

Case Study

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**Case Number Two:
Diagnosis, referral, co-management and follow-up of a patient with map-dot-fingerprint
corneal dystrophy.**

by
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Abstract:

Epithelial Basement Membrane Dystrophy (EBMD) is the most common anterior corneal dystrophy. It is usually bilateral, asymptomatic and only remarked by the patient if a corneal erosion occurs as a result of the dystrophy. EBMD is not familial and not progressive. This report describes the case of a patient with unilateral EBMD. The diagnosis, referral to an Ophthalmologist and treatment of the dystrophy.

Key words: *Cogan microcystic dystrophy, map-dot-fingerprint dystrophy, epithelial basement membrane dystrophy,*

Introduction

Epithelial basement membrane dystrophy (EBMD) is also known as Cogan microcystic dystrophy or map-dot-fingerprint dystrophy. Corneal dystrophies in general have in common that they are genetically determined, progressive, bilateral and develop in the absence of inflammation. EBMD is an exception to the rule since it is neither familial nor progressive.¹

EBMD has a slight female predilection and can manifest at any age, usually before the fourth decade. Intra- and subepithelial basement membrane reduplication causes abnormal epithelial adhesion. As a result there can be seen three different types of lesions: Dots, Cysts, Fingerprints and Maps. All four lesions can best be seen in retro-illumination with the slitlamp. According to Kanski¹ with the passage of time one pattern can frequently change to another. Also the distribution of the lesions can vary.

Often EBMD stays undetected and is only diagnosed because of the patient presenting with recurrent corneal erosions. About 10% of patients with EBMD develop recurrent corneal erosions, whereas 50% of patients with recurrent corneal erosions have EBMD.² Usually recurrent corneal erosions occur after the age of 30. Mostly the patient complains of pain, photophobia and blurred vision after awakening in the morning.³ It is thought that the corneal epithelium adheres to the palpebral conjunctiva while sleeping and is detached from the basement membrane upon opening the eyes in the morning.⁴

Case Report

At 13. April 2000 M.Z. a 36 year old white female (office worker) presented to our contactlens-practice for contactlens fitting. She had no other complaints than blurred vision from her myopia.

She had a 10 year history of occasional wear of glasses. She had not been evaluated by an Ophthalmologist so far. She reported an allergy to penicillin and denied any systemic disease and had no family history of any eye- or systemic disease. She was oriented to time, place and person.

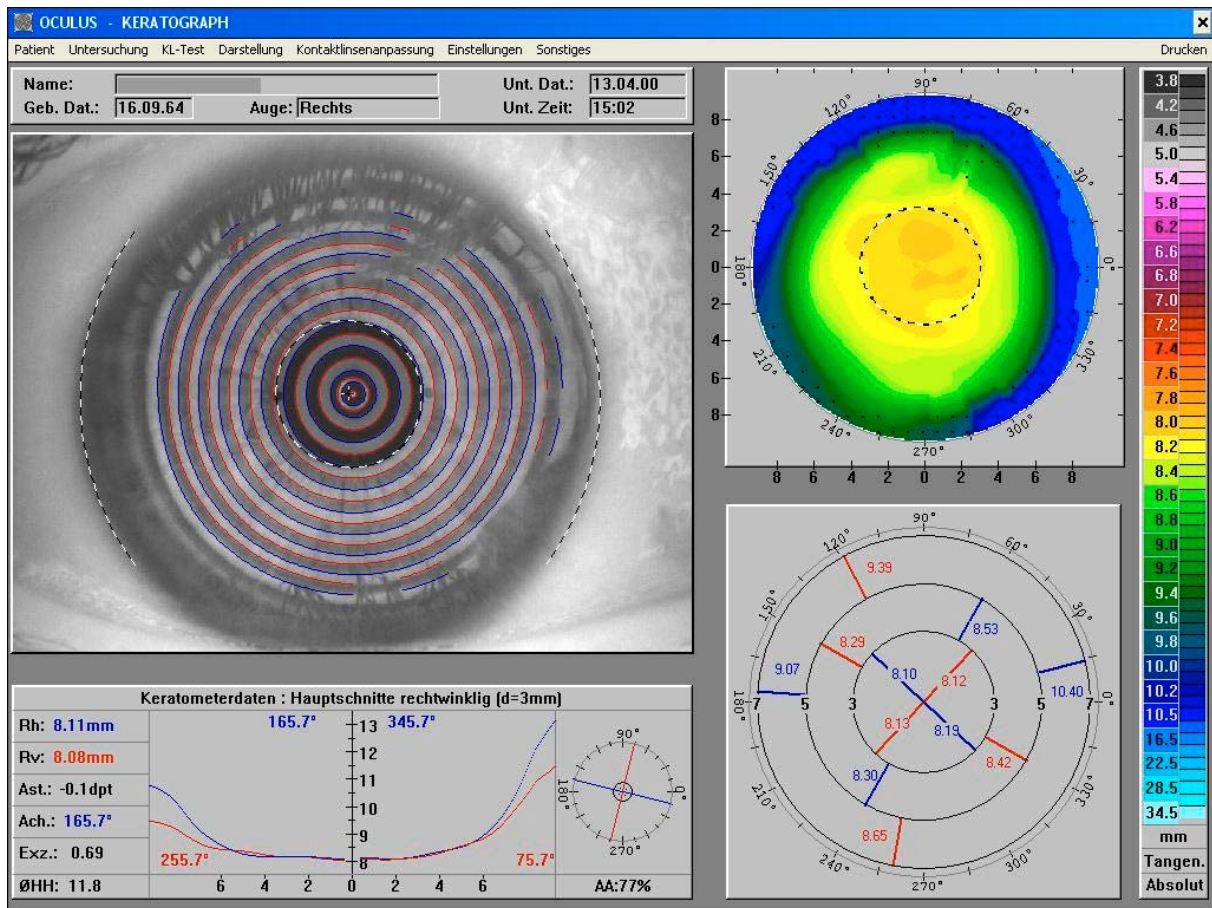
Her uncorrected visual acuity was 20/30 OU. Uncorrected near acuities were .3M OU. Best-corrected visual acuity with glasses was 20/13 OU. The refraction was -0.75 OD and -0.75 OS at a corneal apex to lens distance of 14mm OU. Pinhole acuity at distance was 20/20 OD and 20/20 OS. Color vision testing with pseudoisochromatic plates showed no color vision deficiency OU. Pupils were equally round and reactive to light, no afferent pupil defect was noted OU. Confrontation fields were full to finger count OU. Extraocular muscles were unrestricted in all gazes, and cover test demonstrated orthophoria at distance and near. Due to the law of practice in Switzerland no Goldman Tonometry could be performed. Anterior segment evaluation by slit lamp examination revealed a quiet bulbar and palpebral conjunctiva OU. An even tear film with tear break up time of 15 seconds OU. Clear lashes OU. The corneas were clear and no staining was noted anywhere. Irides were brown OU. The anterior chamber appeared clear without cells or flare and the anterior chamber angles were estimated by the Van Herrick method with the slit-lamp as 4 nasally and 4 temporally OU. Both lenses were evaluated by slit-lamp with undilated pupils and have been noted as clear with no opacities in any region

The evaluation of the posterior segment by slit lamp with 90D lens and undilated pupils revealed clear optic disc margins OU with a cup-to-disc ratio of .3/.3 OU. The neuroretinal rims were healthy and intact. The retinal vessels showed an arterial-venous ratio of 2/3 OU. Both eyes presented with clearly defined macular margins and foveal reflex.

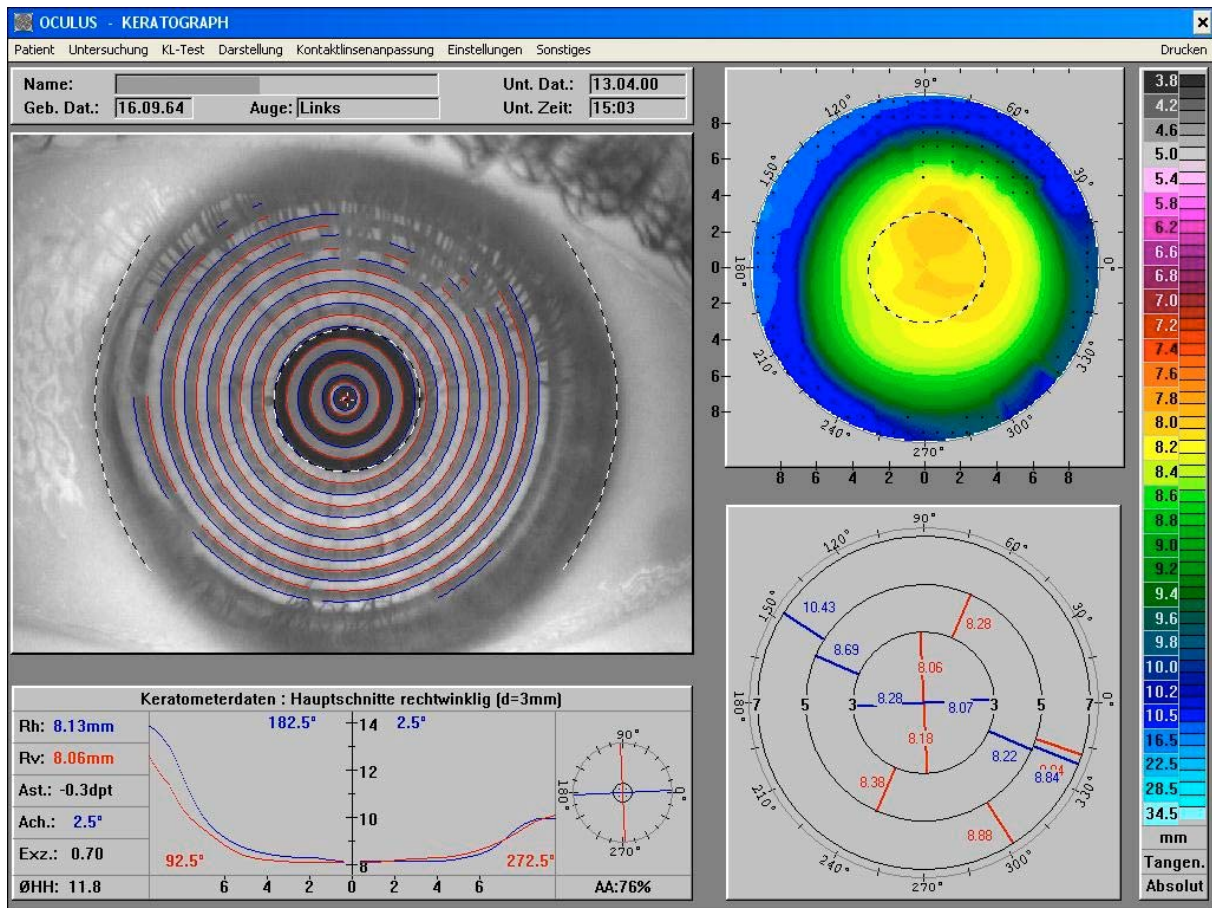
The patient did not bring in her current glasses because they were broken.

Keratography was performed and revealed the following data:

OD:



OS:



The patient was fitted with soft daily disposable lenses (Johnson&Johnson, 1-Day Acuvue) and rechecked several times. She was wearing the lenses about twice to three times per week for sports activity. She agreed to a checkup schedule on a yearly basis.

Emergency follow-up #1, 9 mai 2001

The patient complaints of a sudden reduction of visual acuity OD since this morning. She reports having heart problems but is unaware of her diagnosis and denies any use of medications.

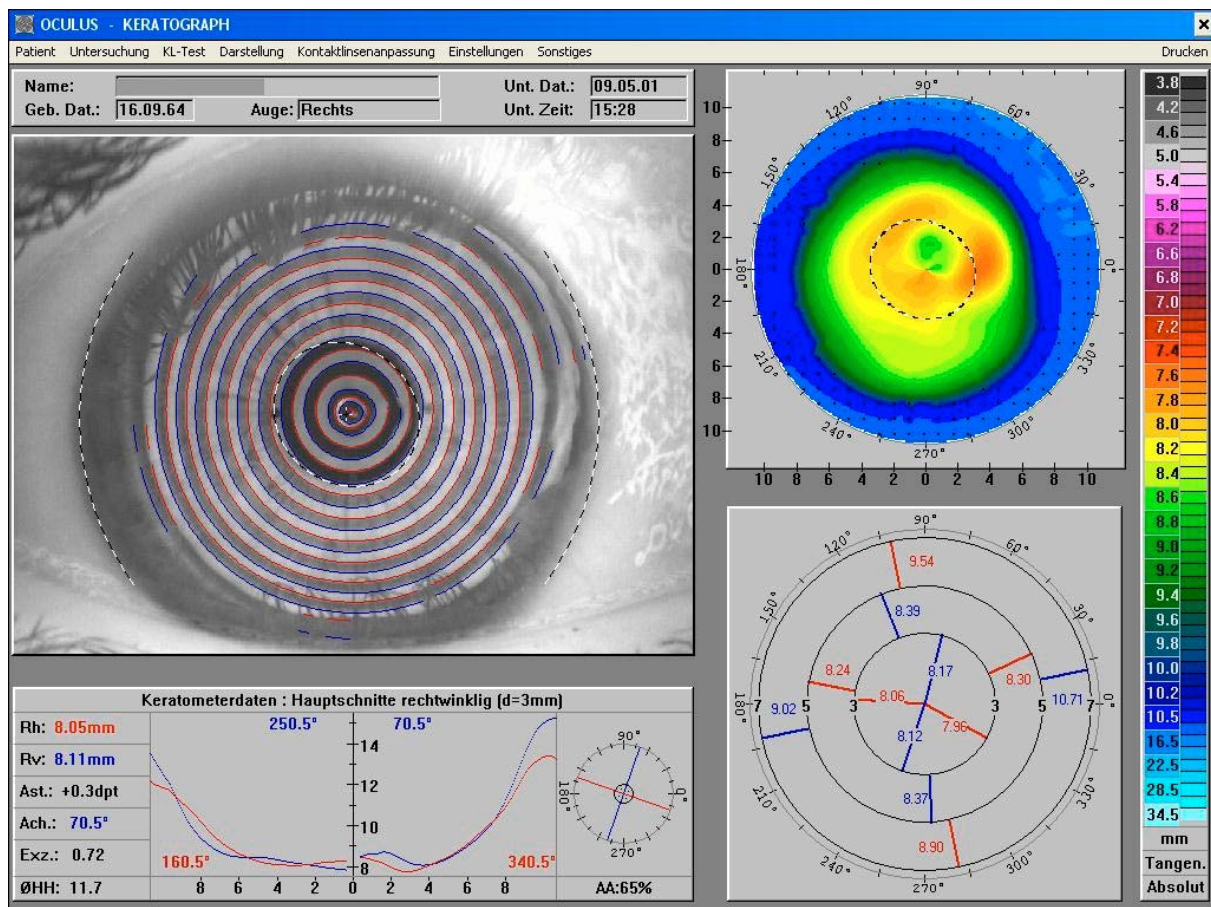
The refraction performed showed no change in myopia with a uncorrected and corrected visual acuity of 20/60 OD and uncorrected 20/30 and corrected 20/13 OS.

Slitlamp examination revealed a quiet, non-hyperaemic conjunctiva. The cornea showed a whitish S-shaped line centrally which was thought to be a Hudson-Staeli-Line OD. The cornea was clear OS. The lens presented clear OU, so did the anterior chamber with no cells and flare OU.

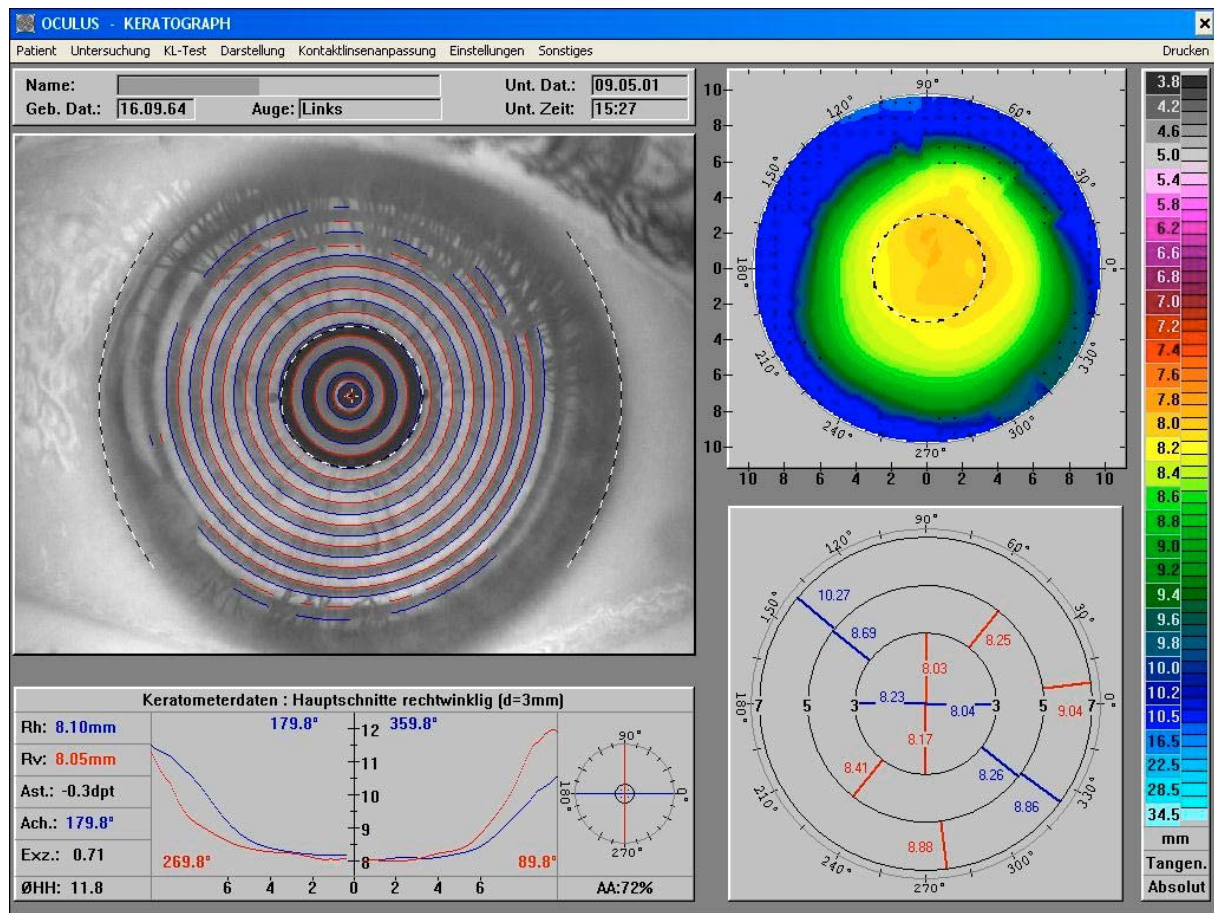
Undilated fundus exam with the 90D lens showed no difference to the initial findings one year ago OU.

Keratography was performed and revealed the following:

OD:



OS:



The keratography readings gave evidence to an irregular corneal astigmatism OD that has not been present one year ago. A rigid-gas-permeable contactlens was inserted into the right eye. The visual acuity with the lens was back to 20/20.

Due to the sudden change of corneal surface and according to the state law the patient was referred to an Ophthalmologist. The Ophthalmologist was unsure of the cause and the diagnosis of this phenomenon and perscribed Vis-Med, unpreserved wetting drops with hyaluronic acid. A checkup with him was scheduled three weeks later according to a phonecall with him. He recomended to temporarily stop the contactlens wear.

Follow-up #2, 16.3.2002

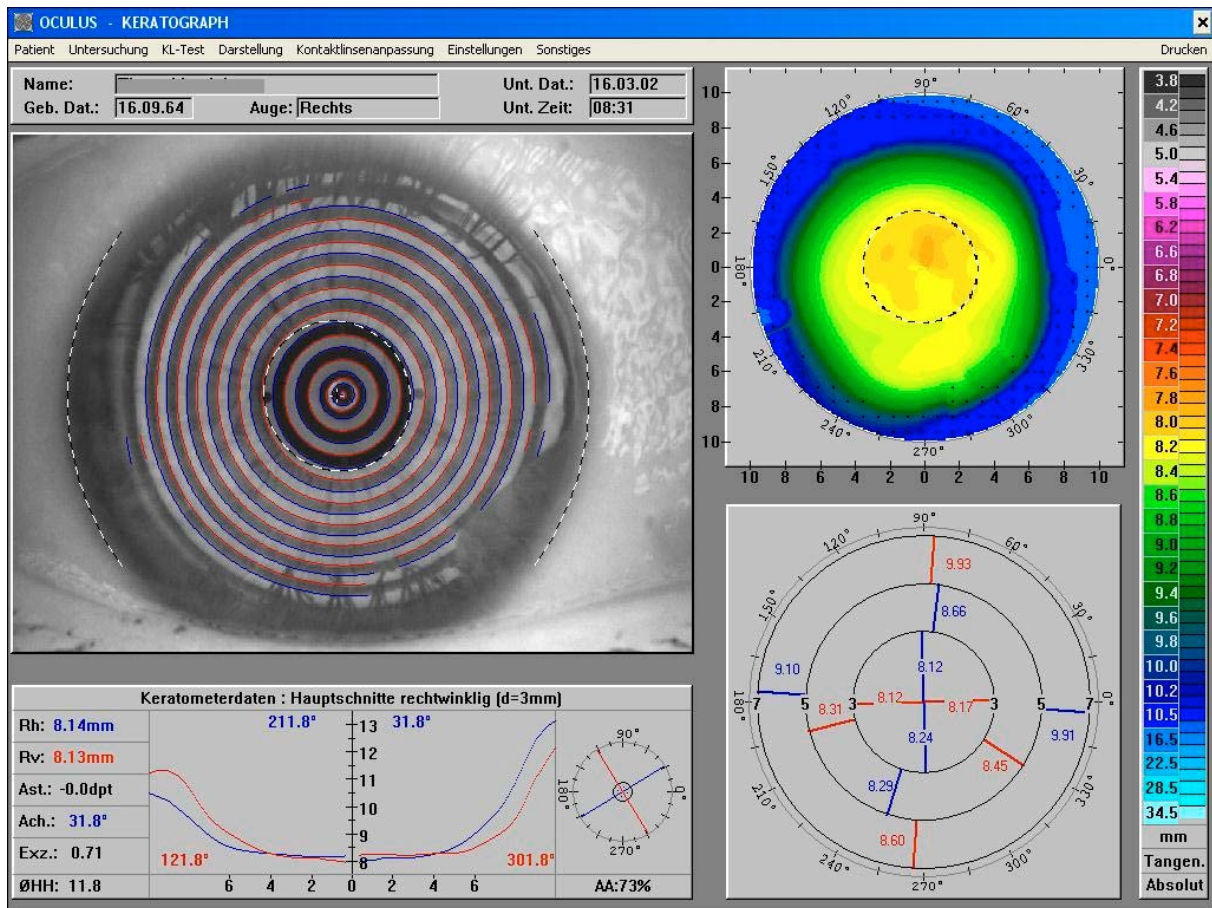
The patient showed up for a routing check-up. She was wearing her lenses only occasionally.

The slitlamp exam revealed the same S-shaped whitish line then one year ago OD and a clear cornea OS.

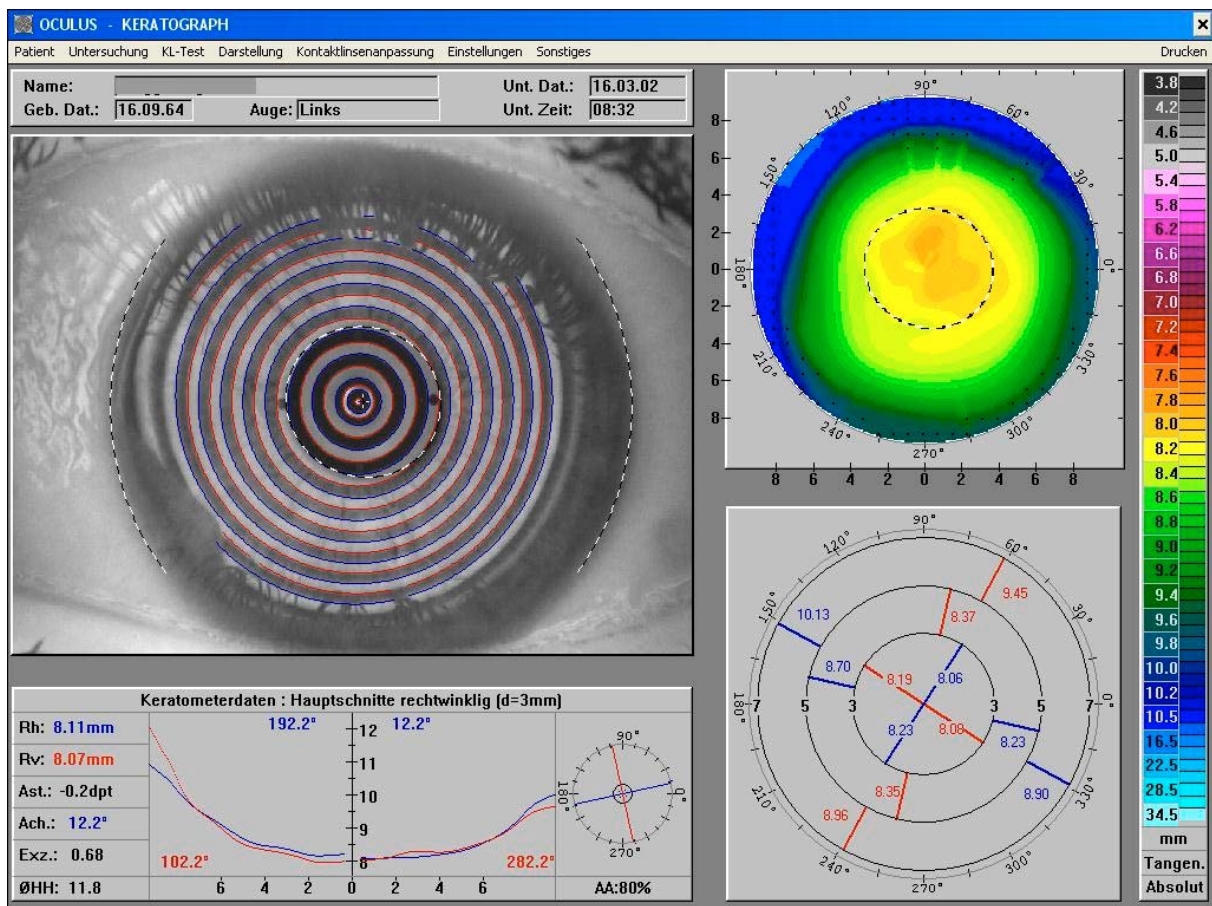
Visual acuities were uncorrected 20/40 OD and 20/25 OS. And corrected 20/16 OU. The refraction was -0.75 OD and -0.50 OS.

Keratography revealed the following:

OD:



OS:



Differential Diagnosis for this case includes the following:

- Retinal Vaso-Obliteration
- Corneal Abrasion
- Anterior Corneal Dystrophy
- Keratoconus
- Cataracts
- Uveitis

Retinal Vaso-Obliteration

Can be caused by atherosclerosis, periarteritis, haematological disorders or retinal migraine. All of these types have in common that the patient presents with a sudden total or partial loss of vision or restriction of the visual field which is usually monocular. The obliteration or its effects will be visible upon a fundus exam at any stage of the disease except with the retinal migraine.

Corneal Abrasion

Corneal abrasion is the result of mechanical trauma to the corneal surface. Usually the patient reports a history of direct or tangential impact trauma to the cornea from objects such as fingernails, paper, tree branch, mascara brush, or contact lens.⁴

Anterior Corneal Dystrophy

These dystrophies are classified by Kanski as follows:

Cogan microcystic dystrophy, Reis-Bücklers dystrophy, Meesmann dystrophy and Schnyder dystrophy.

All of these dystrophies are familial and progressive in character except the Cogan microcystic dystrophy as mentioned before.

Keratoconus

Keratoconus is a bilateral, asymmetric, cone-shaped deformity of the cornea. The underlying process is a progressive thinning of the paracentral cornea. Keratoconus is often associated with systemic disorders such as: Down Syndrome, Turner Syndrome, Ehlers-Danlos Syndrome, Marfan Syndrome, atopy, osteogenesis imperfecta and mitral valve prolaps. Also associations with other ocular diseases are common such as: vernal disease, Leber congenital amaurosis, retinitis pigmentosa, blue sclera, aniridia and ectopia lentis. In addition to the above mentioned possible predisposing factors are also PMMA contactlens wear and constant eye rubbing^{1, 2}.

Cataracts

Are opacifications of the crystalline lens. They are divided into age related and secondary cataracts. All cataracts interfere at a certain stage with the visual acuity and can be destructive to the eye in their end stages.

Uveitis

Uveitis is an inflammation of the uvea and/or adjacent structures. It can be classified anatomically, clinically or aetiologically. An uveitis usually presents with the following symptoms: photophobia, pain, redness (ciliary injection), decreased vision and lacrimation.

Diagnosis

Due to the signs and symptoms mentioned above the conclusion was made that the patient was suffering from a monocular epithelial basement membrane dystrophy.

Plan

The patient was educated about the risk of recurrent corneal erosions. She was told to pay special attention upon removing her contacts not to touch the cornea and to report to us any photophobia, pain, lacrimation or sudden decrease in visual acuity. In case of recurrent corneal erosions a pressure patch, cycloplegic agent and a prophylactic topical antibiotic should be applied. After healing a therapy with hypertonic saline solution (5%) four times a day for a period of 3 month would be appropriate.

Discussion

An anterior corneal dystrophy such as the epithelial basement membrane dystrophy can mimic many more serious diseases. A thorough history and eye examination is therefore essential to rule out any serious eye disease such as vaso-obliterations or uveitis. The EBMD itself is not a sight threatening disease as long as there are no recurrent corneal erosions and no scarring occurs and can just be watched on a yearly basis.

Conclusion

EBMD is the most common of all anterior corneal dystrophies. Therefore chances are very high that in a contactlens practice a patient can be first diagnosed of EBMD during a routine exam. Even though the disease itself is benign in nature it is an advantage for the patient to know his disease and how to react upon it. If in certain cases of recurrent corneal erosions healing does not occur or scars form the patient has to be referred for debridement of the cornea, stromal puncture or excimer laser keratectomy.

Bibliography

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