SCLERO-CORNEA*

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SCLERO-CORNEA is a rare congenital condition in which a part or the whole of the cornea acquires the characteristics of sclera (Rollet, 1933; Lepri, 1949; Goldstein and Cogan, 1962). The process may affect the peripheral parts only or the entire cornea; when it involves the central portion the visual loss is considerable and the condition is noticed earlier. The peripheral type sometimes results in the appearance of an elliptical horizontal cornea, the affected area being vascularized with invading arcades from the superficial scleral vessels. In the total type the whole of the cornea is invaded by a superficial anastomosing network (Duke-Elder, 1964). The vascularization is almost always regular, there is no evidence of inflammation, the condition does not progress, and there is no lucid zone between the opacity and the limbus.

The condition is believed to be due to an aberration of the paraxial mesoderm anterior to the optic cup at the 16 to 20 mm. stage when the cornea is developing (Duke-Elder, 1964). It may occur alone or with cornea plana, microphthalmos, nystagmus, and anomalies of the anterior chamber angle leading to glaucoma (Franceschetti, 1946; Goldstein and Cogan, 1962) or embryotoxon in the other eye (Duke-Elder, 1964). It may be associated with other congenital malformations, such as polydactyly, lacunae of parietal bones, cranial dystrophies, deformities of the ears, and cerebellar anomalies (Goldstein and Cogan, 1962).

Sclero-cornea should be differentiated from arcus juvenilis (anterior embryotoxon), vascular keratitis, and arcus senilis.

It differs from the first by the absence of a lucid zone, the presence of superficial arcades of scleral vessels, and the absence of any hereditary tendency. Histologically the collagen fibres are arranged in an irregular manner while in embryotoxon there is intracellular deposition of lipid granules.

Sclero-cornea differs from vascular keratitis in the absence of inflammation and the presence of scleral vessels in the cornea, and from arcus senilis in that it is present from birth, and is non-progressive.

Two cases of peripheral sclero-cornea seen at the eye department of Irwin Hospital, New Delhi, are presented below.

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Case Reports

Case 1, a 65-year-old Hindu male, attended the out-patient eye department on August 20, 1963, with the complaint of a peculiar appearance of both eyes since birth and progressive bilateral diminution of vision for the past 3 years. According to the patient the horizontal and elliptical shape and small size of the eyes had not changed since birth.

Examination.—Both the corneae appeared to be of normal size. The peripheral 2 to 3 mm. of the cornea was white and sclerotic and there was no lucid zone outside the opacity. The transparent part of the cornea was elliptical, measuring 7 mm. horizontally and about 5 mm. vertically (Fig. 1). The lenses showed nuclear sclerosis which accounted for the diminution of vision.

General examination revealed no abnormality.

FIG. 1.—Case 1. Sclero-cornea with central elliptical transparent cornea.

Case 2, a 45-year-old Hindu male, attended the out-patient eye department on November 11, 1963, with the complaint of gradual, painless loss of vision in the left eye for the past 2½ years.

Examination.—There was a white sclerotic peripheral opacity in both corneae, more marked in the upper part. There was no lucid zone between the opacity and the sclera. There were bilateral immature senile cataracts. The peripheral corneal opacity had been present since birth (Fig. 2).

The ocular tension was 43 mm. Hg in the left eye, the vision was reduced to hand movements at 12 in., and projection of light was defective. The left fundus showed marked nasal shifting of blood vessels with optic atrophy. The secondary glaucoma was not controlled by Diamox and miotics, and as light-projection remained faulty the eye was enucleated.

FIG. 2.—Case 2. Peripheral sclero-cornea with raised intra-ocular pressure in the left eye.

Discussion

The outer coat of the eyeball (sclera and cornea) is transparent in very early intra-uterine life, and the sclerotic process starts in the region of the equator at the 7 mm. stage (Arey, 1954; Barber, 1955) and proceeds both anteriorly and posteriorly. It is possible that there is some inhibiting influence in the region of the limbus which limits its further progress. In cases of sclero-cornea this influence may be lacking or may be exerted at a later stage, so that the sclerotic process continues unchecked into the periphery of the cornea; if it occurs in the region of the angle, it may affect the trabecular meshwork and interfere with the drainage mechanism. This would explain the presence of glaucoma in Case 2.

Summary

- (1) Two cases of peripheral sclero-cornea are presented.
- (2) It is suggested that the condition may be due to a continuation of the sclerotic process into the periphery of the cornea through defect or delay in the normal development which leaves the cornea clear.
- (3) Sclerosis in the region of the anterior chamber angle around Schlemm's canal may predispose the eye to secondary glaucoma.

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