A Case of Congenital Cornea Plana

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This abnormality was first described by Rübel in 1912. It is very rare and in view of the vexed question as to whether it is hereditary or purely spontaneous it seems justifiable to report a new case.

This involves a young man of twenty-one, the eldest of six children; his father had died. When he was six he was operated on bilaterally for alternating convergent squint and from childhood on had worn strong convex glasses.

At first it seemed as though it was a case of microcornea; the strikingly small, almost oval clear parts of the corneae made the most impression. On closer examination, however, the exceptional flattening of the corneae and of the whole anterior segment of the eyeballs sprang to light. The corneal limbus was flattened out and had progressed irregularly centrally, gradually changing into scleral tissue without having a limbal sulcus. At the same time the slitlamp revealed an extremely shallow anterior chamber and the distinct masking of the chamber angle by milkily-opaque conical tissue that in its deeper layers first cleared up centrally and subsequently changed gradually into a transparent corneal stroma, and more centrally an embryotoxon. The rest of the cornea was thin but completely clear.

The extended limbal zones revealed extensive superficial vascularisation which was clearly visible, especially on the upper sides of the corneae. The anterior layers of the irises showed slight atrophy and in R.E. the margins of the pupils were partially blurred; there were also filamentary anterior syn-echiae.

No abnormalities were observed on the lenses, the retinae or the optic disks.

Visual acuity R. E. with Sph + 7 C + 1 axis 170°: 0.5.

Visual acuity L. E. with Sph + 4.5 C + 3 axis 55°: 0.6.

Corneal diameter R. E.: 9.5 horizontally and vertically (including the blurred marginal zone).

Corneal diameter L. E.: 11 mm.

Corneal radius R. E.: at 130°: 10.5 mm, i.e., 32.5 D refraction.

Corneal radius L. E.: at 40°: > 11 mm, i.e., < 30 D refraction.

All the signs of an uncomplicated congenital cornea plana were present.

As far as stronger corneal refraction, respectively a smaller radius and smaller diameter are concerned, border-line cases have

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been described and Fríede uses the term pseudo-microcornea. He also presents the following differences between cornea plana and microcornea.

Cornea plana Microcornea

Abnormal flattening. 1. Normal corneal curvature.

Blurred limbus. 2. Clearly demarcated limbus.
 Corneal refraction 28–29 D. 3. Corneal refraction + 46 D.
 Corneal radius ± 10 mm. 4. Corneal radius ± 7.3 mm.
 Total refraction 7 D hyper- 5. Total refraction 2 D myopia, metropia, 9 D myopia. 18 D
 hypermetropia.
 Eyeball normal or enlarged. 6. Microphthalmos.

Lars and Eriksen (1949) distinguish two groups:

Cornea plana with no other abnormalities (3 sporadic cases and their own series of two families).

Cornea plana with microcornea, glaucomatous aniridia, cataract or other abnormalities.

With regard to heredity, my case offers no points of contact. I examined the brothers, sisters and mother but not one of them had any corneal abnormalities or substantial refraction anomalies at all. The parents are not consanguineous and they know of no “abnormal” eyes among their other relatives.

Larsen, in his series of three generations, affirms dominant heredity and Barkan and Borley (1936) describe a mother and two daughters with the complicated form of cornea plana. They also postulate dominant heredity. Rübel described three brothers, but here there is scarcely any question of a pure form of cornea plana. All the other authors report only sporadic cases.

Cornea plana probably originates from a developmental disturbance during the third or fourth foetal month, while there is still no difference between the sclera and the cornea. Other meso-dermal hypoplasias would accord with this (iris crypts, etc.).

Friede goes still further and believes that the retina and optic nerve would also reveal developmental disturbances (amblyopia, colobomata). Our case does not support this view.

I should like to emphasize once again that the much more frequent cases which at first sight

seem to be microcornea

may possibly; on further examination

turn out to have the characteristics of cornea plana. Description of a larger number of cases would undoubtedly lead to a better insight into their heredity and into the differentiation of the various forms of this anomaly.