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A CASE OF ENDOTHELIOMA OF THE OPTIC NERVE SHEATHS

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Endothelioma of the optic nerve sheaths is a relatively rare neoplasm but according to Byers the commonest of the tumours affecting these structures. Hudson in his paper about neoplasms of the optic nerve and its sheaths quotes 118 cases of gliomatosis, 29 of endothelioma and 6 of fibromatosis among the specimens and reports he collected. This neoplasm is composed of endothelial cells derived either from those adjacent to the dura and lining the sub-dural space or from the cells covering the fine connective tissue and elastic tissue strands which form the arachnoid mater. Mallory does not question this origin but considers that the histological term endothelioma is fundamentally incorrect and that arachnoid fibroblastoma would be more appropriate on the grounds that the endothelial cells are merely differentiated connective tissue cells. This conception is supported by the presence of collagenous and elastic fibrils in these tumours, probably produced by the endothelial cells under conditions of tumour formation. Neame comments on the fact that there is more fibrous tissue than could have arisen by the permeation of the neoplasm into the dural sheath and the sclera with the subsequent expansion of the fibrous tissue layers and lamellae of those structures.

The clinical features described in the literature conform in the main to the following summary. The number of cases reported is too small for statistics to have any significance, so these have been omitted for the purposes of this paper.

Females were affected more often than males and the majority of cases were over 30 years of age, one case recorded by de Schweinitz was 11 and another by Neame was 79. Symptoms of failing vision were gradual in onset in most cases. Impaired movements were more common in cases of this neoplasm than in
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others affecting the optic nerve and its sheaths. At first the limitation was in upward, and upward and outward movements, and later in some cases the movements of the eye in all directions became affected. Exophthalmos developed relatively late and displaced the eye straight forwards. Deviation of the eye down and out was recorded by de Schweinitz in a female child 11 years of age. Increasing hypermetropia was evident in cases where the neoplasm was situated close to the posterior surface of the globe and in a case described by Neame and Wolff this amounted to 7.5 D. Fundus examination revealed changes suggestive of optic neuritis, papilloedema, post-neuritic optic atrophy, and areas of pigmentary disturbance and choroidal atrophy probably caused by interference with the circulation of blood through some of the posterior ciliary vessels. Vision becomes considerably reduced over a course of 4 or 5 years to counting fingers at 1 metre, hand movements or bare perception of light. In a few advanced cases the neoplastic mass could be palpated in the orbit. The operation of choice seems to be partial or total exenteration of the orbit depending on the size and extent of the neoplasm. By the time that symptoms and signs permit of a diagnosis the neoplasm is often too extensive for local excision. In one case the affected part of the optic nerve was resected but the eye became fixed in a position of external deviation and ptosis was an additional sequel.

The histological appearances present certain points of pathological interest. By the time the neoplasm is removed or a post-mortem made it is in most cases impossible to state with any certainty the precise site of origin of the endothelial cells, whether from the layer of these cells that lines the under surface of the dura or from those that cover the connective tissue and elastic strands of the arachnoid mater or from the lining of lymphatics and capillaries. The cells are arranged in ill-formed columns, clumps and whorls and sometimes can be seen occupying the lumen of a blood vessel. Fibrils of collagenous material and fibrous tissue are evident between some of the cell masses. Psammoma bodies with concentric lamination are sometimes present. The neoplasm invades the dura in advanced cases splitting it and extending between bundles of its fibrous tissue. The sclera in the vicinity of the optic disc and posterior pole of the eye is infiltrated in some instances, but in most cases no extension seems to occur through the pia, which remains intact. The optic nerve becomes damaged by compression and the papillomacular bundle seems to suffer most. The myelin sheaths are shown as degenerated in suitably stained sections but the axis cylinders probably retain their function for a long time.

Intracranial extension is a terminal event. Cohen and MacNeal
describe a case in which the neoplasm arose in the optic canal and extended forwards into the orbit and posteriorly into the middle cranial fossa. The neoplasm was adherent to but did not infiltrate the body and lesser wing of the sphenoid on the affected left side. The left optic nerve, chiasma and right optic nerve were displaced towards the right side and were stretched and thin. A very unusual feature of this particular case was the invasion of small islands of metastatic carcinoma in the midst of the endothelioma. Other nodules of secondary deposits of carcinoma cells were in the brain, the primary growth having been in the left breast.

Owing to the origin of the endothelioma in the optic canal there was no clinical evidence of exophthalmos, displacement or impaired movement of the globe.

Cell vacuolation and intercellular spaces which appear as clear clefts in the sections are histological features of endothelioma commented upon by Kettle and Ross and mentioned by Neame in his paper.

Case Report

W. C., a clergyman, 30 years of age, was admitted to the Moorfields Eye Hospital under the care of Mr. P. G. Doyne on March 28, 1928. He gave a history of mistiness of right vision for 5 weeks. Two-thirds of the optic disc on the nasal side was oedematous and the degree of papilloedema recorded was 3 D. The remaining one third of the optic disc on the temporal side was clear. The oedema extended into the adjacent retina on the nasal side. Vision was reduced to hand movements in the temporal field. The late Dr. Adie reported that there was no evidence of disease in the central nervous system apart from the ocular lesion.

The patient was examined by the Honorary Staff of the hospital and the consensus of their opinion was that optic neuritis of local origin was now subsiding.

The condition remained unchanged till August, 1929, when the patient complained of headaches. The right eye showed slight ciliary injection and the pupil was three quarters dilated and immobile. Some small newly formed vessels were present on the anterior surface of the iris; there was an extensive detachment of the lower half of the retina and a dark area adjacent to the optic disc in the lower nasal quadrant was evident. Some retinal haemorrhages were seen in the upper half of the retina; there was no perception of light and the intra-ocular pressure was raised. The left eye was normal and had 6/5 vision. On September 20, 1929 the right eye was excised and in removing the eye the optic nerve and sheaths were found to be much thickened.
**Pathological Report**

*Macroscopic.* (See fig. 1).—The vertical diameter of the optic nerve and its sheath at their junction with the sclera was 10 mm. and the horizontal 9 mm. The optic nerve had been divided 16 mm. posterior to the globe and at this point the diameter of

![FIG. 1.](image)

Microphotograph of vertical sagittal section through the globe and optic nerve. (a) Endothelioma in the arachnoid sheath. (b) Compressed optic nerve. (c) Inter-retinal fluid beneath the detached retina.

the nerve and its sheath was 7 mm. The pear-shaped appearance of the neoplasm, the optic nerve and its sheath is shown in Fig. 1. The dura was thickened, the arachnoid occupied by a neoplasm which completely surrounded the optic nerve and compressed it to a thin cord of irregular thickness varying from 1-2 mm. in diameter. The neoplasm had a yellowish granular appearance, and the remnant of the optic nerve was greyish in colour with its edges sharply defined. In the lower half of the globe the retina was detached from the optic disc to a point about 3 mm. in front
of the equator. Homogeneous exudate occupied the inter-retinal space. The filtration angle was occluded by peripheral synechiae between the iris root and the posterior surface of the cornea. The eye was divided vertically.

Microscopic.—The neoplasm probably arose from the arachnoid mater. It extended forwards as far as the most anterior limit of the arachnoid space where it infiltrated some of the posterior layers of the sclera, several small groups of cells splitting these

![Microphotograph of a section at the anterior extremity of the arachnoid sheath. X80. (n) Optic nerve. (s) Sclera. (p) Pia mater.](image)

and spreading between the lamellae. Posteriorly it extended as far as the line of excision. There was no histological evidence of intra-ocular extension.

The point of maximum distension of the dura was forwards in the vicinity of the posterior surface of the sclera. The neoplasm was composed of oval-shaped endothelial cells such as are found covering the elastic and connective tissue trabeculae of the arachnoid mater. These cells had a granular cytoplasm and a nucleus with a well defined chromatin network and nucleoli. Clear clefts were evident among some of the cells, the nuclei lying adjacent to these. For the most part these cells had an irregular arrangement but some were disposed in definite groups around blood vessels, some in columns and others in whorls. The cell masses
were broken up by vascularized connective tissue trabeculae and collagenous fibrils. The neoplasm had infiltrated the deeper layers of the dura mater but had not perforated it. The pia mater was thickened and the optic nerve surrounded and sharply defined by several layers of newly formed condensed fibrous tissue and collagen fibres; it was infiltrated at several points by the cells of the neoplasm. There were areas of hyaline degeneration at some sites in the connective tissue trabeculae and in the walls of blood vessels. A few areas of cell degeneration with lymphocytic and phagocytic infiltration were present. At some points there were attempts to form concentrically laminated psammoma bodies. The optic nerve was atrophic and hyaline degeneration was present in some parts of the supporting neuroglial tissue. The central vessels were very dilated and engorged with blood just behind the lamina cribrosa and the capillaries traversing the optic nerve were distended with blood. A moderate degree of perivascular lymphocytic infiltration was present around the central retinal vessels at the optic disc. The dural sheath was thickened and the vessels of the dura and posterior ciliary vessels were dilated and engorged with blood, particularly the veins. There was no evidence of thrombosis of any of the posterior ciliary
vessels but it is possible that some mechanical effects such as kinking and narrowing of the lumen of the affected vessels might have occurred at some points where the posterior ciliary vessels ran in close proximity to the distended dura.

The retina was detached below. Degenerative changes were present in all layers but were particularly well marked in the ganglion-cell and nerve fibre layers. Brown pigment deposits were present in and outside the degenerate ganglion cells in the sheaths of branches of the central retinal vessels and distributed sparsely in the molecular and nuclear layers.

Recent retinal haemorrhages were noted, a large one was present in the upper half of the retina near the ora serrata affecting mainly the ganglion-cell, nerve fibre, inner nuclear and inner molecular layers. The branches of the central retinal vessels were abnormally dilated at the optic disc. Thrombosis had occurred in one of the upper branches of the central retinal vein at a point about 3 mm. from the optic disc. Elsewhere the branches of the central retinal veins were dilated and a number of retinal haemorrhages were present, some in all layers of the retina but the majority in the ganglion-cell layer. Exudates were evident in the outer nuclear and molecular layers in the vicinity of the optic disc. Perivascular lymphocytic infiltration of the upper branches of the central retinal vessels at the edge of the optic disc and a moderate degree of lymphocytic infiltration of all layers of the retina just above this point were present.

The filtration angle was occluded by peripheral synechiae. In the lower part of the globe the endothelial cells on the anterior surface of the iris had proliferated excessively at the false angle and some of these had become converted into fibrous tissue. A clump of leucocytes, some of which contained brown pigment granules was adherent to the cornea near the filtration angle below. The entire uveal tract was flattened and atrophic and ectropion uveae was noted at the pupil margin.

The patient is alive and well at the time of writing this paper, 6 years after the excision of the affected eye. There is no clinical evidence of local recurrence or metastasis. Except for occasional headaches over the remaining eye there are no symptoms.

Commentary.—The clinical course of events and the structural changes in the retina, optic nerve and uveal tract terminating in retinal detachment, secondary glaucoma, optic atrophy and blindness may be accounted for by the mechanical pressure effects of the neoplasm on the central retinal and posterior ciliary vessels first and then the damage done by its direct pressure on the optic nerve.

Compression of the central retinal vein as it traversed the arachnoid space caused the papilloedema, the subsidence of which might be explained by subsequent shrinkage of the optic nerve.
from atrophy and compression, by further posterior extension of the neoplasm along the line of least resistance in the sub-dural space and arachnoid mater and by the establishment of collateral channels of circulation. The retinal haemorrhages were also probably brought about by mechanical obstruction to the return of venous blood.

Kinking and obstruction of certain posterior ciliary vessels were responsible for impairment of the choroidal vascular supply and some of the subsequent pigmented disturbance of the retina, a condition which has been effected experimentally in rabbits by dividing some of these vessels. Some of this pigment probably wandered through the external limiting membrane following atrophy in rod and cone elements. Transudation through the endothelium of the chorio-capillaris produced the homogeneous inter-retinal fluid which caused the retinal detachment. The vascular disturbance of the uveal tract and retinal detachment were factors in inducing secondary glaucoma. Other features of interest about this case of endothelioma of the arachnoid sheath of the optic nerve are its relatively slow rate of growth, its confined activity within the dural sheath, and the length of life free from clinical evidence of local recurrence and metastases.

Its behaviour resembles intracranial endothelioma which compresses the brain but does not infiltrate it except in some cases of long duration. The overlying bone of the cranial vault is however invaded by the neoplasm and either becomes absorbed or manifests new bone formation. Similar features in this case of endothelioma of the optic nerve sheath are its comparatively encapsulated state between the dura and the thickened pia mater. The optic nerve is compressed and the deeper layers of the dura and sclera infiltrated at some sites.

It is evident, then, that for some time at least these neoplasms are of relatively low malignancy and in the earlier stages behave more like an innocent encapsulated growth which damages local structures by compression rather than by infiltration and permeation.

I thank Mr. P. G. Doyne for the clinical notes of this case and for his permission to publish them.

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