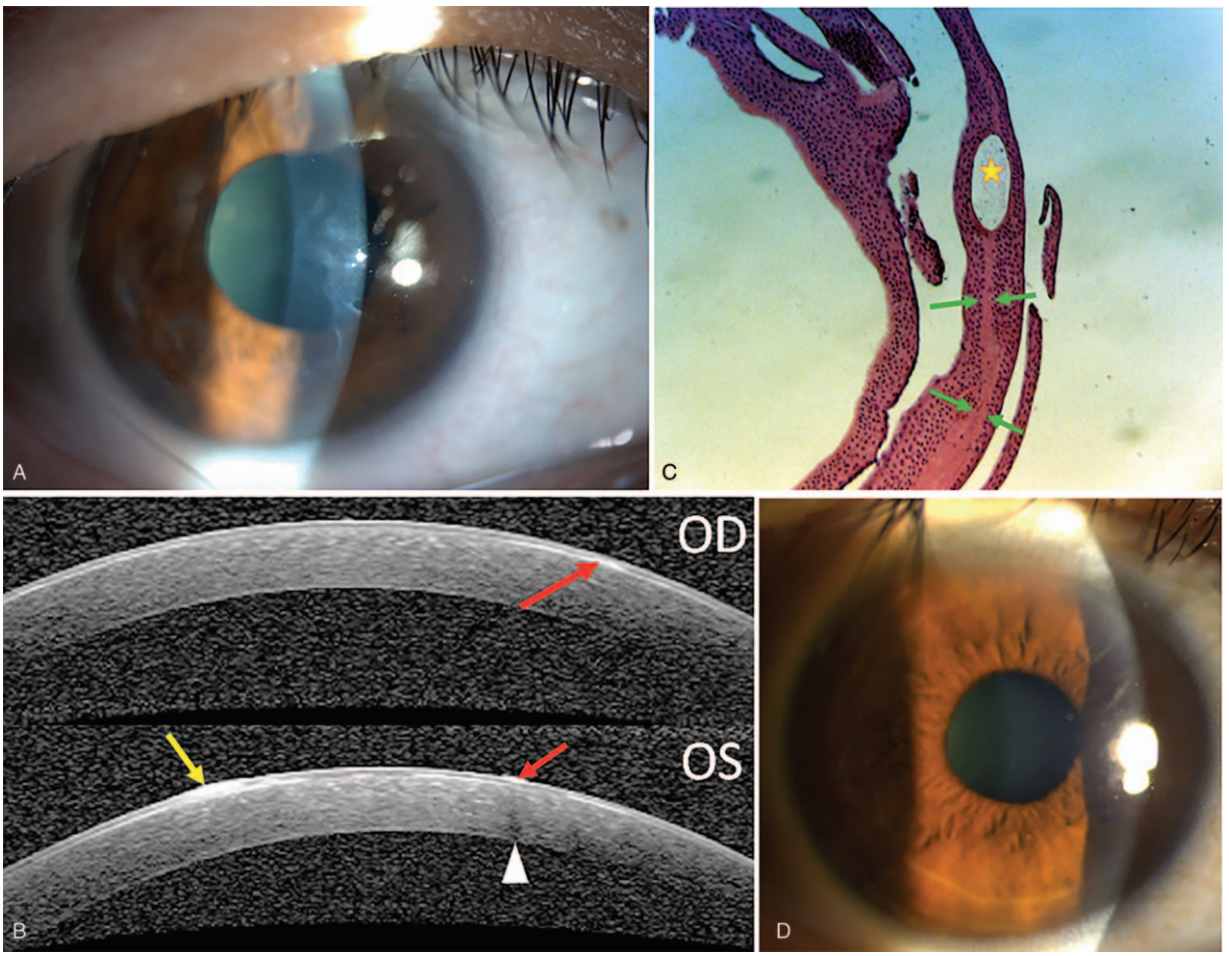


**Figure 1.** A, Multiple irregular, scar-like opacities of anterior cornea were found in her both eyes (only right eye is shown here). B, The SD-OCT demonstrated scattered thickened hyperreflective lesions (red arrows), which were confined to the epithelial layer. Part of left eye epithelial basement membrane became very thick and formed small hyperreflective elevations associated with undulation and elevation of the corneal epithelial layer (yellow arrow). The arrow head pointed the posterior shadow cone. C, The pathological examination showed thickened aberrant basement membrane extending into mid-epithelial layer (between green arrows), with microcyst-like lesions (yellow asterisk) also noted. D, The cornea became clear after epithelial debridement. There were no recurrent opacities in the following 2 years (picture taken 2 years after epithelial debridement). OCT = optical coherence tomography.

(Fig. 1C). There was no recurrence under biomicroscopic examination and OCT imaging in the following 2 years (Fig. 1D).

## 3. Discussion

The basal epithelial cells contain keratin intermediate filaments and secrete a continuous basement membrane, which is about 50-nm thick and composed mainly of type IV collagen, laminin, and other proteins.[3,6–8] EBMD is characterized with an abnormal basement membrane protruding into the epithelium and the presence of intraepithelial microcysts.[7] These anteriorly extending basement membranes hinder deeper epithelial cells from moving forward. The desquamating cells are therefore entrapped beneath the sheet of aberrant basement membrane, forming variable-sized cysts of cellular and nuclear debris. Map-like configurations and fingerprint lines composed of irregular or parallel rows of thickened basement membrane.[9]

In our patient, the corneal opacities did not appear as typical dots, map, or fingerprint. On facing these amorphous putty opacities, an important issue that might affect our treatment decision making is the depth of these opacities. If these opacities involved Bowman layer or deeper, debridement might not be complete or lead to uneven surface and irregular astigmatism. OCT provided a good evaluation of depth of lesions and suggested possible diagnosis preoperatively. The pathological examination of the debrided epithelium confirmed the diagnosis of EBMD.

The ultrastructure of EBMD has been recently demonstrated with confocal microscopy and OCT.[10,11] Under confocal microscopy, the map-like lesion of the cornea presents a shape of high-reflective extracellular deposits, while the fingerprint-like lesion appears as multiple dark striae.[10] The 2 main features on OCT scans are an irregular and thickened membrane duplicating or insinuating into the corneal epithelium layer and the characteristic hyperreflective dots.[11] These hyperreflective dots vary in number and size and located superficially, with or without posterior shadow cone.[11] The resolution of SD-OCT is not as good as confocal microscopy, but SD-OCT provides a better localization and depth of the lesions. Both the confocal microscopy and SD-OCT are beneficial in the diagnosis of EBMD and accurate assessment of the structural changes, and a good correlation between these 2 instruments has been demonstrated.[11]

Apart from diagnosis, the SD-OCT demonstrates the confinement of the lesions and predicts the effectiveness of corneal epithelial debridement.[11–13] Relatively quick and easy, OCT can not only be used for diagnosis and evaluation, but also for follow-up. The recent en face OCT allows fast analysis of large corneal areas,[14] which makes OCT a better tool for early detection of recurrence in the follow-up. Although our report is limited to EMBD, we believe that the advanced OCT will gain an expanding role in the category of corneal dystrophy, including EMBD.

## Author contributions

**Conceptualization:** Yuan-Chieh Lee.
**Data curation:** Yi-Chun Kuo, Yuan-Chieh Lee.
**Formal analysis:** Yi-Chun Kuo, Yuan-Chieh Lee.
**Investigation:** Yuan-Chieh Lee.
**Supervision:** Yuan-Chieh Lee.
**Visualization:** Yi-Chun Kuo.
**Writing - Original Draft:** Yi-Chun Kuo.
**Writing - Review & Editing:** Yuan-Chieh Lee.
Yuan-Chieh Lee orcid: 0000-0002-8486-8549.

## References

[1] Werblin TP, Hirst LW, Stark WJ, et al. Prevalence of map-dot-fingerprint changes in the cornea. Br J Ophthalmol 1981;65:401–9.
[2] Cogan DG, Donaldson DD, Kuwabara T, et al. Microcystic dystrophy of the corneal epithelium. Trans Am Ophthalmol Soc 1964;62:213–25.
[3] Guerry D 3rd. Observations on Cogan's microcystic dystrophy of the corneal epithelium. Trans Am Ophthalmol Soc 1965;63:320–34.
[4] Devoe AG. Certain abnormalities of Bowman's membrane with particular reference to fingerprint lines in the cornea. Trans Am Ophthalmol Soc 1962;60:195–201.
[5] Bron AJ, Brown NA. Some superficial corneal disorders. Trans Ophthalmol Soc U K 1971;91:Xii+.
[6] Qu JH, Sun XG. [Research progress of corneal epithelial basal cells and basement membrane]. Zhonghua Yan Ke Za Zhi 2016;52:703–7.
[7] Cogan DG, Kuwabara T, Donaldson DD, et al. Microcystic dystrophy of the cornea. A partial explanation for its pathogenesis. Arch Ophthalmol 1974;92:470–4.
[8] Rodrigues MM, Fine BS, Laibson PR, et al. Disorders of the corneal epithelium. A clinicopathologic study of dot, geographic, and fingerprint patterns. Arch Ophthalmol 1974;92:475–82.
[9] Ehlers N, Moller HU. Pathology and pathomechanisms of epithelial microcystic and basement membrane abnormalities of the cornea. Acta Ophthalmol (Copenh) 1988;66:318–26.
[10] Kobayashi A, Yokogawa H, Sugiyama K. In vivo laser confocal microscopy findings in patients with map-dot-fingerprint (epithelial basement membrane) dystrophy. Clin Ophthalmol 2012;6:1187–90.
[11] El Sanharawi M, Sandali O, Basli E, et al. Fourier-domain optical coherence tomography imaging in corneal epithelial basement membrane dystrophy: a structural analysis. Am J Ophthalmol 2015;159:755–63.
[12] Pole C, Sise A, Joag M, et al. High-resolution optical coherence tomography findings of lisch epithelial corneal dystrophy. Cornea 2016;35:392–4.
[13] Elhardt C, Priglinger SG, Karakolova Y, et al. [Corneal dystrophies in optical coherence tomography]. Ophthalmologe 2018;[Epub ahead of print].
[14] Ghouali W, Tahiri Joutei Hassani R, Liang H, et al. [Imaging of corneal dystrophies: correlations between en face anterior segment OCT and in vivo confocal microscopy]. J Fr Ophtalmol 2015;38:388–94.