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CASE NOTES

SCLEROMALACIA PERFORANS*

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SCLEROMALACIA PERFORANS is a rare ocular disorder almost meriting the name of an ophthalmic curiosity. A few authentic cases have been reported from various parts of the world, each with minor variations in ocular symptomatology and/or varying associated lesions elsewhere, so that no typical pattern is presented by any two cases. This has inevitably led to confusion in terminology and classification.

Case Report

A male cultivator was admitted to the Victoria Memorial Eye Hospital, Colombo, on November 8, 1957, with a history of defective vision of 15 years' duration in the left eye and of 5 years' duration in the right eye. Lacrimation and smarting had begun in the left eye for no known reason, and about a fortnight later the patient had noticed small whitish areas on the periphery of the cornea which gradually increased while his sight grew misty. As time went on a bulge appeared on the upper part of the left eye above the cornea, but he had had no pain or redness in that eye at any time during the course of 15 years. About 5 years ago similar symptoms occurred in the right eye and he could recall that the right eye underwent the same slow changes as the left, again without redness or pain.

Examination.—The palpebral apertures were unequal in size, the left being the larger. The right eye (Fig. 1, overleaf) showed a centrally clear cornea with a peripheral non-vascularized hazy annulus about 3–4 mm. in width. In the supero-nasal quadrant of the paralimbal zone was an ectasia of uveal tissue such as may be seen after a cataract extraction. In the less bulging parts it resembled an iridencleisis though there was no classical bleb as may be seen in a successful filtration operation. Through an ectopic pupil—drawn towards the limbal ectasia—in a tremulous iris, the lens was seen to be totally dislocated into the vitreous chamber. The fundus was only dimly visible—a proper view being precluded by the alignment of the ectopic pupil with the hazy periphery of the cornea. The visual acuity was 5/60 with +10 D sph. The intra-ocular pressure was 25 mm. Hg Schiötz.

Knobbly ectasiae studded the supero-nasal quadrant of the paralimbal zone of the left eye. The cornea was a total whitish haze, with its curvature flattened by the bulge above. The ectasiae were all of a blue-grey colour showing thinned-out conjunctiva with underlying uveal tissue (Fig. 2, overleaf). The cornea was vascular. No intra-ocular examination was possible. This eye was totally blind, and the intra-ocular pressure could not be recorded.

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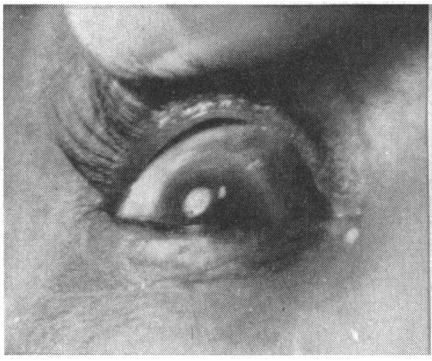

FIG. 1.—Right eye, showing centrally clear cornea and a bulge supero-nasally. This has the appearance of an iridencleisis.

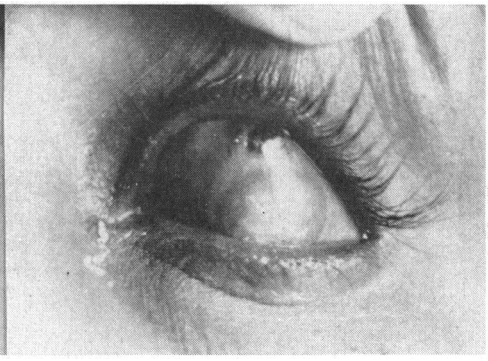

FIG. 2.—Left eye, showing opaque grey-white cornea with a bulge above and slightly nasally. In the centre of the bulge two knobbly masses of uveal tissue are clearly seen.

General Examination.—The man was well nourished, and the teeth and nasal sinuses were normal. There was no swelling or limitation of movement in the joints. The blood pressure was 110/75 mm. Hg.

Laboratory Investigations.—The Kahn test was negative.

White cell count: 8,400 per c.mm.; differential count: polymorphonucleocytes 57 per cent.; lymphocytes 32 per cent.; eosinophils 11 per cent.

Erythrocyte sedimentation rate: 11 mm./hr; 28 mm./2nd hr.

Blood cholesterol: 208 mg. per cent.

X Rays—Spine, sacro-iliac joints, limbs, and chest normal.

Discussion

This case is one of bilateral intercalary staphylomata. In the absence of any history of trauma or acute inflammatory episode this appears to be a case of scleromalacia perforans, unassociated with joint lesions or cholesterolaemia. The absence of scleral holes of the type usually seen makes it resemble the atypical form designated "paralimbal or intercalary scleromalacia perforans" by Franceschetti and Bischler (1950) or "spontaneous scleral intercalary perforation" by François (1951).

The dislocation of the lens may have been due to a slow and steady expansion of the ciliary ring with the ectatic sclera. The iris prolapse may arise because the sclera yields under the normal intra-ocular pressure through softening or divarication of the scleral fibres. In true scleromalacia perforans scleral holes appear with amazing rapidity but without ectasia of uveal tissue, apparently because there is usually a retinal detachment with attendant lowered intra-ocular pressure, and the holes over the choroid do not invite the herniation of that part of the uvea. In cases of scleromalacia perforans in which the cornea is perforated, a cystic swelling has been described (Duke-Elder, 1938).

The case here presented fulfils the criteria of spontaneous scleral intercalary perforation (François, 1951):

(*a*) There is a resemblance to a cystoid cicatrix or an iridencleisis at the limbus,

(*b*) There are no inflammatory symptoms past or present.

It differs from this definition, however, in the following respects:

(*a*) It is bilateral

(*b*) Prognosis is poor

(*c*) The cornea is involved

(*d*) There is no joint involvement

(*e*) There is no cholesterolaemia.

The absence of joint involvement and cholesterolaemia does not prevent this case being regarded as a variant or *forme fruste* of scleromalacia perforans as in the cases of van der Hoeve (1934), Eber (1934), and Arkle and Ingram (1935).

Judging from the non-episodic nature of the condition, the absence of scleral holes, and the very slow progress, it is suggested that it should be called scleromalacia simplex or tarda, or "paralimbal or intercalary scleromalacia", omitting the word "perforans" altogether.

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