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COMMUNICATIONS

TWO CASES OF TUMOUR OF THE OPTIC NERVE*

BY

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TUMOURS of the optic nerve are of such rarity that it seemed that the two cases to be described might be reported in detail.

Case I

Rowland, O., aged 14, was seen by the writer at the Central London Ophthalmic Hospital, in February, 1921.

History: The mother stated that she had noticed during 1915 that the left eye looked larger than the right. Gradually this feature became more noticeable. The boy had always enjoyed good health. There was no pain. There was no history of aural or nasal discharge. Examination on 9/2/21 was as follows:— V.R.=6/6, J1 at 16 inches. Manifest hypermetropia 0.5 D. V.L.=shadows only. The pupils were equal and active; tension normal, R. and L. There was proptosis of the left eye to the extent of 18 mm. as measured from the mandibular joint to the plane of the anterior corneal surface. (R.=82 mm., L.=100 mm.)

Movements of the eyes were good in all directions. The right fundus, disc and macula were normal. The left disc was pale and of a woolly appearance, with no distinguishable physiological cup, and no appreciable swelling. Large choroidal vessels were

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visible in both eyes, but more apparent in the left. In the left there were also several very large choroidal vessels which passed to the margin of the disc and there disappeared.

Mr. Norman Patterson examined the boy and reported that there was nothing in the nose or its surroundings to account for the exophthalmos. X-ray examination at the London Hospital revealed no abnormal shadow in the orbits.

As the proptosis continued to increase, it was decided in October, 1921, that an operation should be performed.

Operation: October, 1921. The external canthus was divided horizontally, the incision being carried to the orbital margin. The conjunctiva was turned forwards and the external rectus

CASE I. FIG. 1.

Stereo-photograph of patient, Case No. 1, eight months before operation.

muscle was exposed. This muscle was divided between two sutures and the eyeball drawn forwards. The orbit was found to be occupied by a mass behind the eyeball. Dissection revealed that a small portion of the optic nerve was apparently uninvolved immediately behind the eyeball. A ligature was passed around this and used as a retractor. The orbit was found to be completely filled by a mass of elastic consistency. It was not found possible to distinguish the superior and inferior recti muscles. The mass was separated from the walls of the orbit by blunt dissection. Curved scissors passed behind the mass were used to cut through its posterior limits at the optic foramen. The mass was then drawn forwards and removed from the orbit, and the optic nerve divided close to the eyeball. Some gummy material escaped from the mass when division was made at the back of the orbit.

The external rectus muscle was then sutured and the wound drained and closed. The edges of the lids were pared and the lids sutured together.

Within twenty-four hours there was very considerable proptosis and swelling of the eyelids. Only a very moderate amount of blood drained along the tube, which was therefore removed. During the next few days the proptosis remained in about the same condition, and the tension upon the sutures in the eyelids was so great as to cause them to cut through the tissues. The cornea was thereby to some extent exposed and quickly developed some opacity in its lower part. The surface stained strongly with fluorescein. By the passage of fresh sutures the lids were made to cover the eye to some extent for a few days longer. Gradually the proptosis lessened, but the corneal condition remained such as to cause considerable anxiety. In the course of about a fortnight some diminution of the extent of staining of the cornea was noticed. Very gradually the ulcerated surface healed, leaving a moderate degree of opacity in the lower half of the cornea. There was complete loss of sensation in the cornea, and this has persisted to the present time. As was to be expected, owing to the size of the orbital mass and the somewhat blind division of the orbital contents, the action of the levator palpebrae was lost. There has been no sign of return of function in this muscle, but a small degree of elevation of the upper lid is now possible through the action of the frontalis muscle. The eyeball took up a position of abduction and was devoid of power of movement. The position of the eyeball remains the same at the present time. The cornea now has slight opacity in its lower third, which is not sufficient to obstruct a fair view of the fundus. There has been no return of sensation in the cornea. But the fact that the upper lid remains paralysed has presumably prevented any fresh development of corneal ulceration. The iris shows marked atrophy, so much so that in places a faint red reflex can be seen through it when the fundus is examined. The optic disc has gradually passed to the stage of optic atrophy, and now is almost dead white in colour with some white areas of choroidal atrophy around. The retinal vessels, from the time at which they were first visible after the operation, have maintained their colour. Some abnormal vessels are present which disappear at the disc margin. There is a slight degree of retinal pigmentation in some parts of the fundus which has developed gradually since the operation.

Pathological Examination: The writer acknowledges his indebtedness to Mr. Williamson-Noble for sections of the specimen. The specimen consisted in a short piece of optic nerve, the anterior extremity of which widened out into a collapsed sac

with walls of from $\frac{1}{4}$ to $\frac{1}{3}$ of an inch in thickness. The sac showed a small opening at its posterior end through which the contents had escaped. (At the time of the operation some thick fluid escaped from this opening which was made with the scissors in separating the mass from the posterior limit of the orbit.) A small portion of optic nerve was removed and embedded separately after fixation in formalin. The remainder of the optic nerve with a portion of the adjoining wall of the sac was fixed in Müller's fluid and embedded in paraffin. Transverse sections of a small portion of the optic nerve were made and stained with Weigert's iron haematoxylin and van Gieson's stain (picric acid fuchsin). Sections of the other piece of tissue, cut longitudinally, were stained with: (1) Ehrlich's haematoxylin and eosin, (2) Weigert's iron haematoxylin, and van Gieson's stain, (3) thionin, (4) polychrome methylene blue.

Microscopic: The optic nerve in cross section at its anterior end shows a normal structure. Sections stained by the 3rd and 4th methods failed to reveal any evidence of the presence of mucin. The longitudinal sections through the optic nerve and part of the sac wall show the following condition (See Fig. 2):—Through the extent of the sections of this portion the dural sheath appears to be free from involvement. In some sections it is interrupted, probably by the process of cutting, but in others it is continuous. At the anterior extremity of this portion the dural sheath is separated from the pial sheath for, on the average, a distance about equal to half the normal diameter of the optic nerve. Here and there traces of arachnoid sheath and its covering endothelium can be seen close to or in contact with the inner surface of the dural sheath. van Gieson's stain shows well the condition of the nerve sheaths. In the anterior half of the sections there is a great increase in the collagen fibrous tissue, internal to the remnants of the endothelial cells of the arachnoid sheath. This appears to have connection mainly with the pial sheath. On one side of the optic nerve the pial sheath can be traced very definitely throughout the length of the section. On this same side the posterior part of the space between pia and dura is occupied by new formation similar to that which involves the nerve, which will be described later, but with this difference—that strands of collagen fibre taking tortuous paths through it break it up into irregular masses. At the other side of the section the pial sheath can be traced from the anterior end to about half the extent of the section, and then is split up into numerous strands by tissue of new formation similar to that to which reference has been made. The optic nerve, sectioned slightly obliquely and not completely in the longitudinal direction, at its anterior extremity in the sections is of normal appearance.

It shows near its anterior end a constriction with interruption of many of the nerve bundles, presumably the site of the ligature

CASE I. FIG. 2.

From a Coloured Drawing of section of part of the tumour and optic nerve of Case No. 1. Low power. Section stained with Weigert's iron haematoxylin and van Gieson's picric acid fuchsin stain. Fibrous connective tissue (collagen) coloured red in original Coloured Drawing. *a*. Dural sheath of optic nerve. *b*. Invasion of subdural space by gliomatous tissue, towards posterior end of section. Proliferation of pial sheath towards anterior end of section. *c*. Pial sheath. *d*. Zone of optic nerve whose interseptal spaces have been widened by new gliomatous tissue. The latter has undergone some form of degeneration. *e*. The cleft indicates the position of the fluid contents of the tumour, the result of degeneration. *f*. Part of pial sheath invaded by gliomatous tissue. *g*. Pial sheath. *h*. Dural sheath. *k*. Anterior end (towards eyeball) of optic nerve, showing part, of normal structure.

which was passed around this part of the nerve during the operation. The normal appearance of the nerve is continued backwards for about half its extent, and from there the interseptal spaces and the nerve bundles within them widen out gradually. Finally, the pattern of longitudinally arranged septal bundles of

collagen fibrous tissue is entirely lost, and the remaining widened mass of tissue, somewhat fragmented, is of irregular pattern and of grey or yellow-grey colour as stained by van Gieson's method. It appears, therefore, from examination with a low power objective that the optic nerve has suffered expansion by a new formation which has destroyed the normal pattern of the nerve, widened the interseptal spaces, and invaded and largely destroyed the pial sheath on one side. New formation noted on the other side of the sections as lying between the pial and dural sheaths is presumably in direct continuity with that which has permeated and destroyed the pial sheath in one part, and has therefore extended in the subdural space so as to surround more or less completely the optic nerve.

Examination with a Zeiss objective A and No. 2 eyepiece shows the mass of new formation to be composed of fairly abundantly nucleated tissue with innumerable fibrils forming a kind of felt-work structure. Where the tissue of the septal interspaces becomes merged in that of felt-work pattern there are many small spaces, as from oedema. Under examination with a $\frac{1}{6}$ th Zeiss objective and No. 2 eyepiece, the nuclei are seen for the most part to be oval or round, and of about the size of red corpuscles or slightly larger, and darkly stained. Among these nuclei are some larger, oval, and less darkly stained. The cytoplasm of some of the cells can be seen to extend into several branching processes. Where the tissue is fragmentary the nuclei are fewer in number and the intervening substance is broken off into granules. The fibrils in the widened interseptal spaces are arranged mainly in a longitudinal direction, but in close proximity to the septa there is a definite transverse fibrillation, but not quite so well marked as in a similar case described by Mr. A. C. Hudson (*Royal London Ophthalmic Hospital Reports*, Vol. XVIII, p. 317, illustrated).

A consideration of the above observations leads one to the definite conclusion that this case falls into the group named by Hudson gliomatosis of the optic nerve.

Case II*

Matilda C. (Royal London Ophthalmic Hospital, Path. No. 423), aged 79, seen 4/11/21. This patient was admitted to Moorfields Hospital under the care of Mr. E. Treacher Collins, to whom the writer is indebted for permission to use the notes of the case. The clinical notes were made by Mr. Basil Graves.

History: There was great uncertainty as to the reliability of the patient's statements, owing to the senile mental condition. It

* On March 20, 1923, that is to say, more than 16 months after the operation of partial exenteration of orbit, news was received of this patient. A district nurse visited her and stated that the socket was quite healthy. The patient, however, was so infirm, owing to her great age, that it was impossible for her to visit the hospital.

was stated that the right eye had been blind for many years, but that it had been prominent only for three weeks. There had been marked pain recently. Clinical examination on November 4, 1921, revealed moderate proptosis of the right eye, in a forward direction. There was some chemosis of the conjunctiva. Slight limitation of movements. The cornea was clear and bright. The anterior chamber was rather shallow; the iris was normal. There was a mature cataract, which was possibly the cause of the long-standing failure of vision in this eye. V. = no perception of light. Tension, normal or slightly raised. At the time of operation excision of the eyeball was started, but a partial exenteration of the orbit was performed, as a mass was felt behind the eyeball.

Macroscopic: The eyeball measured 24 mm. antero-posteriorly, 24 mm. laterally, 23 mm. vertically. There was attached to the

CASE II. FIG. 3.

Drawing of divided eyeball and tumour of nerve.

The plane of section does not pass through the centre of the optic disc.

The growth is seen to involve the sheaths and not the nerve itself.

posterior part around and obscuring the optic nerve, a hard irregular mass, partly damaged during operation (See Fig. 3). This mass measured 14 by 14 mm. in thickness, and 16 mm. from the posterior limit of the eyeball to the posterior extremity of the mass surrounding the optic nerve. Many loose fragments of orbital fatty tissue were with the specimen. The tissues were fixed in 10 per cent. formaldehyde after immersion in Zenker's fluid for one hour. After division of the specimen in the horizontal plane the larger part was placed in Müller's fluid for 3½ months. The cut surface of the growth at the back of the eyeball showed a central zone, grey-yellow in colour, which appeared to be continuous with the nerve fibre layer of the retina. There was a dark area of tissue surrounding this, of semi-transparent gelatinous appearance. Outside of this was the much-thickened sheath (? dural). The cornea was of normal appearance. The anterior chamber was shallow; the iris and ciliary

body normal. The lens showed extensive subcapsular and cortical changes. The retina and choroid were in situ. There was a pale

CASE II. FIG. 4.

From a Coloured Drawing of section of optic nerve and new growth, with posterior part of retina and sclerotic. Low power. The section passes somewhat obliquely, so that the optic nerve does not pass through more than a small part of the section. *a*. Degenerate retina. *b*. Central vessels. *c*. Lamina cribrosa. *d*. Degenerate optic nerve, with increase of fibrous tissue. *e*. Pial sheath, intact. *f*. Sclerotic. *g*. Dural sheath, split up into bundles by the growth. *h*. Extra-dural extension of growth. In the section the growth appears to extend posterior to the nerve, in a broad zone. This mass is seen in this position because of the obliquity of the section, and is in reality a continuation of the growth which lies between the pial and dural sheaths.

area of atrophy, with serpiginous outline, around the disc, equal to about eight disc diameters at its broadest part. In the extreme

anterior part near the ora serrata there were some irregular patches of choroidal atrophy with loss of pigment, and on the inner surface near the equator were some scattered streaks of retinal pigmentation. The disc edges appeared to be rather prominent particularly above.

Part of the anterior half of the globe was embedded in celloidin, and part of the posterior half in photoxylin and paraffin (Jordan's method).

Sections were stained with: (1) Ehrlich's haematoxylin and eosin; (2) Weigert's iron haematoxylin and van Gieson's stain; (3) Weigert-Pal's medullary sheath stain.

Microscopic: Sections of the globe show conjunctival vascularization and slight cell infiltration. The cornea is of normal structure; the anterior chamber shallow; and the angle particularly narrow. The iris and ciliary body are normal; the lens is the subject of advanced subcapsular and cortical cataract. The retina shows much atrophy and loss of pattern. There is a flattened cystic space between the retina and pigment epithelium at the ora serrata on one side. There is some variation in the amount of pigmentation in the pigment epithelium in different parts. The choroid is thin. Sections of the optic nerve stained by the Weigert-Pal method show complete absence of medullary sheath staining of the nerve fibres. A fragment of a posterior ciliary nerve in the section shows medullary sheath staining.

The other methods of staining show marked degeneration of the optic nerve with apparent increase of fibrous tissue. There appears to be considerable increase in the fibrous tissue at the lamina cribrosa. The pial sheath is present and intact. The dural sheath is displaced outwards considerably and is also split up into numerous longitudinal bundles by a cellular new formation. This new formation is present mainly in the subdural space and is bounded internally by the pial sheath. At its thickest part in any of the sections this mass is about 3 mm. in thickness. It is composed of irregularly shaped masses of cells which are separated from neighbouring masses by strands of collagen fibrous tissue. Many of the masses of cells are roughly circular, and in some the cells have somewhat of a whorled arrangement. The most noticeable feature, however, is the presence of numerous small spaces between the cells and within the various masses of cells. Examination with a $\frac{1}{4}$ th objective and No. 2 eyepiece shows the nuclei to be for the most part of size larger than that of a red corpuscle up to a diameter equal to that of two red corpuscles. The nuclei are circular or oval, are mostly lightly stained, and many of them possess a small dark nucleolus. The cytoplasm of individual cells is not clearly demarcated from that of neighbouring cells. Within the cytoplasm may be seen many minute clear

spaces. The larger spaces, already mentioned, are in all cases bounded by cells of the new growth. In these situations the nuclei are generally flattened and elongated. Bridging the larger spaces are frequent delicate strands of cytoplasm dividing these spaces into several parts. There are fairly numerous blood

CASE II. FIG. 5.

Microphotograph, magnification 100. Shows new growth within the retina and a mass protruding into the vitreous cavity. *a*. Mass anterior to optic disc. *b*. and *c*. Branches of retinal vessels at disc. *d*. Growth within the retina.

capillaries present in the growth, but they are everywhere surrounded by a well-formed collagen fibre wall. Although, as already stated, the growth is for the most part within the subdural space or included within portions of the dural sheath, yet processes of the growth have in some places spread through and to the outer surface of the dural sheath. In some sections small masses of growth are seen in close connection with blood-vessels which lie within the sclera in the situation of perforating vessels to one side

of the optic nerve. There are some irregular masses of growth of small dimension within the atrophic retina near the disc margin. These masses of growth should certainly have been visible to ophthalmoscopic examination if the lens had not been cataractous. The growth cells extend to the extreme posterior limit of the sections.

The microscopic appearances of the new growth in this case resemble very closely those of one case described by Mr. Hudson and illustrated with microphotographs. In the latter case, however, the growth did not seem to have formed spaces among and

CASE II. FIG. 6.

Microphotograph, magnification 100. Shows new growth lying between strands of dural sheath. The portion 'a' is shown more highly magnified in the next.

between the cells of the growth. In the writer's case, the cell-vacuolation and the space formation between cells is a striking feature. This point is emphasized by Kettle and Ross. There is no doubt that this is a typical example of endothelioma of the optic nerve, arising almost certainly from the endothelium of the subdural space.

It may be of interest to review briefly the main clinical features of optic nerve tumours.

In Parson's *Pathology of the Eye* (Vol. II, p. 693), optic nerve tumours are subdivided into intradural and extradural. Hudson classifies these growths as: (a) gliomatosis, (b) fibromatosis,

(c) endothelioma. In Hudson's series account is taken under these headings strictly of primary tumours of the nerve; that is to say, tumours, arising outside the dural sheath and extending so as to involve this structure, are excluded. In Hudson's extensive review of the literature of this subject 118 cases were considered as being with fair certainty or probability of the first type—gliomatosis. Six cases were considered to be fibromatosis, and 29 cases as probable endotheliomata.

CASE II. FIG. 7.

Magnification 260. Shows the grouping of the cells, and the formation of many spaces between them, 'a. a.' The nuclei show the light shade of endothelial cells, many with a darkly marked margin and a small dark nucleolus.

(a) *Gliomatosis*: Sex. Of 118 cases 113 had the sex noted: 70 were female, 43 were male. Age: More than 75 per cent. occurred within the first decade. Vision: Primary defect of vision and subsequent exophthalmos was the commonest sequence. Finally, in the majority vision was almost or wholly abolished. Exophthalmos was of very slow development. Protrusion of the eyeball was mainly in the orbital axis. Movement of the eyeball, if but little restricted, was regarded as suggesting freedom of the anterior part of the nerve from growth. The optic disc in most cases showed either papilloedema or post-neuritic atrophy. Hypermetropia was noted in 16 cases, and in 1 case this was

noticed to have increased. Incomplete removal of the growth was carried out in almost 50 per cent. of the cases.

Intracranial growth was found after death in 15. In 9 others cerebral affection was suspected.

Local recurrence in the orbit was not reported in a single case. Freedom from recurrence was noted in 16 cases for a period of from 2-24 years, and in 18 cases for from 1-2 years. In many of these cases removal of the growth was known to be incomplete.

Operation: In 43 removal of the tumour without removal of the globe was carried out. Methods of operating appear to be about fairly evenly divided between the method of Krönlein with bone resection, and one without bone resection. Twenty-eight cases treated by the former method gave 7 first class results, and 22 by the latter method gave 8 first class results. Complications, deviation, ptosis (8 and 3 cases respectively), phthisis bulbi, consequent enucleation, death from meningitis (2 cases to each method).

In the writer's case of gliomatosis the sex was male, the age at onset of symptoms was 8 years. Exophthalmos, however, was noticed before any failure of vision was detected. Final vision was merely perception of shadows. Exophthalmos was of six years' duration before operation, protrusion was mainly forwards, slightly upwards, movement was not restricted, the appearance of the disc suggested very slight papilloedema. Incomplete removal was practically certain for the degenerated cystic mass was opened at the posterior limit of the orbit during operation. This case has remained free from local recurrence or from cerebral symptoms for 17 months. The tumour was removed without bone resection, but resulted in a fixed divergent eye with ptosis.

In the circumstances of this patient it was surely an advantage to him that he should possess his own eyeball, although it be divergent, with paralysis of the levator, but it is open to discussion whether in the case of a patient in better circumstances the advantages would not have been greater with removal of the eyeball and the supply of an artificial eye, provided that the function of the levator palpebrae could have been retained.

(b) *Fibromatosis*: The six cases reviewed by Hudson included 5 in which the dural sheath alone was involved

(c) *Endothelioma*: Sex: 20 female, 7 male. Age: In more than 50 per cent. the onset of the symptoms took place after 30 years of age. Vision: Exophthalmos rather more often preceded visual defect. Exophthalmos was usually slow, but occasionally quite rapid in development. Movement of the eyeball was more frequently limited than in the group of cases of gliomatosis. This indicated probable involvement of the anterior part of the nerve, as is usual in this condition. Intracranial extension was found after death in 7.

Invasion of neighbouring structures was of fairly frequent occurrence. The optic nerve, nerve head, retina, choroid, sclera, orbital fat, and ocular muscles; nasal fossae (1 case). Recurrence was recorded in three, in one of which it was as late as 25 years after operation. Freedom from recurrence was only recorded in 6 cases, with an interval of from 7 months to 4 years after operation. *Treatment*: Removal of the tumour with the eyeball was reported in 17 cases, and without eyeball in 5 cases.

Differential diagnosis of endothelioma from gliomatosis depends upon the following factors:—

In endothelioma (1) the age is greater; (2) exophthalmos more often precedes visual disturbance; (3) limitation of movement; (4) circulatory obstruction in lids and conjunctiva; (5) pain; (6) intraocular extension.

In the case described by the writer the sex was female, age 79, vision was completely abolished and had, as far as could be estimated, been absent for a long period before proptosis was noticed. The exophthalmos, however, was of uncertain duration. Movement was slightly limited. This is in accord with the fact that the growth was situated at the posterior pole of the eyeball and extended into the coats of the same. Invasion of sclerotic and retina was present.

The above-described investigation leads to the conclusion that of two cases of primary intradural tumour of the optic nerve, one is an example of the group of gliomatosis of the optic nerve, the other of endothelioma which had its origin in the endothelium of the arachnoid sheath.

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ENDOTHELIOMA OF THE ORBIT*

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DURING the last two years at the Central London Ophthalmic Hospital there have been two cases of orbital tumour. Although the subject of these tumours is one which has been discussed at length by many writers, it is worth while describing these cases, as they both present some points of interest.

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