Darier's Disease and Corneal Opacity

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Dear Sir,

.- Tear film

– Surface cells

-Wing cells

Epithelium-■-

Basement 7

Bowman's layer

The cornea and conjunctiva can be considered as modified skin, and cutaneous disorders, especially that involving the epidermis, also affect the corneal epithelium. The epithelium, derived from the surface ectoderm is a 5- to 6-cell layer forming 10% of the corneal thickness; the stroma which is of mesodermal origin forms about 90% [1]. The basal cells that are attached to the basement membrane by hemidesmosomes give rise to wing cells which become the surface cells and are shed into the tears (fig. 1); thus, the corneal epithelium is structurally and physiologically somewhat similar to the epidermis. A case of Darier's disease with a peripheral corneal opacity is reported to stress the importance of examining the eyes in this genodermatosis.

A 36-year-old Malay lady was referred to the Skin Clinic for a spreading papular, warty lesion on the body since childhood. She was treated in the Eye Ward for a small left corneal ulcer of undetermined cause. The patient was a gravida 8, para 7, in 34 weeks of gestation with no history of injury or foreign body in the eye. She presented with redness, pain, lacrimation and photophobia of the affected eye which on slit-lamp examination showed fluorescein staining of the cornea at the 6-o'clock position and few cells at the anterior chamber. She was treated with mydriatics and antibiotics, and after a week, the ulcer healed leaving an oval opacity. A skin biopsy confirmed Darier's disease; her second son was also affected. We decided to start her on etretinate after a post-partum bilateral tubal ligation.

There are several skin disorders, especially genodermatoses, which are associated with corneal changes. In a study involving 21 patients with Darier's disease, various ocular abnormalities were observed [2]; unique peripheral corneal opacities, central corneal surface irregularities and keratotic plaques on eyelids were seen in 16 patients. The opacities were teardrop-sized to tiny, clustered oval-shaped lesions, and the irregularities were distributed in a radiating cobweb pattern. In two patients trephine biopsy of the opacities showed epithelial oedema especially of the basal cells and thickened basement membrane – features that are different from the skin pathology because the cornea is not a keratinizing epithelium. Darier's disease in two brothers associated with retinitis pigmentosa has also been reported [3]. In view of the high prevalence of

ocular involvement in this disease , it may be prudent to refer these patients for an ophthalmological examination before systemic retinoid therapy is initiated.

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Stroma

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References

Descemet's _ membrane

Endothelium """"

Fig. 1. Cross-section of the permission.

cornea. From Kanski [1], with

Kanski JJ: Clinical Ophthalmology. London, Butterworth & Co, 1984.

Blackman HJ, Rodrigues MM, Peck GL: Corneal epithelial lesions in keratosis follicularis (Darier's disease). Ophthalmology 1980; 87:931-943.

Itin P, Buchner SA, Gloor B: Darier's disease and retinitis pigmentosa: Is there a pathogenetic

relationship? Br J Dermatol 1988; 119:397-402.