In the present case it seems not improbable, from a consideration of the position of the tumours, and their existence since birth, that they might have arisen in connection with the olfactory lobes, which possibly, as a result of some localised hydrocephalic condition, had protruded through congenital fissures of the skull, and gradually become constricted off with the closure of such defects.

Such a view at least provides the most rational explanation of the various features which have been detailed.

"INFANTILE AND CONGENITAL RETINAL FOLD"*

BY

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HOLLAND

RECENTLY, a number of cases of congenital retinal fold have been noticed. Some of them also showed anterior displacement of the optic papilla into the interior of the globe. Ancona and Kieuwe thought that this condition is caused by persistence of the connecting portion of von Szily's primitive epithelial papilla. Mann suggested that it is due to adhesion of the primary vitreous to the inner layer of the optic cup. Weve considered that the anomaly must arise through contraction of a mesodermal mass of connective tissue and that probably it is hereditary.

None of these writers found signs of inflammation, but Weve observed remnants of haemorrhage in one case. The essential cause of this curious abnormality remained a matter of conjecture and for this reason the following case is interesting.

The parents of a child three-and-a-half years old complained that 5 months before their child had developed a slight divergent squint and that the (right) eye looked strange.

On examination the eye showed no vascular injection, no exudates, the cornea was clear and the iris normal. The lens was turbid in its deep layers, especially at the centre. Behind the lens, temporally and below, a fairly well defined mass was seen against the retina. With dia-scleral illumination no shadow was observed. The intra-ocular tension was +1. Diagnosis uncertain. It was thought best to excise the globe.

Microscopical examination.—The globe measured 22 by 21.5 millimetres. These are normal figures (Fig. 1).

The cornea, anterior chamber and the angle of the anterior chamber were normal. The iris seemed a little more compact

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FIG. 1.

Infantile retinal fold. General view.

FIG. 2.

Retinal fold with delicate strand of connective tissue.

FIG. 3.
Retina showing obvious inflammatory changes.

FIG. 4.

Detachment of choroid by strands of connective tissue. Also note retinal fold.

FIG. 5.

Retina showing marked structural changes.

than usual and its surface seemed smoother, but otherwise no changes could be ascertained. In some places its posterior surface was slightly adherent to the lens near the pupil. The ciliary body and the choroid were quite normal, except in one place temporally and below. This will be described with the retina and vitreous. The lens was not displaced. It was slightly liquefied and vacuolated in the pupillary area of the anterior cortex. The posterior cortex contained very few vacuoles. Delicate fibres of connective tissue were attached to the posterior surface and ran towards the ora serrata.

The retina was completely detached. It showed two folds, one running from the optic papilla towards the pars plana of the ciliary body downwards and slightly outwards (Figs. 1 and 5) and a second just behind the ora serrata running round in a circular direction (Fig. 2). The crests of both folds were attached to strands of connective tissue. The strand adherent to the fold running from the optic papilla (Fig. 5) was thick and ran, like the fold to the ciliary body. The strands adherent to the circular retinal fold behind the ora serrata were delicate and stretched across the globe from one side of the ora serrata towards the other, passing behind and adhering to the lens. All these fibres joined behind the lens in a fairly vascular mass of connective tissue.

One may say that every structure to which these connective tissue strands were attached was displaced into the interior of the globe. The retinal folds have just been mentioned. The optic papilla was found 2 millimetres in front of the sclera, and the optic nerve therefore crossed a subretinal space. The choroid, close to the ora serrata temporally and below, was detached by a strong band of connective tissue joining the central mass (Fig. 4).

The retina showed disturbances of its normal structure. Rods and cones were often missing and the cells of other layers had partly lost their regular arrangement and normal staining properties. Many small spaces were seen filled with a homogeneous coagulum. Scattered leucocytes with segmented nuclei were interspersed between the retinal cells. The retinal veins were surrounded by closely packed lymphocytes. In most cases the vessel wall stained badly while in some cases only remnants of a lumen and of the endothelium were left. Generally speaking the layers of the retina were easily recognised. Close to the large fold, however, the retina suddenly exhibited complete disorganisation and a hole. In this region, below and temporally, the inflammatory changes were most marked (Figs 3 and 5).

The fluid filling the subretinal space contained cellular debris mostly derived from the neural epithelium, also coagulated material and various cells. This coagulated material had been organised here and there to form connective tissue staining red

with van Gieson. Some of the subretinal cells were not easily differentiated because of degeneration, but they were probably derived from the retina. Other cells, however, were undoubtedly leucocytes. This was proved by their finely granular protoplasm, their size as compared to cells inside the blood vessels and by the shape of their (generally segmented) nuclei. The inner surface of the retina was covered with small clusters of similar cells (Fig. 3).

The optic nerve traversed the subretinal space for about 2 millimetres. In its centre a longitudinal tissue-space was seen filled with coagulated fluid. From the optic papilla a strong fibrous band continued forwards (Fig. 1). Its course across the globe towards the ciliary body temporally and below has already been mentioned. This band contained many fibroblasts and cells derived from the retina. It exhibited circumscribed areas of disintegrated tissue (necrosis) and areas densely packed with lymphocytes.

It has already been mentioned that all fibrous membranes and also the large retinal fold joined close to the ora serrata. The fibrous tissue was superficially attached to the choroid which was partly detached (Fig. 4). Although its surface epithelium was disorganised the choroid itself and the pars plana of the ciliary body showed only a slight lymphocytic infiltration, but no inflammatory disturbance. In the angle between the ciliary body and the fibrous bands young delicate connective tissue was seen.

The retina and the connective tissue membranes contained scattered polymorphous leucocytes, especially along their surfaces (Fig. 3).

The central artery divided into several branches close to the optic papilla and was difficult to trace. No remnants of a persistent hyaloid vessel were found.

Discussion

The inflammatory changes were confined almost entirely to the retina and the vitreous body and for this reason it is not very difficult to reconstruct the pathological process. Retinitis, probably metastatic, was followed by vitreous abscess formation. This abscess then subsided and was organised. Fibrous tissue then contracted and pulled every structure to which it was attached towards the centre of the eyeball. Thus the retina, the optic nerve and the choroid were displaced. This could not have happened if the fibrous strands had been attached to the sclera, but it will be remembered that the attachments were superficial everywhere. It will be remembered too that every fold or crest was found to be attached to a band of connective tissue running towards the centre of the eyeball.

Inasmuch as this case closely resembles those described by

Ancona, Mann and Weve it must be decided whether or not its condition represents a stage preceding the condition of the other cases in the literature.

It is quite conceivable that slight and transitory inflammation of the inner layer of the optic cup might arrest normal development. In such cases remnants of inflammation might well be absent. Neither Mann nor Ancona definitely excluded this possibility, but as these writers found no signs of inflammation this problem is left unsolved.

Weve, on the other hand, seriously considered an inflammatory origin as his 8th case seemingly proved this definitely. He then rejected this theory again because the ciliary processes were free while a connective tissue membrane was attached to the pars plana of the ciliary body. A "cyclitic membrane," however, often arises from the pars plana of the ciliary body (*e.g.*, Beets). This case, therefore, was probably caused by cyclitis and the eyeball I described in the previous pages proves Weve's original idea to be correct.

Weve also argued that the majority of his patients were related to each other and such a relationship would render inflammation, of course, very unlikely. This familial occurrence, however, has not been sufficiently proved as only in Weve's 6th and 7th cases is the diagnosis above suspicion.

On the other hand it is very interesting to read that in both Weve's 6th and 7th patients and in the 1st and 4th the condition was observed in both eyes and in the outer and lower quadrant. Moreover, in all these eyes the fundus was pale and three out of these four patients were highly myopic (the refractive error was not mentioned in the first case).

It is tempting, therefore, to suggest a relationship between the high refractive error and the retinal fold, especially as the relationship between myopia and detachment of the retina is so well known in the adult. Moreover, van der Meer, in 9 out of 9 cases of detachment of the retina found a defect of the vitreous body close to the retinal tear. In old cases this defect was covered by a continuous layer of cells. Inasmuch as changes of the vitreous body are common in high myopia (*e.g.*, posterior detachment and sychysis) a lesion of the outer layers of the vitreous body in the very young seems quite conceivable. Regeneration, such as van der Meer found, might be expected to be very active in the very young. In this manner a considerable amount of tissue could be formed which, on contracting, would produce a retinal fold. Such a lesion would have to be close to the ora serrata. It is worth remembering also that this type of retinal fold was observed in the lower temporal quadrant and that "désinsertions" of the retina in the adult are found most frequently in

the same quadrant (Gonin). This theory would also perhaps account for a possible familial occurrence.

As the retinal fold was attached to the optic papilla it could, by exerting only slight traction, prevent the optic papilla from approaching the outer layer of the optic cup. In this way von Szily's primitive optic papilla could persist.

Conclusions

This paper deals with congenital and infantile folds of the retina. In the author's case the anomaly was caused by inflammation; probably retinitis; in Weve's 8th case probably by cyclitis of the pars plana of the ciliary body.

It is suggested that in Weve's 1st, 4th, 6th and 7th patients the condition be related to high myopia.

For all other cases an inhibition of normal development of unknown origin must be accepted; for this group a familial occurrence has not been proved.

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ANNOTATION

Blindness and its Causes

An interesting review of this subject was given at the Annual Conference of the National Society for the Prevention of Blindness at Columbus, Ohio, on December 4, 1936, by Miss C. Edith Kerby, the statistician of the Society. She began by emphasising the importance of an accurate analysis for two reasons. The first is for the sake of the individual, and the second for the sake of those who are planning work for the blind and prevention of blindness. She quotes the case of a man who was reported to an organization