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Francis Delafield*

TUMORS OF THE RETINA.

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BY

FRANCIS DELAFIELD, M.D.,

OF NEW YORK.

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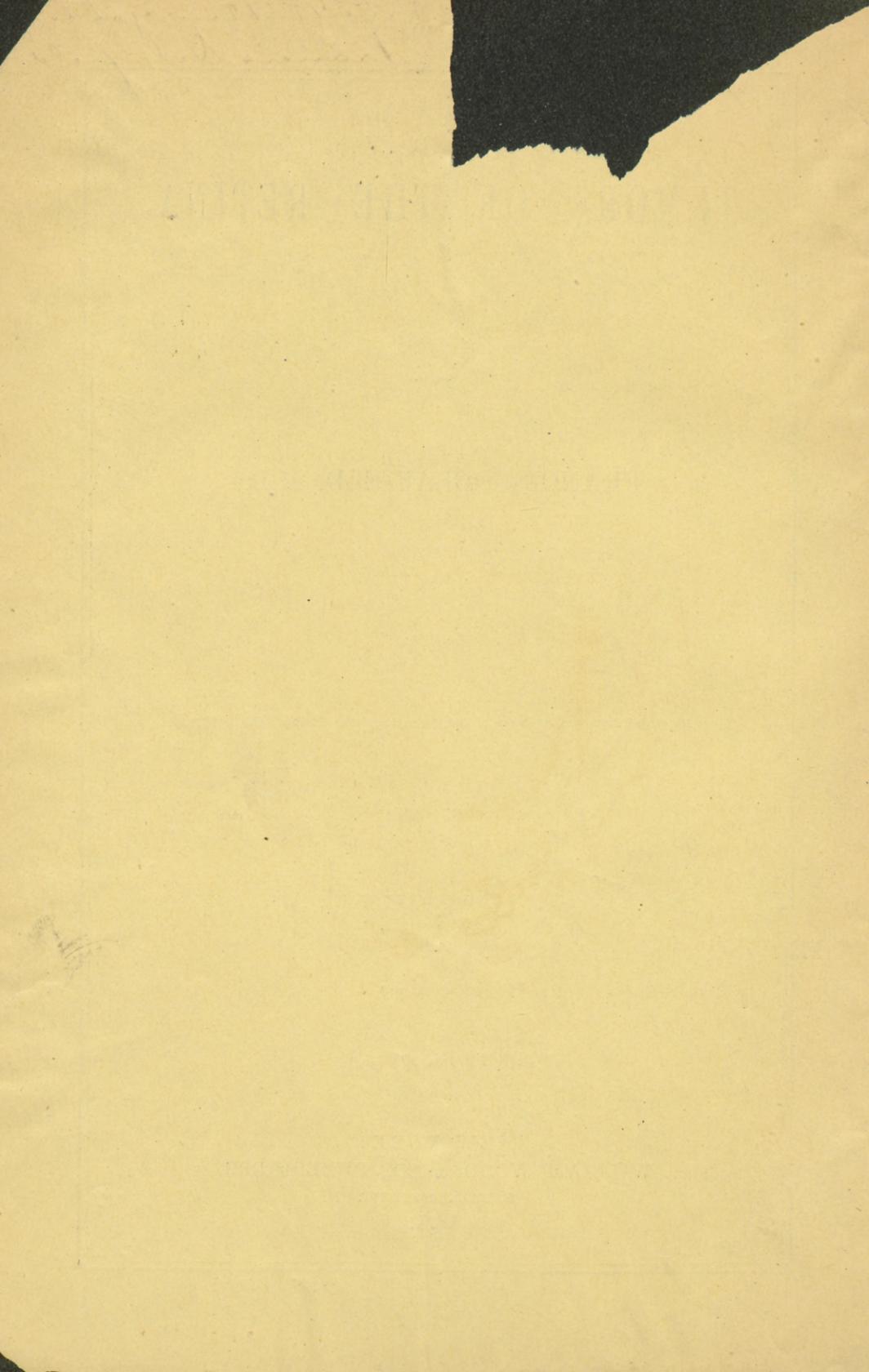
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TUMORS OF THE RETINA.

BY FRANCIS DELAFIELD, M.D., OF NEW YORK.

(PLATES IV. AND V.)

THE following cases of retinal tumors seem to be of sufficient interest to warrant their publication :—

CASE I.—The eye was removed by Dr. H. D. Noyes. It was taken from a girl 18 months old. Her father had first noticed a white spot in the pupil of the eye three months before its removal. The child had no symptoms of a return of the disease for two months after the extirpation. After that time the patient did not return.

The eye, while still warm, was opened by a horizontal section. The cornea, iris, and lens preserved their normal appearance and position. The cavity of the globe, posterior to the lens, was filled with a soft, whitish mass, stained red in some places by hemorrhages, and so soft as to be semi-fluid.

The elements of this mass, examined without the addition of any fluid (Hartnack, No. 9, immers.), were round cells, of very pale, finely granular appearance, exhibiting no nucleus until iodized serum was added, and measuring $\cdot 0116$ mm. Mixed with these cells were

amorphous granular matter, red blood-globules, and shrivelled round cells.

After hardening the eye in Müller's fluid, the tumor was seen to be continuous with the retina over about half its internal surface, and in apposition with it over the other half, the optic nerve being only covered by the tumor (Fig. 1).

Sections were then made of the tumor and retina.

The retina, where it is only in contact with the tumor, is well preserved; the *membrana limitans interna* and radiating fibres are thickened, the other layers are unchanged. In some sections portions of the tumor remain adherent to the internal surface of the *limitans interna*. In sections of that part of the retina which is continuous with the tumor it is found that the rods and cones and *membrana limitans externa* are everywhere unaltered. In other sections the new growth reaches only to the external intergranular layer.

At the line where the tumor ceases to be continuous with the retina the growth of the tumor is seen to take place in two ways.

First: as we follow the layers of the retina towards the tumor, we see round cells making their appearance in the layer of nerve fibres, and a little farther on in the layer of inner granules, the internal intergranular layer remaining intact. These cells increase very rapidly in number, rendering the layers in which they occur much broader, pushing the *membrana limitans interna* inwards, pushing the external intergranular layer outwards, the

cells in the two layers coming in contact with each other, until there is one continuous mass of cells extending from the limitans externa inwards (Fig. 2).

Second: we see round cells making their appearance in the same layers, and increasing in the same way; but the limitans interna, instead of being pushed inwards, and then disappearing in the tumor, is pushed inwards and then turned over outwards against the adjoining portion of limitans.

The retina is not detached from the choroid. The choroid and optic nerve present no traces of new cells.

Robin, Manfrédi, and Iwanoff each describe a case of retinal tumor, in which the new growth was developed inwards. In all the other cases reported the tumors are said to grow outwards towards the choroid. In this case the growth was very markedly inwards, for while the tumor was of sufficient size to fill the globe, the bacillary layer was everywhere perfect and in contact with the choroid.

As regards the precise point in the retina which gave rise to the tumor, we can only say with certainty that it was to the inner side of the external intergranular layer.

Nearly all authors describe the cells which make up these retinal tumors as if they were identical with the granules of the retina, and formed by hypergenesis from them.

The supposition that they are formed by proliferation of the retinal granules seems to be purely hypothetical.

The idea that the cells forming the tumor are identical with the retinal granules seems to be due to the practice

of looking at specimens altered by preservative fluids with low magnifying powers.

The layer of outer granules is said by Schultze to consist entirely of cells connected with the rods and cones. The rod granules consist of a globular, homogeneous nucleus, with small shining nucleoli, and an extremely thin cortex of a pale cell-substance, which is distinctly visible only at the upper and lower ends of the nucleus, and from which fine fibres are given off. Henle, Schultze, and Ritter also describe a peculiar striation of these granules.

Henle describes the inner granules as of two kinds: (1), globular, transparent nuclei, measuring $\cdot 005$ mm.; (2), the same nuclei enclosed by a scanty cell body.

The cells of this retinal tumor, examined fresh with No. 9 Hartnack, do not resemble any of these granules. On the other hand they resemble exactly the bodies commonly called lymphoid cells, which are found in so many new growths.

We can, therefore, leave on one side the question as to whether any of the retinal granules belong to the connective-tissue portion of the retina; for the elements of the tumor do not even resemble the granules, whatever the nature of the latter may be.

This case is one in which the prognosis should be favorable, as the disease was entirely confined to the retina. Unfortunately, the patient did not return after two months. For two months, however, there was no symptom of recurrence.

CASE II.—The eye was removed, by Dr. C. R. Agnew, from a child four years old. The disease of the eye was first noticed by the parents two years before its extirpation. During the last six months there was a great deal of pain in the eye, and the child's general condition became very bad. The child died within a year after the enucleation.

The eye was hardened in Müller's fluid. The cornea was transparent. The lens was pushed forwards, partly through the dilated pupil, into the anterior chamber.

The cavity of the globe was filled with a soft mass, mottled white and red. This mass was continuous with the optic nerve. Around the nerve the choroid was lifted up by a tumor of considerable thickness, and at the equator by another thinner tumor (Fig. 3).

The soft tumor, which fills the globe, evidently has its seat in the retina, leaving, however, no traces of the normal retinal tissue. The anterior portions of this tumor are composed of small round cells imbedded in an amorphous basement substance. The portion of the tumor near the optic nerve consists of larger cells, having a distinct cell body. These cells are contained in a delicate reticulum of connective tissue.

The retinal tumor is in contact with the entire internal surface of the choroid, and a thin white layer adheres in many places to the choroidal epithelium. The stroma of the choroid is thick and fibrous; the epithelium is thickened in irregular patches; many of the epithelial cells are greatly hypertrophied. In many places the epithelium is lifted up from the basement membrane by small, round cells, arranged either in little globular

masses, or forming a thin flat layer. Some of these globular masses of cells break through the epithelium, but none are seen to penetrate the basement membrane.

The stroma also is infiltrated with round cells, mostly arranged in long bundles. In the large tumors around the optic nerve there is nothing left of the choroid but its basement membrane and some irregular epithelium. There is, however, a distinct line of demarcation between the choroidal and retinal tumors. In all the sub-choroidal tumors the cells have a distinct cell body, and are contained in a reticulum of connective tissue.

The optic nerve was cut off close to the globe. It is slightly enlarged, and the bundles of nerve fibres are replaced by round cells, the fibrous septa between the bundles remaining intact.

CASE III.—The eye was removed by Dr. Carmalt, from a girl 4 years old. Ten months before its removal the eye became red and painful, and soon after the pupil gave a greenish reflex. Six months before the removal of the eye there could be seen, with the ophthalmoscope, a tumor the size of a pea, in the outer portion of the fundus. Within four months after the extirpation a tumor appeared in the orbit, and a few months later the child died. A tumor was found in the brain at the optic chiasm.

The eye was hardened in Müller's fluid.

The cornea, iris, and lens are unchanged. The anterior chamber is filled with fibrine. There is an episcleral tumor extending from the optic nerve to just in front of the equator.

The cavity of the globe is nearly filled with a soft white tumor continuous with the optic nerve. No retina can be seen (Fig. 4). Around the nerve the choroid is lifted up by flat tumors, extending on one side as far forward as the ciliary body. There are the same changes in the epithelium as were described in the last case. The subchoroidal tumors do not break through the basement membrane, even around the optic nerve.

The optic nerve was cut off at a distance of 23 mm. from the sclera. It is uniformly increased in diameter. The nerve fibres have entirely disappeared and are replaced by round cells. The fibrous septa between the bundles of nerve fibres preserve their normal arrangement, and in a few places contain oval and round cells. The inner sheath of the nerve is thickened and contains a few round cells; the outer sheath appears normal. There are no cells between the two sheaths (Fig. 5).

The episcleral tumor has the same structure as the subchoroidal growths. There are several small aggregations of cells in the sclera between the choroidal and episcleral tumors, but there is nowhere any regular cell communication between the intra and extra ocular growths.

CASE IV.—The eye was removed by Dr. Roosa, from a boy two years old. An opacity in the pupil of this eye was first noticed one year before its enucleation. Within four months the vision of the eye was destroyed, and the eye-ball began to protrude. A tumor situated in the upper and outer part of the orbit, and attached to the globe, was removed with it.

One month after the extirpation the tumor reappeared in the orbit. Soon after this the sight of the second eye was lost; there was hemiplegia of the left side, marked evidences of cerebral disease, and the child died in a short time.

The orbital tumor has the size of a pigeon's egg, is of ovoid shape, and is attached to the sclera.

The eyeball measures 30 mm. in its antero-posterior diameter, and between the tips of the ciliary processes 12 mm. There is an annular staphyloma in the sclera, between the corneal insertion of the iris and the ciliary body. The iris is carried forwards with the cornea.

The lens is transparent; it is held in place by the suspensory ligament, which is lengthened, thickened, and of a white color. The anterior surface of the lens is 2 mm. posterior to the plane of the iris.

There is a large white mass, extending like a pillar from the posterior surface of the lens backwards to the optic nerve. This mass does not touch the choroid, except at a point close to the nerve.

The internal surface of the choroid is covered by a thin white layer, composed of round cells. The epithelium is in many places lifted up by aggregations of round cells.

On the side of the nerve where the retinal tumor is in contact with the choroid, the latter is lifted up by a tumor reaching forwards as far as the equator.

The attachment of the episcleral tumor is over the sub-choroidal growth. The sclera between the two is thinned, and in some places infiltrated with round cells;

but neither the retinal, choroidal, or episcleral tumors are anywhere continuous with each other (Fig. 6).

The optic nerve is cut close to the globe, the nerve-fibres are replaced by round cells.

The new growth has the same characters as in the previous cases. The retinal tumor is composed of small round cells. The choroidal and episcleral tumors are composed of cells having a larger cell-body, and contained in a fibrous stroma.

CASE V.—The eye was removed by Dr. Mason from a girl $2\frac{1}{2}$ years old. Six months before the operation it was first noticed that the pupil of the right eye was enlarged. When the eye was removed the orbit was scraped and washed with ac. acetic. Six weeks after the operation the tumor recurred in the orbit. Afterwards tumors appeared on the temples and inferior maxilla; the vision of the other eye was destroyed, and death followed three months after the operation.

The anterior two-thirds of the globe has its normal shape, the posterior third is continuous with an irregular mass of new tissue.

The cornea is opaque; the lens is pushed forwards against the posterior surface of the cornea.

The cavity of the globe is filled with a tumor, soft and white at its centre, firm and gray at its periphery. The choroid and retina cannot be distinguished (Fig. 7).

The sclera is unaltered over the anterior two-thirds of the globe, over the posterior third it is lost in the tumor.

The central soft portion of the tumor is composed of small round cells, the peripheric portion contains in

addition a fibrous stroma. Where the tumor has replaced the sclera, it can be seen that the latter has been split up into layers of fibrous tissue separated by cells.

The optic nerve is surrounded by the extra-ocular tumor. There are groups of round cells between its two sheaths, and replacing the bundles of nerve-fibres.

The extra-ocular tumor is continuous with the posterior portion of the globe. It is surrounded by thickened connective tissue and by the ocular muscles. The muscles are not invaded by the new growth.

Remarks.—The last four cases exhibit the methods in which the new growth spreads from the retina to the neighboring parts.

The choroid, optic nerve, episcleral tissue, sclera, brain, and periosteum of the cranial bones become the seats of secondary tumors.

The optic nerve was involved in four cases, and in all presented the same changes. The nerve was increased in size, but was contained in its sheaths. The two sheaths were very little altered in any of the cases, and in only one case were there cells between the two sheaths. The bundles of nerve-fibres disappeared, and were replaced by round cells. The fibrous septa remained unaltered. The cells formed a continuous growth with the retinal tumor. It will be noticed that this description differs from that given by most authors, for they describe the new growth as advancing along the fibrous septa and sheath of the nerve, leaving the nerve-bundles untouched until the last.

In the choroid the secondary tumors occur by discontinuous infection. Separate foci are formed beneath the epithelium and in the stroma. Those beneath the epithelium increase in size until they break through it, but were nowhere seen to break through the basement membrane. Even over the large sub-choroidal tumors around the nerve the same small sub-epithelial tumors existed. In addition there were irritative changes in the epithelium, causing hypertrophy of its cells.

The tumors in the stroma also increase in size, pushing the basement membrane inwards. They reach their greatest size around the nerve, and come closely in contact with the retinal tumor; even here, however, the basement membrane is not broken through. In Case V. we can no longer distinguish between retinal and choroidal tumors, but it seems probable that there were sub-choroidal tumors which broke through the basement membrane, and became continuous with the retinal tumor.

That these discontinuous choroidal tumors were produced by cells from the retinal tumor seems probable, but not certain.

The secondary tumors in the sclera were of small size, formed of aggregations of cells separating the fibrous tissue. This cell-growth reached its greatest development in Case V., where the sclera is split into many layers, so as to form a large mass continuous with the posterior part of the globe.

The tumors on the outside of the sclera reached in Cases IV. and V. a considerable size. In Cases III. and

IV. these tumors were completely separated from the intra-ocular growths.

In Case V. there were metastatic tumors growing from the periosteum of the cranial bones.

From the study of these cases I venture to draw the following conclusions :—

1. The rule laid down by Hirschberg, that retinal tumors grow outwards towards the choroid, has many exceptions.

2. The elements of these tumors only resemble the granules of the retina when altered by reagents and seen with low powers. When examined fresh and with high powers, they are seen to be identical with the so-called lymphoid cells which compose so many new growths.

3. If we dismiss from our minds the superficial resemblance between the elements of these tumors and the retinal granules, they at once take their place in the class of round-celled medullary sarcoma.

4. The variation in the size of the cell-body and in the proportion of stroma in different parts of these tumors, is almost always found. This variation is the same which we find in most sarcomata, and is no reason why we should speak of a tumor as beginning as a glioma, and then becoming sarcomatous.

5. Not only the anatomy, but also the clinical history of retinal tumors corresponds exactly with those of the medullary sarcomata.

6. The development of secondary tumors follows the rule laid down by Virchow for sarcomata, and occurs :
(1) By continuous infection of the retina, optic nerve,

and, perhaps, the brain ; (2) By discontinuous infection, forming the choroidal, scleral, and episcleral tumors ; (3) By metastasis proper, forming tumors on the bones, lymphatic glands, and liver.

The idea that the choroidal tumors are formed by continuous infection from the retinal tumors, and that the scleral and episcleral tumors are formed by continuous infection from the choroidal tumors, seems to be erroneous.

In the very great majority of sections these tumors are evidently discontinuous. And, even if in some few instances a scanty cell communication is found between the two, it seems most probable, from what we know of other sarcomata, that this is a cell-growth posterior to the development of the secondary tumor.

That, however, these secondary tumors, although discontinuous, are produced from cells which have wandered from the parent tumor, is possible.

EXPLANATION OF TABLES IV. AND V.

FIG. 1.—Represents a horizontal section of the eye described in Case I. The tumor is seen to arise from about half the retina and fill the globe. The lens is pushed forwards.

FIG. 2.—Is a section of the retina at the point where it becomes continuous with the tumor, from Case I. The layers of inner granules and of nerve-fibres and ganglion cells become filled with round cells, and these cells become so numerous that they entirely replace all the layers of the retina as far as the *limitans externa*.

FIG. 3.—Is described in Case II. The cavity of the globe is filled with a tumor continuous with the optic nerve. On each side of the nerve the choroid is lifted up by tumors of some size.

FIG. 4.—Is described in Case III. The globe is filled with a tumor continuous with the nerve. On each side of the nerve the choroid is lifted up by small tumors. On the outside of the globe is a tumor of the same nature.

FIG. 5.—Is part of a cross-section of the nerve from Fig. 4. The neurilemma and fibrous septa are unaltered, while the nerve-fibres between them are replaced by round cells.

FIG. 6.—Is described in Case IV. The cornea and iris are carried forwards by an annular staphyloma, while the lens remains in place. A tumor, continuous with the nerve, extends from behind forwards through the centre of the globe. Beneath this the choroid is lifted up by a flat tumor, and the attachment of the episcleral tumor can be seen on the outside of the sclerotic.

FIG. 7.—Is described in Case V. The globe is entirely filled with the new growth, which has posteriorly split up and broken through the sclera so as to form a projecting tumor outside the globe.

FIGS. 2 and 5 were drawn from my preparations by Dr. Eno.

The other figures are from photographs of the specimens, by O. G. Mason.

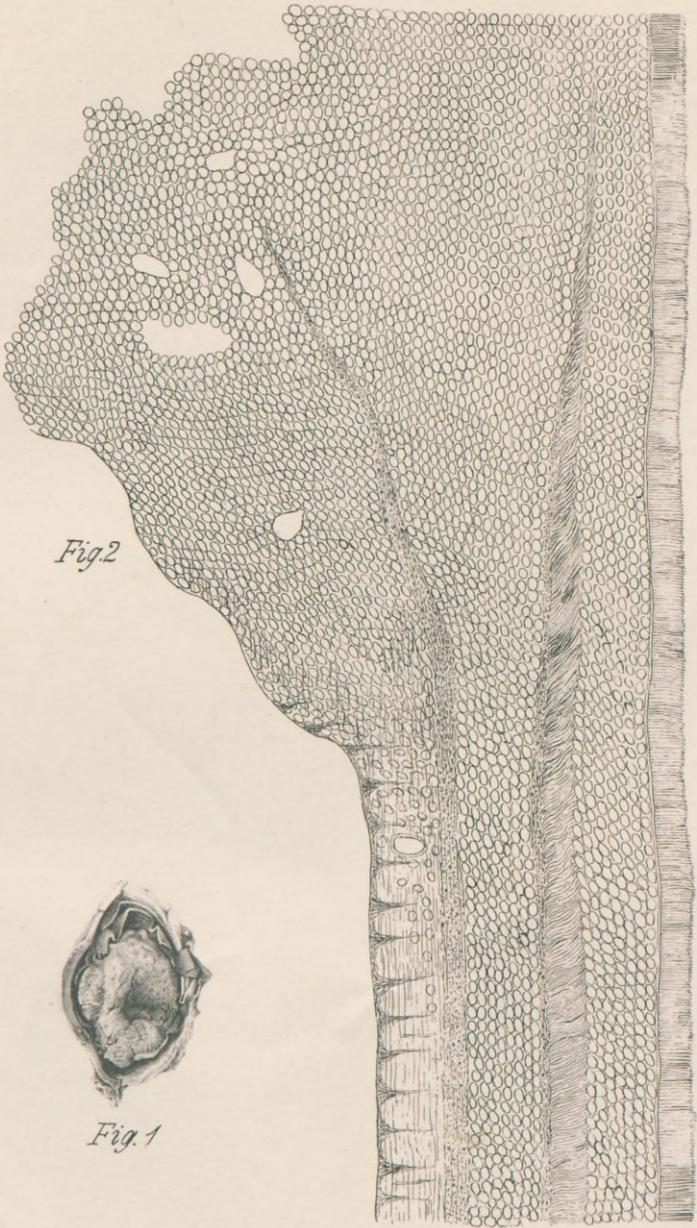


Fig. 2

Fig. 1

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Fig. 3.



Fig. 4.

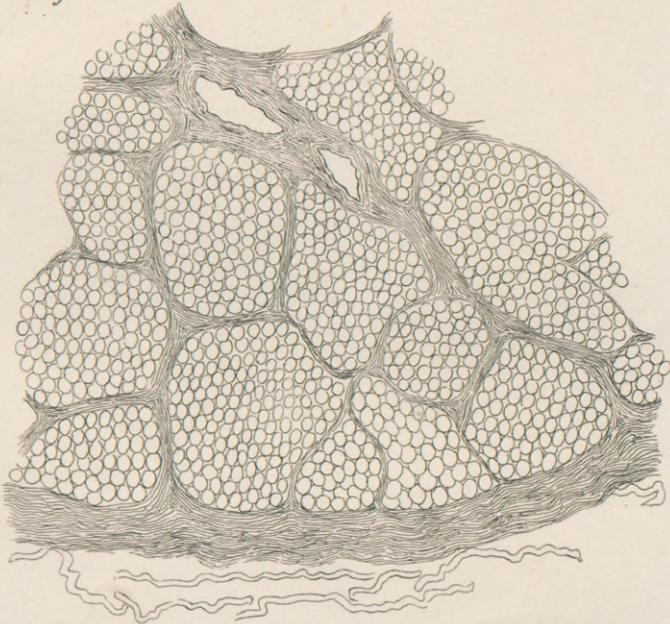


Fig. 5.



Fig. 6.



Fig. 7.

