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COMMUNICATIONS

MICROPHTHALMOS*

BY

GILBERT FORBES

FROM THE DEPARTMENT OF FORENSIC MEDICINE,
THE UNIVERSITY, SHEFFIELD

A FEMALE child was born after it had completed the sixth month of intra-uterine life but before reaching the seventh month. It lived for thirty hours and then died due to prematurity. A post-mortem examination was made three days after death. It was found that the child had a supernumerary digit on the ulnar side of the right hand, that the eyeballs were deeply sunken into the orbits and were smaller than normal. Behind the pupils an opacity could be seen. No other congenital defects were found.

One eyeball was removed, fixed and cut in serial sections. Unfortunately, for various reasons, it was not possible to make a dissection of the contents of the orbit.

The eyeball was found to measure 8 mm. in antero-posterior diameter and on naked eye examination was observed not to be absolutely spherical.

Histology

Sclera.—The sclerotic is of average thickness and of normal structure. Muscular insertions into the globe can be seen. In the lower anterior part of the eye there is a bridge of scleral fibrous

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FIG. 2. KEY TO FIG. 1.

tissue cutting the cavity of the eye into two parts, one large and one small (Figs. 1 and 2). This smaller part is the reason for the asymmetry mentioned above and if it were amputated the section of the eye would be much more nearly circular. The bridge is of rather finer and more vascular fibrous tissue than the sclera proper, and in it portions of hyaline cartilage can be seen. This partition arises near the sclero-corneal junction anteriorly. It is apparent therefore that there is a cyst attached to the lower anterior part of the eye,

Cyst.—This is lined by choroid and by the pigment layer of the retina in its anterior one third. The choroid is thrown into irregular folds which protrude into the cyst. The pigment epithelium, which is present only on those parts which are covered by choroid, is very heavily pigmented and is irregular in depth. The appearance suggests that the pigment layer has been thrown into secondary folds on the folded choroid. The cyst is not completely cut off from the cavity of the eye and fragments of disorganised retinal tissue can be seen passing through the gap. The interior of the cyst contains a mass of plicated disorganised retina, in some places opposed to the choroid, in others to the fibrous wall of the cyst, and also lying apparently free in the cyst cavity. The outer wall of the cyst is continuous with the sclera. In places

FIG. 1.
Section of eye.

it appears to be of the same consistence as the sclerotic, and in other places it is much less dense. This suggests that the wall was imperfect and has been reinforced by condensed orbital fibrous tissue.

Conjunctiva and cornea.—The structure of the conjunctiva is normal. No clear Bowman's membrane can be seen in the cornea. The superficial corneal layers are unduly cellular and the fibrous strands are less dense and less homogeneous than in normal corneal tissue. The structure of this part corresponds to that of the sclera and is vascularised. Descemet's membrane and the epithelium covering it are normal.

Choroid.—The choroid lines the interior of the globe from the upper ciliary body to the opening into the cyst where it ends abruptly. The anterior one third of the cyst is lined by choroid of irregular depth and thrown into many folds. An irregular broken semi-circle of what appears to be vascular fibrous tissue passes from one free edge of the iris to the other behind the lens (Figs. 1 and 2) separating it from the disorganised retina. The pigment of the choroid is scanty. In sections beyond the periphery of the cornea and iris the lens can still be seen, and the choroid is found to line the whole of the inside of the sclera, including the inner surface of the fibro-cartilaginous partition separating the cavity of the cyst from that of the eye proper. There is of course a break in the continuity of the choroid where the retina passes out into the cyst and this gap can be clearly seen in peripheral sections. Beyond the gap the retina is directly opposed to the inner surface of the sclera. In the anterior part of the eye, the choroid is thrown into folds and it projects as vascular papillae into the interior of the globe.

Ciliary processes and iris.—These structures are small and poorly developed but the ciliary process on the side nearest the cyst shows an hypertrophied irregular outgrowth projecting backwards. The ciliary muscle is present but the muscles of the iris cannot be identified. The canal of Schlemm was not found. The iris appears to be turned backwards behind the lens and, as already mentioned, a bridge of vascular fibrous tissue passes between the free margins of the iris behind the lens.

Lens.—The lens is roughly spherical in shape and shows irregular projections here and there. No suspensory ligament can be identified. In the serial sections the lens disappears from the picture after the iris. The reason for this may be that the lens is abnormally large, or has prolapsed into the posterior chamber, or that there is a gap in the iris corresponding to that in the sclera and choroid. The superficial layers of the lens show some fibrillar

structure but the interior appears to be composed of structureless homogeneous vacuolated material. In other words there is a rind of more or less normal lens tissue with a degenerate cataractous centre.

Retina.—The pigment layer of the retina is opposed to the choroid from the ciliary body in the upper half of the eye round roughly one third of the circumference. The anterior half of the cyst is similarly lined. In the interior of the eye proper the retina is detached and crowded towards the lower anterior part of the posterior chamber (Figs. 1 and 2). In places there is some attempt at the formation of the normal layers but the rods and cones are not demonstrable. The detached retina is thrown into folds and it is difficult to be certain whether the layers of the retina are normally arranged or inverted relative to the choroid. In other parts the retinal tissue is completely disorganised. Where the choroid is absent within the cyst (*i.e.*, in the posterior two thirds of the sac) the retina is opposed directly to the cyst wall. The interior of the cyst contains imperfectly formed irregularly arranged retinal tissue. The ciliary part of the retina is detached and folded back behind the lens. On the side nearest the cyst this part of the retina is thrown into many complicated folds. Occasional masses of vitreous humour can be seen intermingled with retinal débris.

Comment

This was at first thought to be a case of pure bilateral microphthalmos but, as in so many of these cases, histological examination has demonstrated the presence of a cyst attached to the lower anterior part of the eye. The cyst is in fact quite small and was not obvious on macroscopic examination. It is quite common for congenital defects of the eye to be associated with deformities in other parts of the body. Swan *et al.* (1943), Swan (1944) and Gregg (1941) have pointed out the frequency with which congenital cataract is associated with congenital defects of the heart, and an abnormality of the limbs as an added defect is not uncommon (Mayou, 1904). Apart from abnormal topography the tissues are imperfectly developed. Vascularisation of the cornea has previously been referred to by Cruise (1905), the presence of hyaline cartilage in the sclerotic by Collins (1897), the hypertrophy of the ciliary body on the side of the eye next the cyst by Mayou (1908), and the failure of development of the rods and cones in the outer layer of the retina by Collins (1897). In many of the cases previously reported, the lens has been abnormal in shape, often tending to be spherical, with a degenerate centre, containing

vacuoles and inclined to prolapse. In this case the lens shows all these features and is undoubtedly cataractous.

Normally the foetal fissure in the human embryonic eye begins to close at the 11 mm. stage. In this case things must have begun to go wrong after invagination of the primary optic vesicle otherwise a congenital cystic eye would have resulted, no lens would have developed and differentiation of the retinal layers would not have occurred. Furthermore, defective development must have been initiated before closure of the foetal fissure. The critical period must have been between the 7 mm. and the 14 mm. stages, or in other words between the fourth and sixth weeks of intra-uterine life. Clearly, in this case, there has been a failure of closure of the foetal fissure at its anterior end. Through this gap the inner layer of the invaginated optic cup has protruded to form a small cyst. Mesodermal condensation has occurred round the deformed vesicle, including the cyst, to form a scleral coat, and from this a septum has grown backwards in an attempt to segregate the cyst from the cavity of the eye proper. This septum is incomplete and is partly composed of cartilage. The choroid is absent over the area of eversion. According to Ida Mann (1937), there is a close association between the presence of the pigmented layer of the retina and the development of the choroid. The everted portion (retina) represents the inner layer of the optic cup, no eversion of the outer layer (pigment layer) taking place. One would therefore not expect choroidal development between the sclera and the retinal contents of the cyst.

The condition is certainly not an ectatic coloboma. If it were there would be no fibrous septum partially separating the cyst from the cavity of the eye, and the cyst wall would be lined by an undifferentiated, flattened, atrophic neural layer arising from the apposition of the two layers of the optic cup.

The layer of vascularised connective tissue passing from the free edge of the iris behind the lens corresponds to the condition described by Terry (1945).

Developmental abnormalities of this kind are attributable to a variety of causes. It is common ground that, whatever the noxious agent responsible, it must act early in embryonic life—in fact, during the first six weeks of pregnancy. The abnormality may be intrinsic to the embryo, *i.e.*, due to a defect of the chromosomal or genic pattern, and this explanation is said (Ida Mann, 1937) to account for hereditary and familial abnormalities and often for sporadic cases also. Again the causal factor may be a toxin transmitted through the placenta from the maternal circulation. Lastly, mechanical pressure or trauma to the embryo

may be responsible. In this particular instance the abnormal influence must have acted so early that it is not conceivable that mechanical factors could have been responsible. The history of the pregnancy and the family history were investigated. The parents are perfectly healthy, normally developed people. They have had three children, including the child which forms the subject of this paper. No congenital defects are present in either parent or in the two surviving children, and there is no history of deformities of any kind on either side of the family. It appears, therefore, that this case represents a sporadic developmental defect in an otherwise healthy family. The mother has not previously aborted or been delivered of a premature child. During her last pregnancy her health was undisturbed. According to the work of Swan *et al.* (1943), Gregg (1941) and Swan (1944) there appears to be some sort of association between congenital defects in general, and of the eye in particular, and the occurrence of an attack of rubella in the mother during the early weeks of pregnancy. In most of the cases reported by these authors the child was born prematurely, was puny, difficult to feed and in many cases only survived its birth for a few days or weeks. The most frequent ocular defect found was congenital cataract, and the suggestion is advanced by Hurst (according to Swan, 1944), that, because of the association of congenital cataract and developmental defects of the heart, the toxin affects the embryonic vascular system and damage to the hyaloid artery is responsible for the cataractous condition of the lens. Damage to the eyes is unknown in rubella, but the theory is advanced that the virus responsible need not necessarily cause the same manifestations in the developing foetus as in the adult. In view of this work careful enquiries were made as to the possibility of the mother of this child having suffered from any of the exanthemata during pregnancy. There was no history of any sort of illness during this time. It would appear, therefore, that this case originated either from a genetic defect, or that the mother suffered a sub-clinical infection during the early weeks of pregnancy which passed unnoticed but left its mark on the foetus.

Summary

1. A case of microphthalmos with cyst is described.
2. The embryology and aetiology of the condition are briefly discussed.

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ADHESIVE EPISCLERAL REACTION IN THE OPERATIVE TREATMENT OF RETINAL DETACHMENT*

BY

L. WEEKERS

LIEGE

THE various procedures used in the operative treatment of detachment of the retina aim at causing adhesive choroiditis at a spot corresponding exactly to the place of the tear: the tear is to be engulfed by the area of the operative chorioretinitis and thus indirectly obliterated.

We believe that in addition to the choroiditis, one should endeavour to promote an adhesive episcleral reaction, as this contributes greatly to the immediate operative result and helps to preclude relapses.

1.—Experimental observations

We would first briefly recall certain experimental observations which we published previously: ¹²³

Fig. 1 refers to a simple but significant experiment. We used a cataract knife, considerably thinned by wear; the point was sharpened as much as possible so that the blade was but a thin flat needle, extremely sharp. After having instilled a few drops of cocaine, the point of the knife was driven into the eye of a rabbit at a point distant from the limbus, adjoining the *superior rectus*,

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