PRE-DESCEMETIC CORNEAL DYSTROPHY*†

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The pre-descemetic corneal dystrophies, apart from cornea farinata, are rare disorders which have not been shown to be an inherited trait.

Maeder and Dahis (1947) described a 40-year-old woman with small white curved filaments occupying a disc in the deepest layers of the corneal stroma. The peripheral 2 mm. of cornea remained unaffected. The corneae of this patient showed keratoconus, clinically manifest in the left eye with a forme fruste in the right.

D’Ermo (1950) annotated a similar case in a 19-year-old girl who was also affected by keratoconus, and in whom the middle layers of the stroma were affected in the central area.

Franceschetti, Chodos, Dieterle, and Forni (1957) described a case in which punctate and filiform opacities were found in the deepest corneal layers, again leaving the corneal periphery unaffected. The corneal nerves were prominent, but keratoconus was not present. The conditions were designated deep filiform corneal dystrophy.

A similar condition, deep punctate corneal dystrophy, was first described by Franceschetti and Maeder (1954) in a 20-year-old patient. Bluish-white translucent points, some of which were prolonged into filaments, were scattered in the pre-descemetic layers. They were most densely distributed half-way between the centre and the periphery of the cornea. Under high magnification, they appeared like a knot of fine fibrils. No other corneal layers showed pathological change. The patient suffered from ichthyosis of moderate severity. The same condition was reported in a 50-year-old man with chronic simple glaucoma, and primary band-shaped degeneration of the cornea, who was also affected with congenital ichthyosis. Some of the opacities in this instance were comma-shaped and some were short rods, some straight and some angled; they occupied a central disc in the deepest stromal layers.

Kraupa (1924) described deep parenchymatous corneal changes in five patients with congenital ichthyosis. The opacities were grey points and flecks appearing like snowflakes under high magnification and were less marked at the periphery of the cornea.

Franceschetti and Schläppi (1957) reported, in a 56-year-old man with congenital ichthyosis and band-shaped corneal degeneration, bilateral pre-descemetic dystrophy where the opacities took the form of short irregular lines.

Case Report

A 62-year-old man was referred to the ophthalmic clinic because an optician had been unable to supply satisfactory glasses. He had a mild degree of myopic astigmatism which, when corrected, brought the visual acuity up to 6/12 partly in each eye. Slit-lamp examination showed both corneae to be affected by punctate and irregular opacities, mainly located in the deepest corneal layers just anterior to Descemet’s membrane. The peripheral part of the cornea was not affected. In the central area, some opacities extended into the middle and superficial layers of the stroma (Fig. 1). Some of the larger pre-descemetic opacities were situated in small, smooth, stromal protuberances, which caused the posterior surface of the cornea to be irregular, gave a distorted view of the fundus ophthalmoscopically, and broke up reflected light from the fundus (Fig. 2).

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The patient was of low intelligence and it was not clear whether he had suffered a worsening of vision. There was no evidence of keratoconus or of ichthyosis (examination by Dr. E. Raffles, Department of Dermatology, Dundee Royal Infirmary).

One parent, three siblings, nine nephews and nieces, and thirteen great-nephews and nieces were examined by the author, but none showed any corneal pathology.

Comment

This case shows differences from the cases of pre-descemetic corneal dystrophy previously described, in particular the irregular posterior corneal surface, but appears to resemble most closely the pre-descemetic dystrophies.

Summary

A case is described of pre-descemetic corneal dystrophy with some unusual features.

REFERENCES