ABERRANT INTRA-OCULAR LACRIMAL GLAND TISSUE*

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THE occurrence of aberrant lacrimal gland tissue within the eye is a great rarity and presents problems of diagnostic and embryological interest. A search of the literature has revealed only three reports of this condition. The first was that of Puech (1887), who briefly described an adenoma of the choroid in an adult female: it had the microscopic characteristics of lacrimal gland tissue, and was bounded by and was adherent to the sclera and to the retinal pigment epithelium, both of which were normal. The patient was otherwise healthy both before and after operation. The second was that of Christensen and Anderson (1952), who described aberrant lacrimal tissue within the sclera, ciliary body, limbus and iris in the upper temporal quadrant of the left eye of a full-term infant of 2 weeks; the tumour was globular and pinkish in colour with solid and cystic portions. The third was that of Bruce (1952), who demonstrated lacrimal tissue in the excised iris of the lower nasal quadrant of the left eye of a 2-months-old male infant born 2 weeks prematurely: this tumour was also pinkish-white in colour, irregular in contour, and appeared cystic. No other abnormalities were noted in these infants.

This present report adds to the foregoing a further case recently submitted for pathological examination at the Institute of Ophthalmology.

Case Report

Unfortunately only a few clinical details are available. At the age of 7 weeks numerous cysts were noticed in the outer and lower quadrant of the left iris of an otherwise completely normal and healthy male baby. It was not possible to examine the lesion with the slit lamp; 8½ months later the eye was removed.

Pathological Report

The specimen was a left eye in which a cystic mass and some small white opacities occupied the lower outer quadrant of the iris from 4 to 6 o'clock. Just posterior to the limbus at this site a small nodule presented in the outer sclera anterior to the insertion of the inferior rectus.

A vertical section was made through the globe and the interior showed an enlarged ciliary body and iris root displacing the lens antero-superiorly and narrowing the anterior chamber. Posteriorly the globe appeared to be normal.

Histology

Macroscopic examination of serial sections of the whole eye showed the enlargement of the ciliary body and the iris root to be due to a cystic lesion, which pushed the iris slightly forward and indented the inferior equator of the lens. The scleral nodule was a solid lesion in the middle and outer scleral layers.

* Received for publication November 26, 1959.

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Microscopic examination showed the cystic lesion to consist of a glandular tumour lying in the ciliary body, iris, and adjacent limbus and peripheral cornea. It contained three large retention cysts communicating with one another and with dilated acini lying in a semi-circular rim around their postero-inferior margin (Fig. 1).

FIG. 1.—“Normal” aberrant glandular tissue in sclera, and cystic glandular tissue in ciliary body and iris. Two of the large retention cysts may be seen, the dilated and normal acini lying postero-inferiorly around their rim. Haematoxylin and eosin. $\times 9$.

These cysts contained serous eosinophilic material and were lined with a double row of flattened epithelial cells having large oval basophilic nuclei. A few normal serous acini were present but no normal collecting tubules or ducts could be differentiated (Fig. 2, and Fig. 3, opposite).

FIG. 2.—Higher-power view of dilated and normal acini seen in Fig. 1. Haematoxylin and eosin. $\times 36$.

FIG. 3.—Higher-power view of acini seen in Fig. 2. Haematoxylin and eosin. $\times 375$.

The normal architecture of the area was distorted, the tumour extending into the iris to within 0.25 mm. of its tip, into the cornea to within 0.25 mm. of its surface bulging the superficial lamellae outwards over it (Fig. 4), and into the ciliary body as far as its posterior third, the ciliary muscle being split around it. The filtration angle, Schlemm's canal, and the trabecular meshwork were obliterated, and fibrous tissue had bound the tumour to the posterior corneal lamellae; the endothelium and Descemet's membrane were reflected around the false angle on to the tumour surface, and the whole iris was drawn forward, narrowing the anterior chamber.

FIG. 4.—Medial to plane of section seen in Fig. 1, cysts extend into cornea and obliterate structures of distorted angle. A second cyst is seen extending almost to the iris tip. Haematoxylin and eosin. $\times 30$.

The nodule of tissue in the outer sclera closely resembled normal lacrimal gland and at its closest point was only 1.25 mm. from the glandular tissue in the ciliary body. Antero-posteriorly the nodule measured from 1 to 3.75 mm., and inwardly it extended into the inner third of the sclera. The acini appeared normal and several ducts were contained (Figs 1 and 5).

FIG. 5.—Higher-power view of "normal" glandular tissue in sclera. A duct is seen centrally. Haematoxylin and eosin. $\times 100$.

No duct tissue could be found connecting the glandular tissue in the iris and ciliary body with the corneal or conjunctival surfaces, or with the scleral portion, while episcleral tissue was not available for examination to determine whether a duct system connected the scleral portion with the conjunctival surface.

Discussion

Developmentally the occurrence of lacrimal gland tissue in these abnormal sites is not completely understood, but that it should occur is not surprising; indeed it is remarkable that abnormalities of this type are so rare for the ectodermal structures of the eye and lids arise from a very small area, the cells of which at an early developmental stage must be multipotent. The stimulus or influence determining the development of the normal lacrimal gland is unknown and therefore the initiation of aberrant development is similarly obscure; however, budding of the surface ectoderm into the underlying mesoderm must occur initially from an abnormal site or in an abnormal direction.

Various abnormal locations for normal lacrimal tissue have been reported (Hughes and Ballen, 1956; Boase, 1954, François and Rabaey, 1951; Dame 1946, Duke-Elder 1932, 1938), and, as Reese (1951) points out, lacrimal gland tissue may normally be found at almost any site in the lateral half of the

orbit, and sometimes as far away as the lower fornix. Furthermore, the lacrimal gland in its phylogenetic development moves from the inner canthus along the lower lid to the outer canthus, and then to the upper lid; the ducts which persist under the lower lid remain as an indication of this course.

In the human embryo the lacrimal gland first appears usually at about the 25-mm. stage, although it has been noted as early as the 22-mm. stage. Budding continues up to the 60-mm. stage, and the histologically similar glands of Krause appear at the 55-mm. stage. The extent of the inward growth of the budding aberrant lacrimal tissue will depend on the time that budding begins, and on the stage of development of the underlying mesodermal and neuro-ectodermal tissues. As Mann (1957) points out, the more gross abnormalities of the eye tend to arise in the earlier stages of development, for after the definitive structures are established, abnormality can never be so extensive. Aberrant lacrimal tissue in the iris would suggest an anomaly in early development, whereas aberrant tissue only in the conjunctiva or outer cornea indicates a later developmental defect.

In explanation of the case here reported it is suggested that the aberrant tissue grew into the undifferentiated mesoderm probably at the 25-mm. (7-weeks) stage in an atypical direction and from an atypical ectodermal site (conjunctival), to abut against the early choroidal and outer neuro-ectodermal layers near the rim of the optic cup, and in front of the scleral condensation for the insertion of the inferior rectus muscle. Further distribution of the tissue into the situation seen in Fig. 1 might be explained by considering the development of the area into which the aberrant tissue had grown. The neuro-ectodermal cup extends forward at about the 50-mm. (10-weeks) stage accompanied by its overlying mesoderm, to form the epithelium and the inner mesodermal layer of the ciliary body and iris. This forward movement would tend both to direct growth of the tip of the aberrant tissue forwards and also to carry small portions of it anteriorly. Somewhat later, in the 5th month, the mesodermal differentiation of the ciliary body and limbal areas would tend to accentuate this displacement of the glandular tissue. The ciliary body grows unequally (Allen, Burian, and Braley, 1955), and the greater growth which occurs forwards and outwards would carry the tissue into the future area of the chamber angle. The forward movement of the limbus, from its early position over the ciliary body to its final location immediately anterior to the angle, would assist. With this forward passage, the tip of the glandular ingrowth would be stretched and thinned through elongation, apparently enough in our case to sever its connexion with the original site, thus leaving a nodule in the sclera discharging superficially *via* a duct system, and a component in the ciliary body, iris, and limbus, which having no outlet developed retention cysts.

The aberrant growth inwards in the case reported by Puech (1887) apparently took place into mesodermal tissue surrounding the optic cup

posterior to its rim, and was restricted thereby to development in the choroid; however, the lack of detail in this report does not allow of any further conclusions.

In the case reported by Christensen and Anderson (1952), the aberrant growth inwards probably occurred somewhat more anteriorly and from at least two separate buds. The mass of glandular tissue in the ciliary body was a conglomerate one, and probably became separated from the scleral mass as the sites of origin in the limbal and conjunctival epithelium drifted apart with differentiation and enlargement of the globe.

In Bruce's case the local excision precludes comparison; however, it would appear unlikely that closure of the foetal fissure played any part in the displacement of the aberrant tissue as he suggested.

A less likely explanation of aberrant glandular tissue within the eye is differentiation *in situ* of epithelial islands carried into the mesoderm with or by the lens plate. This would not account for the duct system seen in the case reported by Christensen and Anderson; nor, in considering the development of the anterior eye, does the distribution of the aberrant tissue in their case or in the one now reported, lend itself to this explanation. The suggestion that aberrant tissue may grow through scleral defects is similarly less satisfying.

The last three reported cases, including the present case, show aberrant lacrimal gland in three differing quadrants of the left eye. It is likely, however, that any quadrant of either eye may be the site of election. They all presented as partially cystic and partially solid iris tumours, pinkish-white in colour, and enlarging. Presumably with a patent duct system to drain the tissue the lesion could be entirely solid. A nodule was present on the sclera in one case and limbal cysts were present in two cases.

Summary

The occurrence of aberrant lacrimal gland tissue is less remarkable than its rarity, only three intra-ocular cases having been found in the literature. A fourth case is here described.

As the stimulus for normal development is unknown, it is impossible to explain exactly how this abnormality occurs, but the site and time of the aberrant stimulation probably determine the area and extent of the ocular involvement. Reasons are given for the belief that a late aberrant stimulus causes involvement of only the epibulbar and superficial layers of the globe, whereas an early aberrant stimulus will involve the deeper structure. Aberrant ingrowth of lacrimal buds into the mesoderm adjacent to the rim of the optic cup will probably involve the limbus, ciliary body, and iris, and will disturb the normal structure of this area, presenting at or shortly after birth, the clinical signs of a gradually enlarging solid, or more likely a cystic, tumour.

The pathology of these cases suggests that local excision, if feasible, may be all that is required when surgical treatment is indicated.

I am indebted to Professor Norman Ashton for permission to report this case and for his helpful suggestions in its presentation.

REFERENCES

- ALLEN, L., BURIAN, H. M. and BRALEY, A. E. (1955). *A.M.A. Arch. Ophthalm.*, **53**, 783.
- BOASE, A. J. (1954). *Brit. J. Ophthalm.*, **38**, 380.
- BRUCE, G. M. (1952). *Trans. Amer. Acad. Ophthalm. Otolaryng.*, **56**, 47.
- CHRISTENSEN, L., and ANDERSON, E. DEM. (1952). *A.M.A. Arch. Ophthalm.*, **48**, 19.
- DAME, L. R. (1946). *Amer. J. Ophthalm.*, **29**, 579.
- DUKE-ELDER, S. (1932). "Text-book of Ophthalmology", vol. 1, pp. 315-369. Kimpton, London.
- (1938). *Idem.*, vol. 2, pp. 1775-7.
- FRANÇOIS, J., and RABAËY, M. (1951). *Brit. J. Ophthalm.*, **35**, 237.
- HUGHES, W. L., and BALLEEN, P. H. (1956). *A.M.A. Arch. Ophthalm.*, **55**, 271.
- MANN, I. (1949). "The Development of the Human Eye", 2nd ed. B.M.A., London.
- (1957). "Developmental Abnormalities of the Eye", 2nd ed., p. 7. B.M.A., London.
- PUECH (1887). *J. Méd. Bordeaux et du Sud-Ouest*, Feb. 27.
- REESE, A. B. (1951). "Tumors of the Eye", p. 488. Hoeber, New York.
- WOLFF, E. (1954). "The Anatomy of the Eye and Orbit", 4th ed. Lewis, London.