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Optic Nerve.

By F. BULLER, M.D.,
MONTREAL, CAN.

[*Reprinted from American Ophthalmological Transactions, 1899.*]



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A CASE OF PRIMARY TUMOR OF THE OPTIC NERVE.

By F BULLER, M.D.,

MONTREAL, CAN.

In the course of twenty-three years' practice I have chanced to meet with three primary tumors of the optic nerve, two in children: a boy and a girl about six years of age, and one in a young adult female. The latter has not been reported as yet. These three cases have been furnished by a material of about 45,000 patients.

The present report is that of the boy six years of age, J. McN., who was brought to me in June, 1892. *I have not been able to learn the ultimate result in this case, but hope to do so before this report is published. The history shows that the growth was probably of congenital origin: no other member of the family, nor any relative on either side of the family was ever known to be similarly affected. The history is as follows:

The child is apparently in fairly good health and well nourished. When about three months old, the right eye was thought "to be set," as the parents expressed it. There was probably some fault in the position of the eye, as the family physician is reported to have said he could "cut the cord and make it all right." No intervention of this kind, however, was permitted. The enlargement of the eye was noticed soon afterwards, and progressed very gradually until the age of four years, when the eye became rather suddenly much more prominent.

About this time he had a severe attack of general convulsions. During one of these convulsive seizures he was for a time unconscious. Since that illness he has had no actual convulsions, but is subject to attacks of fear or terror, and swallows frequently at such times: he does not fall or lose consciousness. These attacks are supposed to be brought on readily by errors of diet. In other respects his general health is good. The left eye is normal. The right eye presents a high degree of proptosis, being pushed forward approximately half an inch or more, but is little,

if at all, restricted in its movements. The edges of the lid rest almost on the equator, and at no time fully cover the eyeball. This exposure causes a constant redness and irritability of the eye, evidenced by pericorneal congestion and a slight marginal keratitis, but corneal sensation is good. The pupil is dilated 5mm. and inactive to light. Media clear enough to permit a perfectly good view of the fundus. Although the child is restive and difficult to manage, the ophthalmoscope showed the optic nerve entrance apparently much larger than it should be, of a pale, bluish white tint, with oval outline, and frayed-looking margins. The retinal vessels are very small, but cannot be studied in detail.

Palpation reveals within the orbit a large somewhat elastic mass without distinct outlines, situated quite behind the eyeball.

The complete blindness, the expanded appearance of the optic nerve, and the character of the proptosis, all pointed to the presence of a tumor involving, if not originating in, the optic nerve, and the removal of the growth was advised.

The operation was performed under anæsthesia by ether. The internal rectus being freely divided, secured with a suture, and drawn out of the way, the growth was reached and found to fill the apex of the orbit completely. It was detached with scissors as far back as possible, turned forward and separated from the eye. After this had been accomplished, a considerable mass of soft material was still recognizable at the orbital apex, the removal of which involved much disturbance of the surrounding tissues, as well as a rather copious hemorrhage, on account of which all idea of saving the globe had to be abandoned. After removal of the globe and tumor, chloride of zinc paste was applied in the usual way to ensure complete destruction of any remaining tumor substance at apex. The patient made a good recovery, and was able to return home at the end of two weeks. One month after the operation, he was reported to be in excellent health and quite free from nervous attacks, the orbital cavity only discharging very slightly.

The growth when removed was found to be 10 x 7 millimeters in diameter at its attachment to the eyeball. At this point

the nerve appears as a small circle in the center of a yellowish, gelatinous-looking, rather firm mass. The portion at first removed constitutes about $2/3$ of the entire mass, that which was taken away after removal of the eye the remaining third, this portion being concave anteriorly and perforated at its center. This disk-like segment when fitted over the other completes the growth.

In separating the parts with the scissors, they must have passed into a sulcus in the soft tissues, and so divided the tumor into the two portions just described.

The chief interest in this case is the pathological report prepared by my friend and colleague, Dr. Byers, and herewith submitted *in extenso*.

I wish particularly to emphasize the apparent, sudden increase in the proptosis at the age of about four years. This seems to coincide with the observations of Dr. Byers in reference to the œdematous condition he describes in the extraocular lymph-spaces, and which he considers a prime factor in the production of the proptosis. This and the whole subject of primary tumors of the optic nerve will be treated in greater detail in a paper of Dr. Byers now in course of compilation.

PATHOLOGICAL REPORT.

By W. GORDON M. BYERS, M.D.

[From the Pathological Laboratory of the Royal Victoria Hospital.]

The eyeball and tumor in two pieces detached from the globe were handed to me in Müller's Fluid.

MACROSCOPICAL EXAMINATION.

The anterior and larger portion of the tumor was composed first of all of a 15 mm. long piece of enlarged optic nerve having a transverse diameter of 12 and a vertical diameter 10 mm. The whole was encapsuled by two distinct fibrous sheaths. The inner of these was thin (less than 0.5 mm.), smooth, dense, and loosely



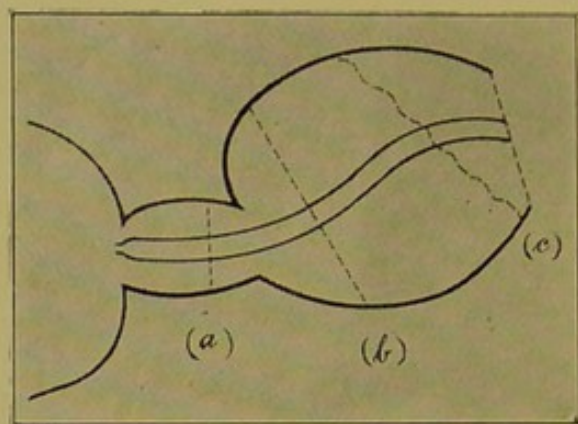


FIG. 1.—Purely diagrammatic to show points of section *a*, *b*, and line of cleavage (*c*) between two portions of tumor.

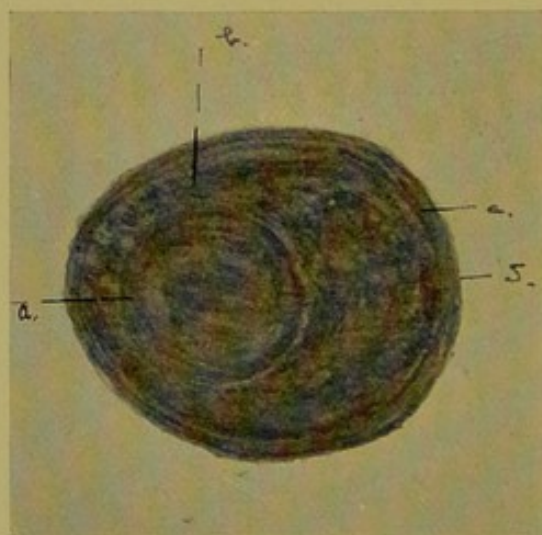


FIG. 2.—(*a*) optic nerve. (*b*) intervaginal overgrowth. (*c*) dural sheath. (*d*) connective tissue sheath external to this.

connected with the outer surface of the spongy layer described below. It was everywhere surrounded by the outer, looser, shaggier sheath to which it was likewise connected by delicate adhesions. This also had a thickness of 0.5 mm.

Viewing the anterior somewhat rounded (from retraction of the outer sheath) cut surface of this piece, the optic nerve proper, 3 mm. in breadth, could be seen occupying a nearly central position, and surrounding this a layer of somewhat spongy tissue which varied in thickness from 3 to 4 mm.

Transverse section at (A, Diagram 1) 8 mm. behind this end (sketch 2) showed in the main the same arrangement as on the anterior cut surface. The chief differences were in the optic nerve proper, which here obtained a breadth of 6mm. in both the vertical and transverse meridians. It was also more eccentrically placed owing to the irregular distribution of the sub-vaginal tissue. This had on the nasal side a thickness of about 4 mm., whereas on the temporal side it measured only about 1 mm. in breadth.

Fifteen millimeters from the globe the tumor made a sharp bend upon itself and became greatly increased in size. It here had a diameter of from 22 to 24 mm.

On transverse section (B, Diagram 1) of this portion (v, sketch 3), the increase in size was seen to be due mainly to a further increase in the sub-vaginal tissue, which had a breadth on all sides of from 6 to 7 mm.; and to a marked thickening of the outer fibrous sheaths, C and D, which were still everywhere intact, but more firmly adherent to one another and to the substance of the enlargement. The optic nerve proper at this point measured the same as further forward, in other words, 6-7 mm.

On the upper and posterior surface of this portion was a large, irregular gap in its continuity. Into this fitted the remaining tumor bit which had been evidently torn away during the operation. The end of this portion which formed the posterior cut surface of the tumor was dense and firm, apparently from great thickening of the outer sheaths, and formed a firm, rubbery collar around the optic nerve, which here measured 5 mm. in the vertical by 6 mm. in the transverse diameter.

As nearly as could be made out, the course of the optic nerve throughout the enlargement (and hence more or less of this as a whole) was at first backwards and nasalwards, then upwards and slightly towards the temporal side, and finally backwards to the optic foramen. On transverse and longitudinal section it was seen that the optic nerve maintained throughout its whole course practically the same dimensions which it attained after leaving the globe, in other words, 6-7 mm. Only in its last backward course was it less clearly demarkated from the surrounding tissue, but even here its size could be made out with considerable exactitude. The pial sheath seemed to merge at every point in the sub-vaginal overgrowth.

TEASED SPECIMENS.

Teased Preparations. The teased specimens showed cells of the type of embryonic connective tissue. Some of these were bipolar (a, b), and the terminal filaments, often long and wavy, were given off from an oval or spindle-shaped body. In others, the processes were numerous, and the cell-body more or less irregular in shape (c, d, e). Between these two types were numbers of intermediate forms (*e. g.*: f, g). These cells contained a rounded or oval nucleus, which was centrally or eccentrically placed, and stained deeply with hæmatoxylin. Under the microscope were seen, also, a large number of mononuclear cells, unprovided with a termination of any kind (H), but these were not confused with the numerous oval or spindle-shaped bodies (I), which seemed free only because of the detachment of their delicate processes.

OPTIC NERVE.

The nerve bundles varied little if any in regard to size and shape from those of a normal section. The enlargement of the nerve proper noted macroscopically seemed at first sight to be due to a marked increase in the perineurium, but further examination showed that the chief cause of the increase was not so much a hyperplasia of the connective tissue as a pressing apart from one another of the individual septal fibers. The areas between the nerve bundles were filled by a loose branching mesh-

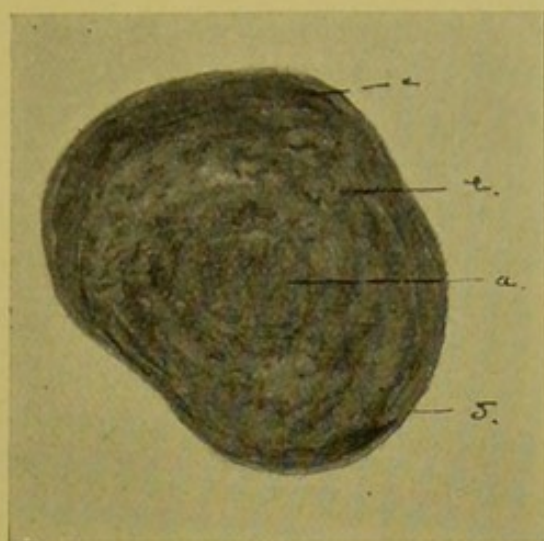


FIG. 3.—(a) optic nerve. (b) intervaginal overgrowth. (c) dural sheath. (d) connective tissue sheath external to this.

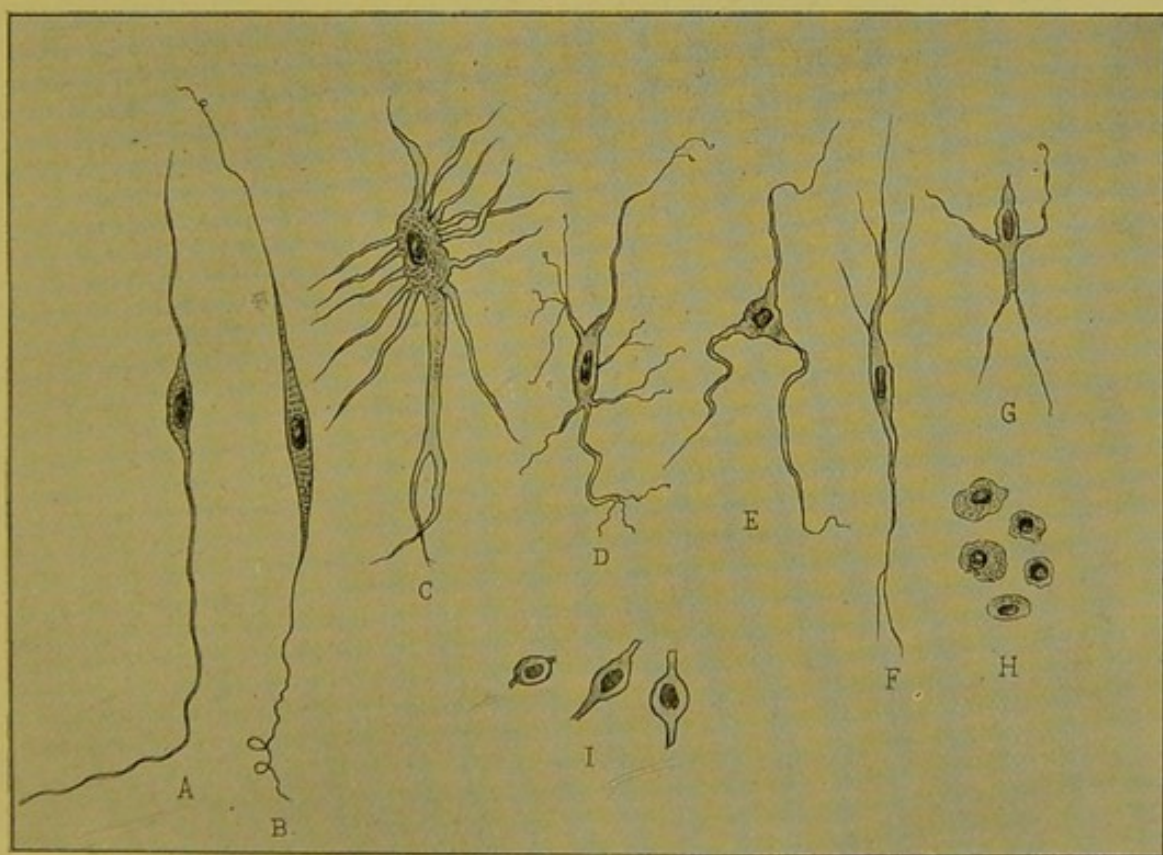
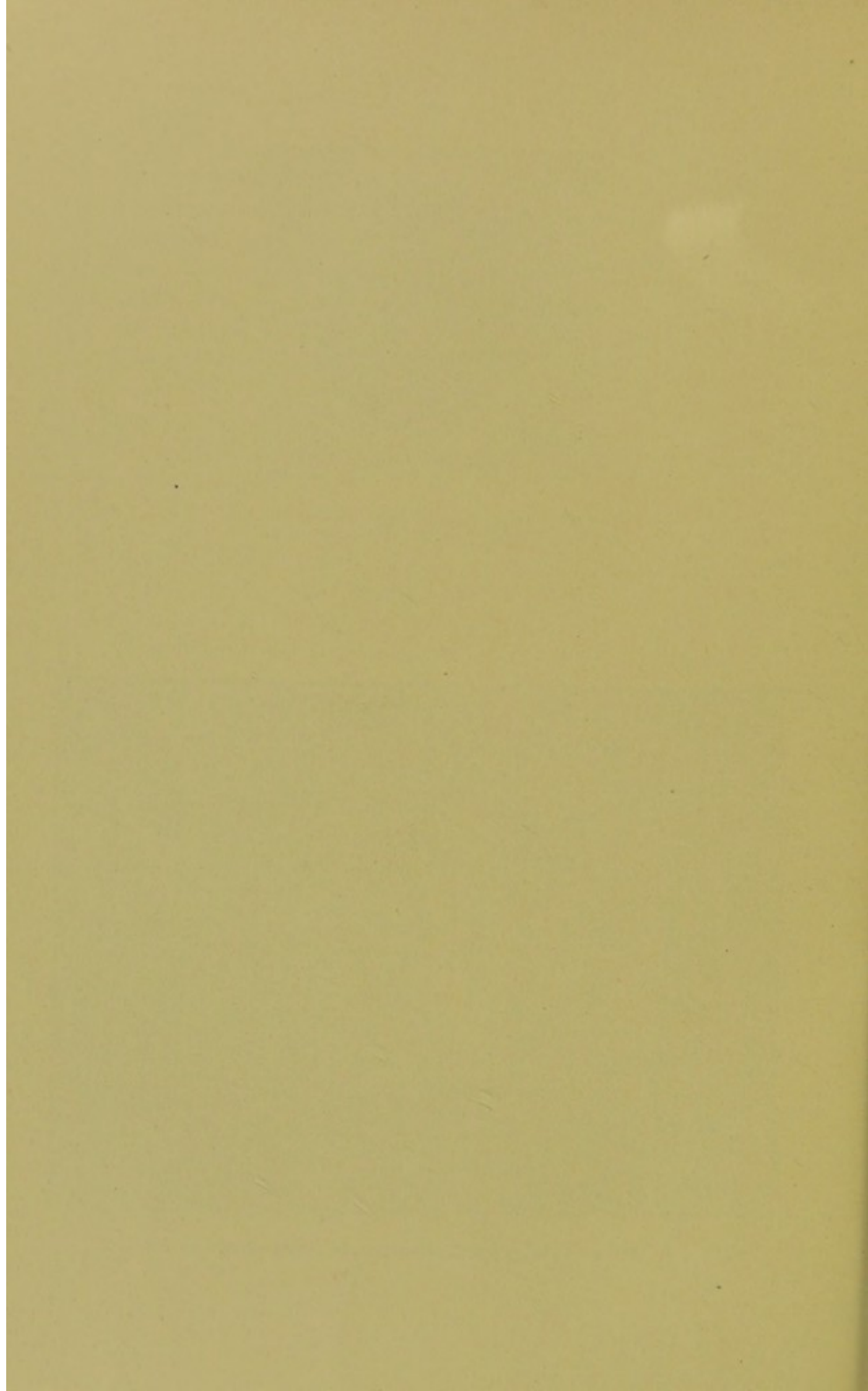


FIG. 4.— Showing types of cells found in teased preparation.



work of connective tissue in which were seen a large number of rounded or oval nuclei staining deeply with hæmatoxylin. Another striking feature in the nerve proper was the presence of large, clear spaces, which existed everywhere throughout its whole extent, between the outer surface of the nerve bundles and the inner surface of the peri- and epi-neurium. These were of irregular shape, and surrounded the bundles in their circumference to a variable extent.

Both the foregoing findings, though made out in celloidin specimens, were especially well seen in thin paraffin sections. Dr. Shirres, neuro-pathologist to the hospital, kindly stained specimens for nervous elements, but we were unable to find traces of axis cylinders in any part of the nerve. The parenchyma in some places, apparently normal, in others and very commonly, was represented only by the endoneurium which formed a honey-combed meshwork, the spaces of which seemed entirely free from tissue substance of any kind. This condition was perhaps more marked in the periphery of the bundles, and was only well made out in thin sections.

Evenly distributed throughout the whole nerve substance were rounded or oval nuclei, which were generally situated over or in the endoneurial fibres, but sometimes appeared attached to the side or lay free in its center.

MICROSCOPICAL EXAMINATION.

In the periphery of the sections were seen densely packed, often wavy, bands of connective tissue corresponding to the outer fibrous sheaths noted macroscopically. These ran for the most part in a concentric manner around the whole of the section, the innermost layers being involved in the changes described below.

The area between the optic nerve proper and the outer sheaths was occupied by a flowing, irregular, branching, network of connective tissue strands, which stained deeply with eosin. The spaces of the meshwork were occupied by large numbers of cells with rounded, oval, or elongated, deeply staining nuclei, and fine processes, which branched or interlaced in an irregular way.

Sometimes the cells had a stellate appearance and the picture was then characteristically myxomatous. At other times the spaces were filled with large numbers of more or less rounded closely packed cells which had an almost epithelial appearance. These masses occurred especially in the region corresponding to the inter-vaginal space, and here the cells often showed signs of degeneration, becoming granular, ill defined, and difficult to stain. In places they formed merely areas of amorphous debris, and here and there clear spaces were seen which had evidently arisen from their complete disintegration — cystic formation.

The individual fibers of the connective tissue strands were often pressed apart from one another much in the same manner as was described in connection with the perineurium. The cleavage spaces thus formed were occupied by the myxomatous cells, and by delicate processes, which extended respectively between the different septa and the individual fibers which composed them.

The pia mater and especially the arachnoid sheath were the structures mostly involved in the changes described above, but the dural sheath in its inner layers was also affected, and this structure was not therefore cut off from the general process.

EXAMINATION OF GLOBE.

The measurements of the eyeball were as follows: Antero-posterior, 20 mm., transverse, 22 mm., vertical, 21 mm. The nasal side of the globe was smooth and shiny, but the whole of the temporal half was covered by a loose layer of tissue which blended forward with the conjunctiva. Behind, at from 5 to 8 mm. from the optic nerve entrance, it became adherent to the sclerotic which was thickened in the same way all over the posterior pole of the eyeball. The external surface of the covering was shaggy, but the internal smooth and shining, and but loosely connected with the surface of the sclerotic. The sac thus formed could be dilated with water, and when filled the appearance was as if a large bleb covered two-thirds of the temporal side of the globe.

The superior, inferior, and exterior recti muscles pierced this covering forwards and received reflections from its substance.

Near the posterior pole of the globe was a distinct pit $5 \times 5 \times 2.3$ mm. in depth. In the bottom of this about its center was the optic nerve of normal size, and above this a conical-shaped piece of tissue in the inter-vaginal space.

The marked pitting was chiefly due to the extensive thickening of the posterior pole of the globe in the way described above.

MICROSCOPICAL EXAMINATION.

At many points, the surface of the sclerotic was covered by a loose mass of curling, intermixed, elastic, and connective tissue fibers filled in with granular debris, a few leucocytes, etc. At a few points, towards the surface, the sclerotic showed signs of œdema, and its vessels were distended.

There is marked enlargement of the optic papilla. The retina, ciliary body, and iris were also marked by moderate œdema, and in the two latter was an excessive proliferation of pigment cells. The retinal pigment layer of the iris was greatly thickened. The sub-conjunctival tissue showed very marked signs of œdema and an infiltration of small round cells in places.

SYNOPSIS OF REPORT.

The following then are, in brief, the pathological findings:—

Moderate increase in the size of the anterior 15 mm. of the optic nerve, and great enlargement along the rest of the structure.

The specimen everywhere contained by the dural sheath and a second connective tissue layer, in all probability the orbital septa bounding the supra-vaginal space.

The optic nerve, uniformly enlarged, was followed throughout the whole length of the tumor, which was principally due to an overgrowth of tissue in the sub-vaginal space.

The enlargement of the optic nerve was due to a marked separation of the fibers of the connective tissue between the nerve bundles; and this was associated with clear, irregular spaces present everywhere between the outer surface of the nerve bundles and the inner surface of the peri- and epi-neurium. The axis cylinders completely atrophied.

The temporal half of the globe was covered by a portion of Tenon's capsule; over the whole of its surface were signs of an œdema, which was present also in the retina, ciliary body, iris, and sub-conjunctival tissue.

REMARKS.

This tumor agrees in its microscopical details with what former writers have described as fibro-myxomatous or myxo-sarcomatous growths. It might, perhaps, be placed with reason in either of these categories. But the changes in the optic nerve, at least, are to be looked upon as due to a chronic distension of the lymph spaces of that structure; and it is just possible that we have here in general to deal with some localized disturbance of the lymphatic system.

A point which seems to have been generally overlooked in connection with these growths, is the part played by the extra-ocular lymph spaces in the production of the principal symptom of the disease, namely, the exophthalmos.

As a result of the tumor growth, occlusion of the supravaginal space is brought about, and, as a result, a passive distension of it (varying with the extent to which it is filled by the tumor) and of Tenon's space occurs. In this way the development of the proptosis is augmented.

In conclusion, it is a pleasure to acknowledge the advice and kindness which I have received from Dr. Adami in the preparation of this report; and to thank my brother, Mr. Roddick Byers, student of medicine, for the execution of the drawings.

DISCUSSION.

DR. REEVE. — I would like to ask Dr. Buller whether the sight was lost in the other two cases he referred to. Last summer I saw an interesting preparation in the Birmingham Ophthalmic Hospital with preservation of excellent, if not normal, vision.

DR. BULLER. — There was no sight in this eye, and in the other two cases the ophthalmic picture was entirely different. In neither was there any sight. I consider the early loss of vision with monocular optic neuritis one of the diagnostic features of tumor of the optic nerve.

DR. REEVE. — That is the point of interest, because it was not the case in the interesting specimen I saw.



