A case of microphthalmus with upper-lid cyst : clinical report / by Charles H. May ; pathological report by Ward A. Holden.

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(*With six figures on Text-Plates XII. and XIII.*)

Clinical Report.

The patient, William Gordon, was born December 21, 1904; precipitate labor, full term; mother has tuberculosis; mother's eyes normal.

Present Condition (December 28, one week after birth): The right eye appears normal. The left eye presents a large cystic protrusion, pink in color, occupying the entire palpebral aperture and appearing to consist of the everted conjunctival surface of the upper lid. (Fig. 1, Text-Plate XII.) The cystic mass measures 18*mm* in width, 12*mm* in height, and projects 10*mm*. Upon puncture with a hypodermic needle there is an escape of a single drop of clear, yellow fluid, without any apparent reduction in the size of the cyst. The lids can be separated with difficulty by the aid of retractors, and when this is done it is seen that the protrusion is attached to the upper lid. When the cystic mass is raised with a lid retractor and the lower lid drawn down, a microphthalmic globe can be seen. The cornea measures 7*mm* in width and 6*mm* in height, is flattened, but perfectly transparent; the iris presents an eccentric, pear-shaped pupil, with the narrow extremity pointing upward and inward; there is a complete posterior synechia; the area of the pupil is grayish and opaque (cataract), so that no view of the interior of the eye can be obtained.

Five months later (May 3, 1905) the conditions were the same,

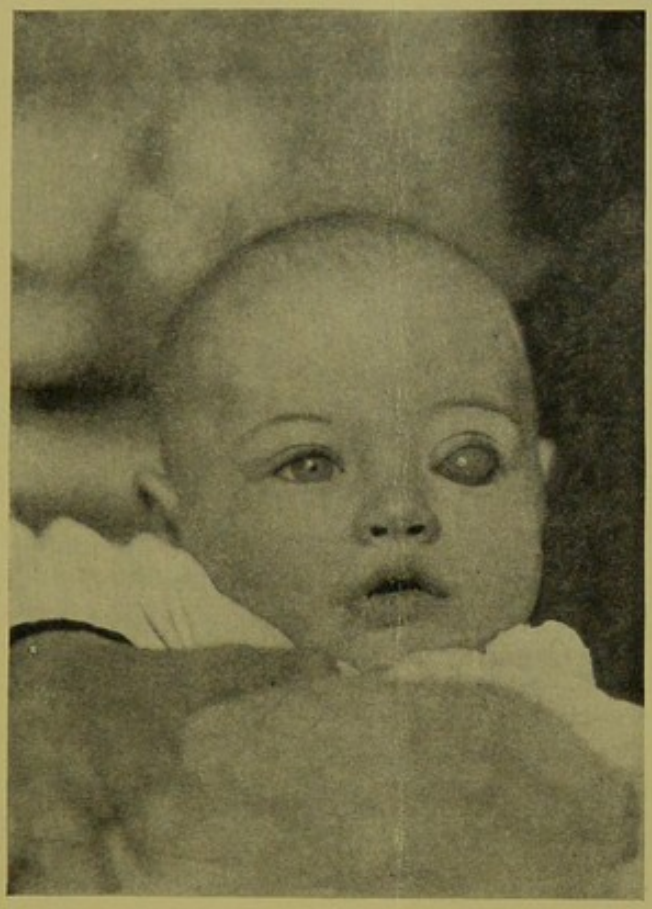

FIG. 1. Head of the child, with the cyst in the left upper lid.

FIG. 2. Upper-lid cyst. Natural size.

FIG. 3. The pedicle of the tumor, showing proliferating large cells at various points on its outer surface and numerous rosettes near these points ; Nissl stain.

excepting that the mass had grown slightly, and now measured 20*mm* in breadth, 14*mm* in height, and projected 12*mm*.

Operation, June 8, 1905.—Under chloroform anæsthesia, the conjunctiva covering the cyst was divided transversely and separated from the mass by blunt dissection. The separation was effected quite readily everywhere, excepting over an area about 4*mm* in diameter, where there was a direct connection between the mass and the eyeball. The situation of this attachment was practically at the equator of the eyeball and about 3*mm* above its horizontal meridian. This connecting portion was of dark color, and was found upon division to be formed partly of pigmented and partly of non-pigmented tissue. At the seat of the attachment a probe could be passed into the eyeball for a distance of 15*mm*, but it was impossible to determine whether between the tunics of the eyeball or into the vitreous.

July 19, 1906.—The microphthalmic eyeball has enlarged since the last note, and there is not a great deal of difference in the size of the globe on the two sides. The left cornea measures 8*mm* in the horizontal and 7*mm* in the vertical diameter; the right cornea measures 12*mm* in the horizontal diameter. The width of the palpebral aperture is 20*mm* on the left side and 24*mm* on the right.

Pathological Report.[1]

There was a small eyeball without coloboma of the iris and with a cataractous lens. From a point on its upper-inner surface, near the equator, issued a narrow stalk which expanded into a large tumor lying behind the upper lid (Fig. 2, Text-Plate XII.). This tumor was removed by Dr. May from the patient, a boy of six months, and since then the eyeball has grown until a year after the operation it is nearly normal in size. Lower-lid tumors are common enough, but the upper-lid tumors have been seen only a few times (Purtscher, Snell, Parsons, Collins.)

Development.—Evidently a knuckle of the secondary optic vesicle forced its way upward into the overlying mesoblast and, continuing to develop in extent, formed a mass of folded rudimentary retina which is surrounded by a fibrous sheath continuous with the sclera. The stalk is a tube of retina with a narrow lumen connecting the cleft-like cavities of the tumor with the vitreous

[1] Presented at the meeting of the American Ophthalmological Society, June, 1906.

chamber of the eyeball (Fig. 3). The lumen surface corresponds to the inner surface of the retina within the eye. The stalk is surrounded by a layer of pigment continuous with the pigment epithelium, and is enclosed in a fibrous sheath continuous with the sclera.

On passing from the stalk into the tumor proper the relations of retina, pigment epithelium, and outer fibrous coat are modified. There has been an invagination and a folding of the retina, so that what corresponds to the outer surface of the retina lines many of the clefts in the tumor as if it were the inner surface of the retina (Fig. 4, Text-Plate XIII.). The pigment epithelium surrounding the outer surface of the retina has mostly disappeared, and the fibrous coat and the retina pass one into the other without any line of demarcation.

But where a fold in the retina has allowed the fibrous sheath to pass into the central portion of the tumor, pigment epithelium has in several places been carried in also, and now lies in an anomalous position as regards the retina.

The rudimentary retina in this case is composed simply of a network of glia fibres and scattered nuclei, enclosing numbers of ganglion cells, large and small. There is no division into layers and there are no distinct outer and inner limiting membranes.

The feature of this case to which I wish to call attention is the presence of numbers of the rosette formations which are found in some gliomas and in some retinas of arrested development, which were considered neuro-epithelial formations analogous to the rods and cones by Flexner and by Wintersteiner, who first independently described them. Since then Ginsberg, Brown Pusey, and Verhoeff, among others, have studied these rosette formations. There has been considerable discussion as to the nature of their component cells. Their genesis and development are well shown in this case. Here the primitive neuro-epithelial cells, which compose the inner layer of the secondary optic vesicle, have become differentiated into spongioblasts and neuroblasts, which, attaining their full development, have formed, on the one hand, glia cells and fibres, and, on the other, ganglion cells. Of these elements, jumbled together without definite arrangement, this rudimentary retina is chiefly made up.

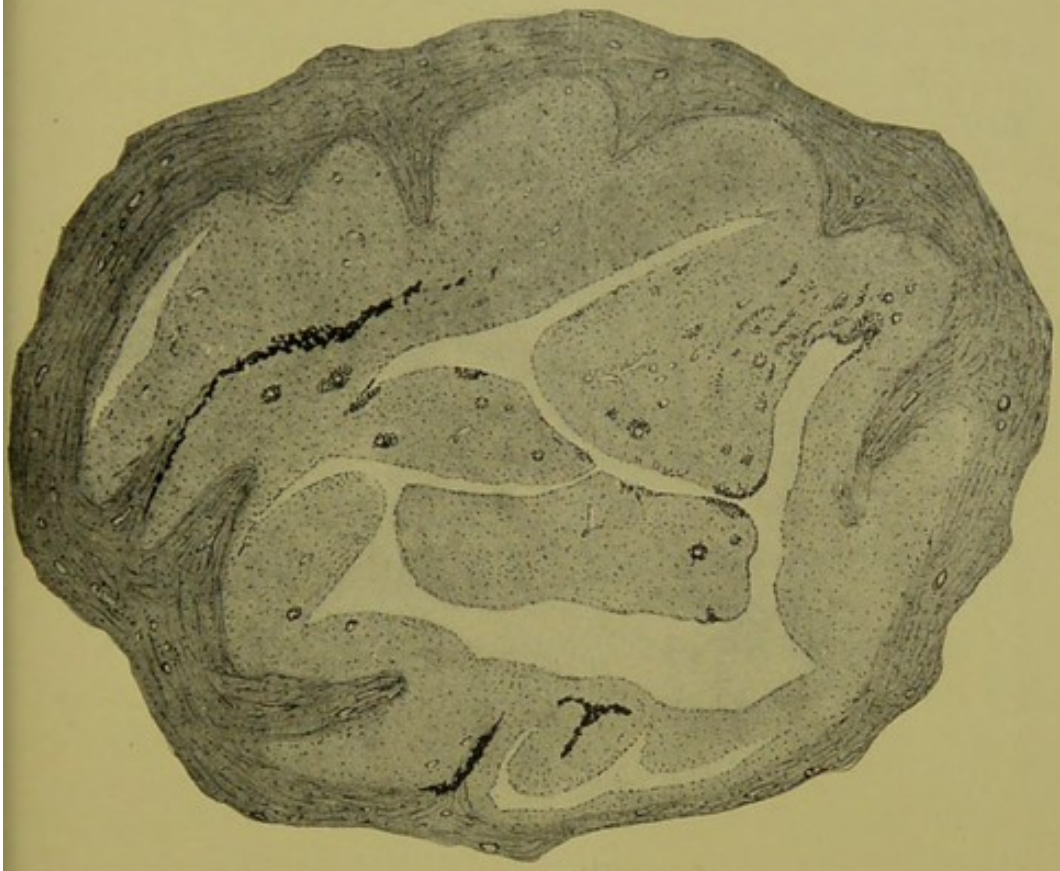

FIG. 4. A section through the middle of the tumor. The outer portion connective issue, the inner rudimentary retina.

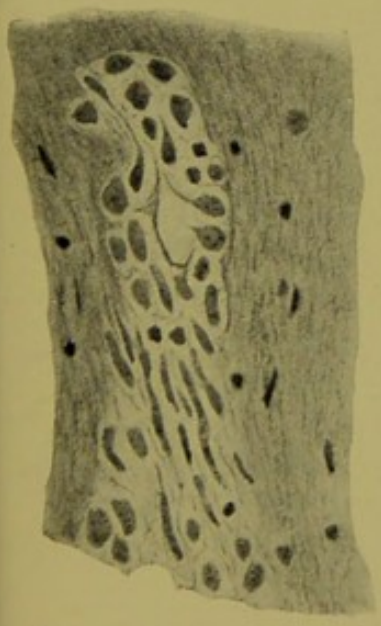

FIG. 5. Large cells growing inward from the outer surface of the retina to form a rosette.

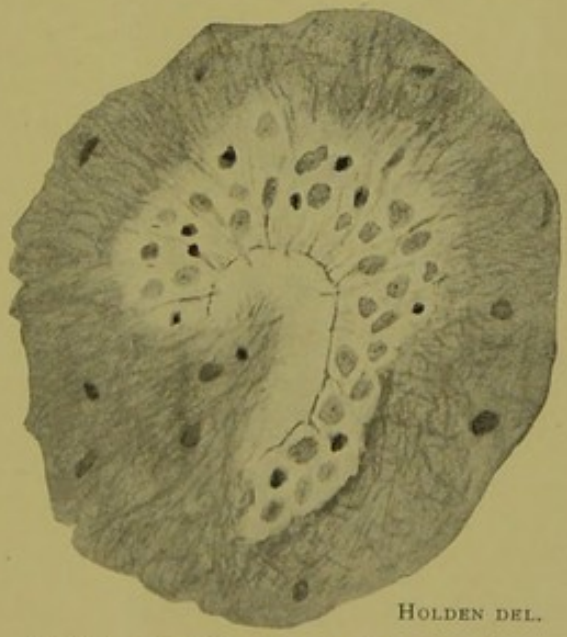

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FIG. 6. An almost complete rosette surrounded by ordinary glia tissue.

But here and there along the margins of the retina are single cells, or groups of cells, with large nuclei and large bodies which are to be considered embryonic retinal cells which have not yet developed. Now at various points on the outer surface of the retina these cells have proliferated, and, assuming a spindle form, have pushed perpendicularly into the retina like ependyma cells, and then have formed what in sections appear as spirals or as circles with a fine basal membrane, through which the large basal cells send filaments into the cavity (Figs. 5 and 6 Text-Plate XIII.). Rows of smaller cells, gradually assuming the characteristics of ordinary glia cells, surround the large basal cells, and the rosette is formed. The larger cells push into the neuroglia tissue without being directly connected with it, but the smaller peripheral cells in the rosettes send their processes out into the neuroglia with which they are in intimate connection.

The principal cells in these rosettes are seen to develop atypically from embryonic cells which in their natural course would have become rods and cones.