

CASE NOTES

BILATERAL ESSENTIAL ATROPHY OF THE IRIS*

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ESSENTIAL atrophy of the iris is a rare clinico-pathological entity; its aetiology is ill-understood, almost all cases reported have been unilateral, and treatment is of no avail. We report this case not only because it is rare, but also because bilateral cases are rarer still. This case, which appears to be closely allied to buphthalmos, like that of Barkan which was also bilateral (Duke-Elder, 1940), also appears to shed some light on the probable aetiology of this condition.

Case Report

A male farmer, aged 26, was admitted to hospital on July 20, 1954, with a history of progressive blindness. The left eye had been blind for 3 years and the right for 3 months. Neither redness of the eyes nor ocular pain was a feature of his illness. A slowly progressing baldness was the only extra-ocular complaint—this he had noticed for 5 years.

Family History.—Two brothers and parents healthy.

Examination.—A healthy muscular individual whose only extra-ocular defect was baldness of the frontal area of the scalp with inroads to the vertex. The skin over these areas was of normal texture; the hair, which was predominantly black, was brownish over the vertex, and was greying at the sides. The Wassermann reaction was negative.

Ophthalmic Investigation.—Both eyes were buphthalmic and executed a series of fine nystagmoid movements in the horizontal direction. Vision in each was perception of light. The corneal diameters were: Right horizontal 14 mm., vertical 13mm.; left horizontal 14 mm., vertical 14 mm.

The ocular tension was 26 mm. Hg (Schiötz) in the right eye with pressure of 7.5 and 10 mg., and 44 and 37 mm. Hg (Schiötz) in the left eye. The left cornea was cloudy because of oedema of the epithelium and substantia propria. The anterior chambers were deep. The irides (Figs 1 and 2, overleaf) showed two shades of brown pigmentation, the paler areas having a washed-out appearance; there were multiple holes—pseudopolyoria. The pupils were eccentric and ectropion uveae delineated their borders. Strands of atrophic iris tissue showing lines of traction radiated from the pupils. No contraction furrows, crypts or collarettes were seen. The irides looked as though they had been ironed out. The left iris was tremulous opposite the three holes below the pupil (Fig. 2). The pupils reacted sluggishly to light. There were no keratic precipitates or deposits of iris pigment on the back of the cornea.

Through the slight dilatation that could be effected with homatropine 2 per cent. drops, the lenses were seen to be sclerosed but the fundi could not be seen.

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FIG. 1.—Right iris showing eccentric pupil with ectropion uveae, two holes, atrophy, depigmentation, and lines of traction.

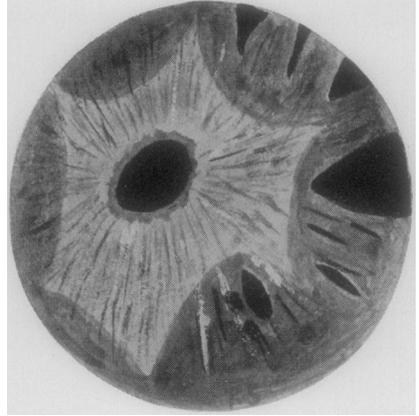


FIG. 2.—Left iris showing eccentric pupil with ectropion uveae, and multiple holes peripherally and in the mid-zone of the iris.

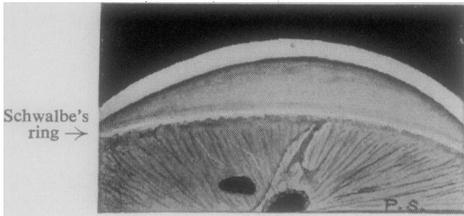


FIG. 3.—Gonioscopic view of the upper angle of the right eye, showing a false angle, moth-eaten appearance of the periphery of the iris, Schwalbe's ring, and a thick band running from the angle to the middle of the iris.

and there could be seen thickish bands coated with a whitish mantle running from the pseudo-angle to the mid-zone of the iris into which these bands dipped rather like roots of a plant. At the site of insertion of these bands there were large islands of iris atrophy. There was no deposit of iris pigment in the angle (Fig. 3).

Comment

The interesting features of this case are its bilaterality and its association with buphthalmos; otherwise it conforms with the description of this condition given by previous writers. The partial baldness suggests a dystrophic aetiology. The association of baldness with uveal disease is known in the Vogt-Koyanagi syndrome, though this condition is inflammatory rather than dystrophic.

The state of the irides, the buphthalmos, and the degree of blindness show that the glaucoma must have been antecedent to the atrophy of the iris, for in a man of this age buphthalmos could not appear if the raised tension were of recent origin.

Buphthalmos is a common concomitant of congenital aniridia in which there is an abnormal insertion of the fringe of iris into the angle of the anterior chamber. In essential atrophy of the iris it would appear that there is an abnormal insertion of the root of the iris into the cornea instead of into the ciliary body. This anatomical anomaly may be connected with the cleavage of the mesoderm at the angle of the anterior chamber. The whitish bands seen running from the false angle probably arise from the site of termination of Descemet's membrane. It is probable that, owing to the abnormal insertion of the iris into the cornea, the blood supply to the iris is either absent or insufficient.

Gonioscopic findings in this case support the view that there is an abnormal adherence of the iris to the cornea (Duke-Elder, 1940). An anatomically blocked angle caused the raised tension, and this was responsible for the loss of sight. The enlargement of the globe probably caused traction on the iris through the abnormal extensions of Descemet's membrane. Continued traction would not only tear off the iris at a point opposite to the site of traction but would also cause a form of pressure atrophy. An attenuated blood supply would add an ischaemic factor and aggravate the atrophic process.

It is suggested that essential atrophy of the iris is a variant of buphthalmos determined primarily by an abnormality at the angle of the anterior chamber. The parallel with congenital aniridia also points to the same conclusion. This may account for the fact that many of the cases so far reported are unilateral.

REFERENCE

DUKE-ELDER, S. (1940). "Text-book of Ophthalmology", vol. 3, p. 2404. Kimpton, London.