Two cases of primary neoplasm of the optic nerve / by E. Treacher Collins and C. Devereux Marshall.

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Two cases of primary neoplasm of the optic nerve.

By E. Treacher Collins and C. Devereux Marshall.

(With Plate V.)

During the last session of this Society two papers were read dealing with cases of tumour of the optic nerve or its sheaths, but so rare are these, and so different are the appearances which the growths present microscopically, that we have not hesitated in bringing two fresh cases before you this evening.

To show how rare they are, we may mention that during the last fifteen years (that is from 1885 up to the present time) only two growths of the optic nerve have been removed at the Moorfields Hospital, whilst during this time 388,000 patients have been treated.

As to the difference in the microscopical characters of the neoplasms, we find that of the five cases which have been recorded in this Society's 'Transactions,' one is described as a fibroma (Brailey, vol. ii), one as a sarcoma of the sheath (Brailey, vol. vii), two as gliomata (Rockliffe, vol. xiii, and Bullar and Marshall, vol. xix), and one as a myxo-fibroma of the sheath (Arnold Lawson, vol. xix).

Case 1 is that of a healthy-looking boy, Harry L—, æt. 5 years, who came under the care of one of us at the Moorfields Hospital on August 24th, 1899.

His father stated that the boy's right eye had been more prominent than its fellow during the last eight months, that the amount of prominence had during that time gradually increased, and that the sight of that eye had failed.

On examination it was found that his right eye was

markedly proptosed and at the same time displaced a little downwards. The movements of the globe laterally and downwards were unrestricted, but there was slight defect in the movement upwards. No swelling could be felt in the orbit.

The pupil of the affected eye was larger than that of the other, and was inactive to light. The lens and vitreous were clear. The optic disc was somewhat pale, and its margin was ill-defined; the retinal veins were large and tortuous. In the region of the optic disc it was estimated that there were 4 D. of hypermetropia. The tension was normal, and the vision was hand movement only.

The presence of proptosis with but slight eccentric displacement of the globe, and but little limitation of movement, together with loss of sight, pallor of the disc, and tortuosity of the retinal veins, led, at that time, to a tumour of the optic nerve being suspected. It was, however, thought well, before resorting to any operative procedure, to try first the effect of a course of treatment with iodide of potassium. Five-grain doses were given three times a day for a month. At the end of that time the proptosis was found to have increased, and the amount of hypermetropia in the region of the disc to have become 9 dioptres.

It seemed impossible to conceive of any condition, other than a new growth in or around the nerve, capable of producing proptosis and an increasing amount of hypermetropia.

On September 21st, 1899, the patient having been anæsthetised, an exploratory incision was made through the skin and palpebral ligament at the outer margin of the orbit, and the finger was then passed back to the optic nerve, which was felt to be enlarged. The eyeball was then enucleated in the ordinary way. At the first cut with the scissors through the nerve only a portion of the growth was removed, the remaining part was felt for and divided close up to the optic foramen. The patient

was last seen on November 6th, 1899, and was in good health with no sign of recurrence.

Pathological description.—The eye to external appearance is healthy. The optic nerve is greatly enlarged, it being at its thickest part 13·5 mm. in diameter. The enlargement is spindle-shaped, and commences close to the sclerotic in front, reaching backwards 23 mm. At its posterior extremity the optic nerve is slightly larger than normal, and the cut surface has a gelatinous appearance, rather like that of the tumour. The dural sheath is not involved, and can be stripped off as in a healthy nerve.

On section the eye itself is normal, though somewhat compressed in its antero-posterior diameter by the tumour.

The optic nerve, which is so much enlarged, has, on section, the following appearance. Exactly at the lamina cribosa it differs but little from normal, but immediately it leaves the sclerotic it commences to enlarge. The central portion of the new growth is dense and looks like a very hypertrophied optic nerve. The peripheral portions appear less dense, and seem to be composed of tissue surrounding the nerve, probably thickened pial sheath.

The central artery can be seen in its usual situation in the centre of the nerve.

Microscopical appearance.—Transverse sections of the growth show it to be mapped out into areas by a thick network of bundles of connective tissue containing small blood-vessels. This fibrous-tissue framework resembles that of the normal nerve, except that its component parts are much thicker and denser.

In the interspaces of this network instead of bundles of nerve-fibres supported by delicate connective tissue, the nerve-fibres seem to have all disappeared, leaving empty spaces very much reduced in size, whilst the supporting connective tissue is everywhere increased, thickened, and more cellular than normal (Plate V, fig. 1).

In longitudinal sections the spaces left by the atrophied nerve-fibres do not show, and the spaces between the

bundles of fibrous tissue appear filled with long spindle-shaped cells, which at the borders of the trabeculæ merge into and mix with the fibrous tissue (Plate V, fig. 2). The thickened pial sheath is also seen to be composed of collections of spindle-cells and strands of fibrous tissue, the latter less dense and thick than in the nerve itself.

Case 2, Mrs. P—, æt. 46, was under the care of Mr. Austin Reynolds and Mr. Poulett Wells, who stated that the patient was suffering from detachment of the retina with hæmorrhages and proptosis. The vision of the right eye had been failing for nine months, and the condition was gradually getting worse. Tension ? + V = no P. L. The movements of the eye were quite normal and not impaired in any direction.

The presence of an intra-ocular growth extending through into the orbit was suspected, and it was decided to exenterate the orbital contents.

Owing to illness Mr. Poulett Wells asked one of us to operate for him, and on November 30th, 1897, enucleation of the eyeball was performed in the usual manner. On division of the optic nerve it was found to be considerably enlarged, and although it had been divided far back, the incision had gone through the middle of a new growth, and a large mass was still left. As it was considered probable that it might be of a malignant nature, the contents of the orbit were removed, the optic nerve being divided as close to the foramen as possible.

Pathological examination.—After hardening the following condition was noted. The anterior part of the eye is healthy, except that the lens has numerous congenital opacities on it and in it. The vitreous is clear. The optic disc does not appear to be altered, there are numerous hæmorrhages in the retina; but there is no intra-ocular neoplasm.

The optic nerve is very much enlarged, and forms a fusiform swelling measuring 7·5 mm. in diameter at its thickest part. The swelling commences immediately the

nerve leaves the eye, and is continued backwards nearly up to the point of section.

Microscopical appearances.—Transverse sections at the thickest part of the tumour show a somewhat thickened pial sheath, immediately beneath which is some round-celled infiltration. From the pial sheath dense processes of connective tissue extend inwards, and separate off spaces in which optic nerve-fibres were originally located; but these are now occupied with a network of irregular branching cells in which not a single healthy nerve-fibre is visible (Plate V, fig. 3). The vessels are not unduly enlarged.

We see that the growths in these two cases, though somewhat similar, are in certain respects different. In the first case the pial sheath in much involved, and in the second it is but slightly thickened. In both cases the fibrous-tissue trabeculæ which extend into the nerve from the pial sheath are thickened, but much more markedly so in the first case. In both there is an increase in the supporting tissue network of the nerve-fibres in between the fibrous-tissue trabeculæ, which is also much more cellular and more like embryonic tissue than normal; but in the first case the cells are definitely large and spindle-shaped, while in the second they are indefinite, many of them being irregular and branching. In both, in the region of the growth there has been complete destruction of the nerve-fibres, small circular spaces being left where they formerly extended.

A point of interest in connection with the second case is that the appearance which is presented clinically had suggested the possibility of its being a case of sarcoma of the choroid.

A case of new growth of the optic nerve accompanied by retinal detachment, which was also clinically taken to be a sarcoma of the choroid, has been recorded by Dr. Brailey in vol. ii of this Society's 'Transactions.'

In his case, as in ours, no new growth in the eye was

found. Microscopically the growth in the optic nerve was seen to be principally due to a new formation of fibrous tissue within it. "A hypertrophy of the trabeculæ evidently forming the basis of the new growth."

The names and descriptions which have been given of the pathological characters of tumours of the optic nerve are exceedingly numerous and varied, but it is highly probable that many of them which have been described under different names are essentially of the same nature.

Tumours of the optic nerve are so rare that few observers have had the opportunity of examining sections of more than one.

We are fortunate in being able to show to the Society this evening the macroscopic and microscopic characters of six of these growths collected from different sources. Careful study of these leads us to believe that they are all primary growths of the connective-tissue framework of the optic nerve. This consists of the pial sheath, prolongations of the fibrous tissue forming trabeculæ in the substance of the nerve, and a delicate connective tissue (the so-called neuroglia) supporting the nerve-fibres in the spaces left between the trabeculæ.

In some cases the growth seems to have involved all these elements (Case 1, Lawson's case, and Bullar and Marshall's case). In others, one or other part is more extensively affected (Case 2 and Rockliffe's case). In some the development of the tissue composing the new growth is more highly organised than in others; thus, whilst in some a large amount of fibrous tissue is met with which would justify their being described as fibromata, in others the tissue is of a more embryonic type and the cells vary from spindle-shaped to branching or round, as is met with in the developing optic nerve. Thus the terms glioma or sarcoma would appear applicable. In others, again, tissues of varying degrees of organisation are met with in the same growth, suggesting such compound names as fibro-sarcoma, etc.

We have arranged six sections of the collected cases in

order, showing the relative degree of development of the tissue composing the new growth. Thus we have :—

Case 1 composed of fibrous tissue and long spindle-cells ;

Cases 2 and 3 (W. W. Sinclair) with thickening of the fibrous trabeculæ and a network of irregular cells ;

Case 4 (W. C. Rockliffe) with hardly any increase of fibrous tissue, but many large and branching cells.

Case 5 (G. Lawson) is somewhat similar to Case 4, but the cells are rounder in shape and less branching.

Case 6 (Bullar and Marshall's) is the most lowly organised of any, and is composed almost entirely of round cells.

In studying the published reports of optic nerve tumours it will be noted that in many of them myxomatous changes are described. In none of the six specimens which we have had the opportunity of examining have we been able to discover any true myxomatous changes. In most of them vacant spaces are to be seen scattered throughout the growth, some small, some large, but in none of them have we been able to detect any mucoid material.

Some of these spaces are certainly channels in which the nerve-fibres were contained, but which, owing to their complete atrophy, have been left vacant.

In conclusion we would urge that much of the confusion which has arisen with regard to the nature of optic nerve tumours is due to the nomenclature which has been adopted, and that this confusion is only likely to be cleared up by recognising the essential principles which account for the varied appearances which they present, such as those which we have endeavoured to indicate.

(*November 9th*, 1899.)

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PLATE V.

Illustrates Messrs. E. Treacher Collins and C. Devereux Marshall's paper on Two Cases of Primary Neoplasm of the Optic Nerve.

FIG. 1 shows the microscopical appearances of a transverse section of the growth from Case 1.

FIG. 2 shows the microscopical appearances of a longitudinal section of the growth from Case 1.

FIG. 3 shows the microscopical appearances of a transverse section of the growth from Case 2.

Fig. 1.

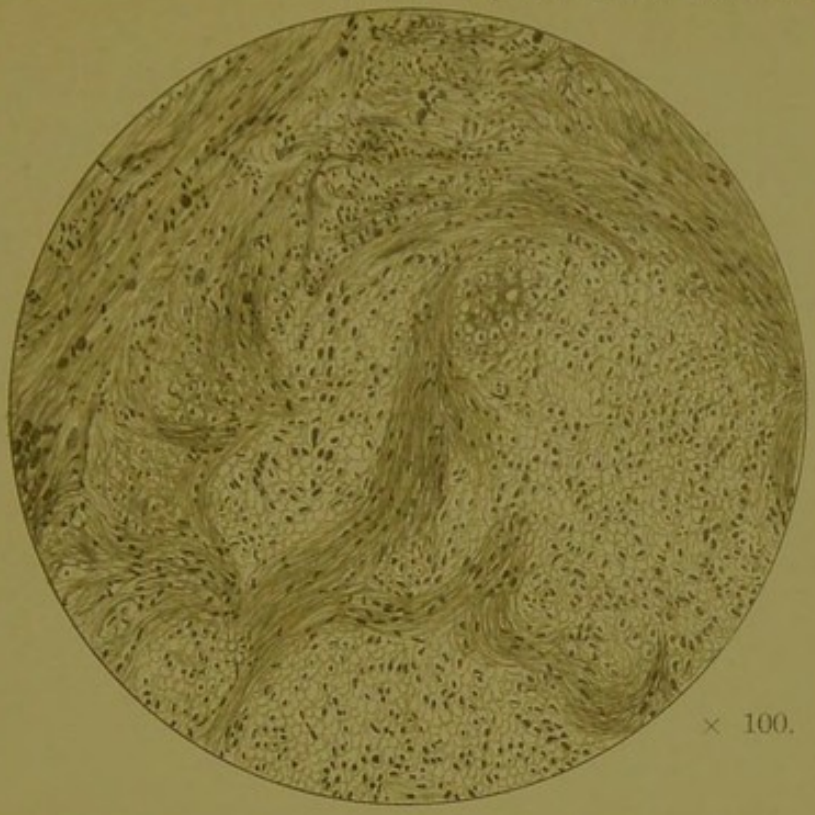

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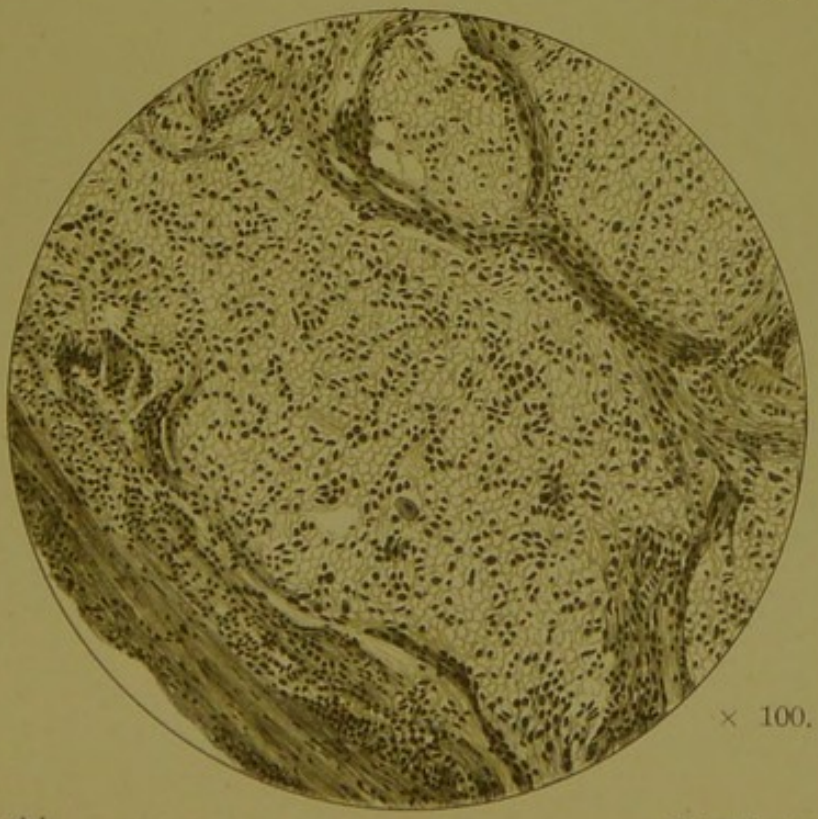
Fig. 2.

× 100.

Fig. 3.



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