

Corneal Epithelial Basement Membrane Dystrophy

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Abstract

A 38 year old female presented with recurrent redness, pain, watering in both eyes since last 6-7 yrs having taken antibiotics, steroids, lubricating drops, anti-viral (acicvir), hypersol, (bandage contact lenses) BCL for treatment with no positive family history. Her best corrected visual acuity were 6/9 in both eyes for distance and N/6 for near with right eye showing lid oedema and conjunctival congestion. Corneal sensations and Schirmer's test were normal.

Introduction

Corneal map-dot-fingerprint dystrophy is by far the most common corneal dystrophy and is named from the appearance of its characteristic slit lamp findings. Map-dot-fingerprint dystrophy also is known as epithelial basement membrane dystrophy, and Cogan microcystic epithelial dystrophy. It usually is classified as a dystrophy but fits more accurately into the corneal degeneration category.

Corneal dystrophies usually are hereditary, bilateral, progressive, and not associated with systemic or local disease.¹ Map-dot-fingerprint dystrophy has been found in several families with a presumed autosomal dominant pattern, but in most cases, it is not familial. It is not progressive but rather variable and fluctuating in its course. Usually, it is bilateral but can be unilateral or very asymmetric in presentation.²

Case Report

A 38 year old female presented with recurrent redness, pain, watering in both eyes since last 6-7 yrs having taken antibiotics, steroids, lubricating drops, anti-viral (acicvir), hypersol, BCL for treatment with no positive family history. Her best corrected visual

acuity were 6/9 in both eyes for distance and N/6 for near with right eye showing lid oedema and conjunctival congestion. Corneal sensations and Schirmer's test were normal.

On examination, slit lamp findings suggested of an oval corneal epithelial defect measuring 3 mm x 1 mm, with positive fluorescein staining and inferior limbal vascularisation in the right eye and left eye showing sub-epithelial map and dot pattern opacities.

Patient was treated locally with mild steroid-fluromethanole and lubricating eye drops. Systemic Doxycycline 100 mg BD was prescribed for 6 weeks.

Pathophysiology

Corneal abnormalities associated with map-dot-fingerprint dystrophy are the result of a faulty basement membrane, which is thickened, multilaminar, and misdirected into the epithelium. Maps histologically represent areas of multilaminar basement membrane, which extend into the epithelium. Dots are intraepithelial microcysts that contain nuclear, cytoplasmic, and lipid debris. Fingerprints are curvilinear clusters of reduplicated and thickened basement membrane and fibrillogranular material. Blebs, a less common manifestation are localized areas of fibrillogranular material or thickened basement membrane.³

Frequency. In the US prevalence of map-dot-fingerprint dystrophy range from 2-43% of the general population and is found to be

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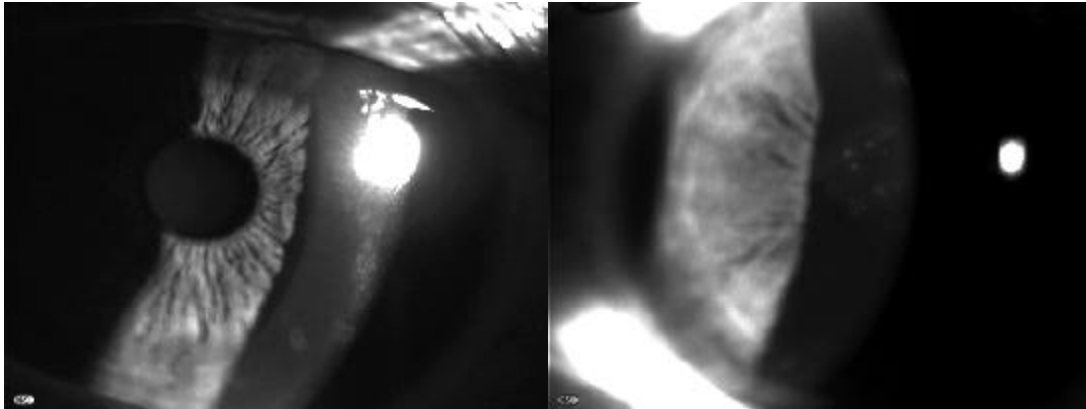


Fig. 1 : Oblique illumination showing corneal subepithelial map and dot dystrophy.

more common in females and is in all age group except children.

Mortality/Morbidity: Patients may be asymptomatic, experience painful recurrent erosions, decreased vision, or both.

Clinical Relevance

Refraction is uncertain due to irregular astigmatism.

Slit lamp findings includes the following:

Corneal maps seen as irregular geographic shape, faint gray-white patches that may contain clear oval areas. They vary greatly in size (usually 1 mm to several mm) and are seen best with broad oblique illumination.

Corneal dots seen as gray-white, puttylike opacities, which can be round, comma-shaped, or irregular. They usually are 0.05-1.0 mm in size.

Corneal fingerprints seen as clusters of contoured concentric lines 0.25-4.0 mm long.

They are seen best with retroillumination.

Corneal blebs are clear, round, bubble like defects 0.05-0.2 mm in diameter. They are seen best with retroillumination.

Keratometry shows irregular astigmatism.

A placido disk or keratometer often demonstrates irregularity better than

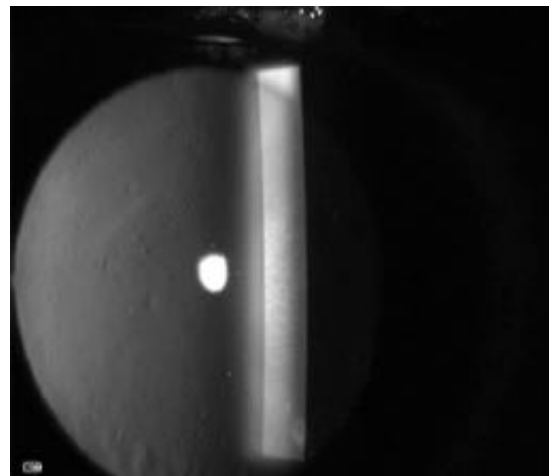


Fig. 2 : Retro illumination view of basement membrane dystrophy.

computed tomography.

Conditions of elevated intraocular pressure and corneal decomposition which gives rise to corneal epithelial oedema mimicking corneal pseudo fingerprints or shift lines can cause diagnostic dilemma.

Treatment

Medical Care

Hypertonic drops or ointment help both irregular astigmatism and recurrent corneal erosion problems. Sodium chloride (5%) drop four times a day and ointment at bedtime is

recommended.

Nonhypertonic lubricating drops or ointment is also preferred as it is found that there is no difference between hypertonic and nonhypertonic ointment.

Capsule Doxycycline (100 mg) BD for six weeks helps in the adhesion between cells of epithelial basement membrane, mechanism of which is not known.

Patching is done in case of acute episodes of corneal erosions.

Bandage extended wear soft contact lens is useful but risk of infectious keratitis makes this a secondary choice.

Hard or gas-permeable contact lens is used to improve vision by masking corneal irregular astigmatism but is poorly tolerated because of increased corneal fragility/erosion problems.

Surgical Care

Debridement/superficial keratectomy is done in case of significant visual loss from associated irregular astigmatism and recurrent corneal erosions.

Diamond burr superficial keratectomy: After epithelial debridement a diamond-dusted burr is used to polish the basement membrane.

Excimer laser phototherapeutic keratectomy is an excellent treatment for recurrent corneal erosions associated with map-dot-fingerprint dystrophy.⁴

Corneal anterior stromal needle puncture: This procedure is not as successful for recurrent erosions associated with map-dot-fingerprint dystrophy, which is usually more diffuse and often migratory.

Prevention

Lubricating hypertonic saline or bland ointment at bedtime is most of the time

helpful to prevent recurrent erosions.

Complications

Recurrent erosions predispose the cornea to infection.

Prognosis

Map-dot-fingerprint dystrophy findings fluctuate but tend not to progress over time. Majority of patients are able to maintain sufficient vision and comfort for reading, driving, and other visual tasks, except during episodes of corneal erosions.

Special Concerns

Patient with this dystrophy may be bothered by painful recurrent erosion episodes and or decreased vision but are most frustrated by the unpredictability of the condition.

Map-dot -fingerprint dystrophy is a relative contraindication for refractive procedures, such as LASIK or LASEK. Trauma from the microkeratome sliding over the epithelial surface or from flap manipulation is more likely to occur in patients with map-dot-fingerprint dystrophy because of the poorly adherent epithelium. Epithelial sloughing can lead to epithelial ingrowth and stromal melts. Surface ablation [PRK]) may be a better refractive procedure option for these patients.

References

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