

## OLIGODENDROCYTOMA OF THE OPTIC NERVE.

### Report of a Case.

A.-M. Zia Shamsa, M.D. ★ and M.H. Lashkari, M.D., F.I.C.S., ★★

Definition, nomenclature and nature of lesion.

The optic nerve structure is different from those of the other peripheral nerve tracts. It is similar to the white matter of the brain. The sheath is formed by dura-mater, arachnoid and pia-matter. Its tract differs from the intracerebral pathways, being sub-divided into bundles by fibrous septa which enter from the pia-mater. The cells in these bundles are of three types:

- 1— Astrocytes.
- 2— Oligodendrocytes.
- 3— Microglia.

Studies made by Enriquez show that 75% of glial elements of the optic nerve are oligodendrocytes (6). According to Lundberg the majority of the cells composing glioma of the optic nerve are of the oligodendrocytic type (6). Based on this, Lundberg names the tumour **Oligodendrocytoma**. On the other hand, Davis (2) believes that most of the cells forming the tumour are astrocytes. Del Rio Hortega (6,8) indicates the possibility of the tumour being astroblastoma or oligodendrocytoma, but the more noticeable kinds are mono or bipolar types of spongioblastes. Oberling and Nordman (6) think that the lesion is arising from leptomeninges and the tumour cells are analogous to the cells of Schwann. This opinion is also confirmed by Satanowsky and Adroque (6).

Hudson (6,7) views the glioma of the optic nerve as a degenerative lesion and uses the term gliomatosis to describe it; Verhoeff (6) and Weigelin (1) concur in this view.

Davis (3) points out that in patients suffering from glioma of the optic nerve, atrophy is the prime concern, while the increase of glia is of secondary importance.

#### Age

Oligodendrocytoma of the optic nerve observed among children and

★ Cancer Institute,  
Pahlavi Hospital. Tehran University  
Tehran

★★ Department of Ophthalmology,  
Farabi Hospital. Tehran Uni-  
versity. Tehran

adults. Sixty per cent of the patients are in the first decade of life and ninety per cent of all cases occur before the age of twenty (8).

#### Location

The tumour may occur anywhere along the optic nerve in front of the chiasma (4), however it usually originates approximately from 1 cm. behind the globe (8,9). The tumour starts within the central portion of the optic nerve, because it produces, at the beginning, a central scotoma.

#### Multiplicity

Emanuel (6) notes the multiplicity of the tumour and describes ten cases in the literature which demonstrates the multiplicity of the tumour.

#### Frequency

In four hundred cases of optic nerve tumours, observed by Davis (2) two-thirds were of glioma type. Collins and Marshal (8), reported two cases among 388,000 ophthalmologic patients.

#### Case report

A. A, a seventeen-year old white male student, was admitted to Faraby Hospital with symptoms of exophthalmos (Fig. 1) and loss of vision in the right eye.

Five years prior to admission, the patient noticed a gradual progressive diminution of vision and a protrusion of the right eye. At the time of examination, he had been blind in the right eye for four months.

The physical examination revealed the following positive and significant negative findings:

Right eye: Direct exophthalmos, resulting in extrusion of the lower lid.

Lateral movements normal, but upward movements showing some limitations.

Ability to close the right eye but an inability to cover the eyeball due to exophthalmos.

No inflammation of the conjunctiva.

Presence of an almond shaped, mobile painless and palpable tumour behind the globe.

Dilatation of pupil with no reaction to light.

Vision: Nil.

Biomicroscopy: Cornea, anterior chamber, lens and vitreous body normal.

Ophthalmoscopy: Colour of the retina normal.

The pale disc with a distinct circumference, indicating the complete atrophy of the optic nerve.

Intraocular pressure: 30 mm. Hg. Schiötz.

**Left eye:** Visual acuity: 20/20.

Fundus: normal.

Intraocular pressure 20 mm, Hg. Schiötz.

The remainder of P.E., revealed no pathological findings.

C. B. C. and urine analysis were normal.

**The Operation,** (March 10, 1957)

On opening the orbit, through an incision 8 millimeters, below and parallel to the margin of the lower lid, an oval shaped tumour along the course of the optic nerve was exposed. It was brownish-red in colour with a maximum diameter of 35 mm. and a minimum of 20 mm. It was quite mobile and unattached to the surrounding structure except for the optic nerve, which was embedded in the tumour. The tumour was completely removed. The patient's postoperative course was uneventful.

Exophthalmos disappeared (Fig.2) and ptosis of the upper lid occurred.

He was discharged three weeks later. Lack of vision and ptosis of upper lid persisted in the right eye.

Three months later, he returned for a check-up. His left eye was quite normal and ptosis of the right upper lid was improved. No information of the patient is available since then.

#### Pathology:

**Macroscopic:** The tumour was oval shaped about the size of a pigeon's egg (35 mm. X 20 mm.). The optic nerve protruded at both ends (Fig. 3). The colour was brownish-red with a rubbery consistency. It was completely covered by an intact sheath neovascularization around the tumour was observed.

On a longitudinal section of the tumour, the surface displayed two distinct parts: 1- The central area was fusiform with a greyish-white colour (Fig. 4 ). 2- The periphery was red, with non evidence of hemorrhage or degeneration.

**Microscopic:** In general the lesion showed four properties:

1. Hyperplasia of the pia-mater in periphery or between the distinct bundles (Fig. 5,6,7,).

2. Proliferation of the cellular elements of the nerve bundles, and a consequent increase in the spaces between the lamellae originating from the nerve sheath (Fig. 8).

3. Invasion of proliferated glial tissue into hyperplastic pia-mater (Fig. 9).

4. The replacement of pia-mater by glial tissue in a number of areas, notably in the center of the tumour (Fig. 10).

The glial elements composing the major part of the tumour were

of oligodendroglial type. These cells manifested a distinct line between the clear cytoplasm surrounding the nucleus and the dim periphery, resulting in a honeycombed pattern (Fig, 11,12,13,14,15,16,17).

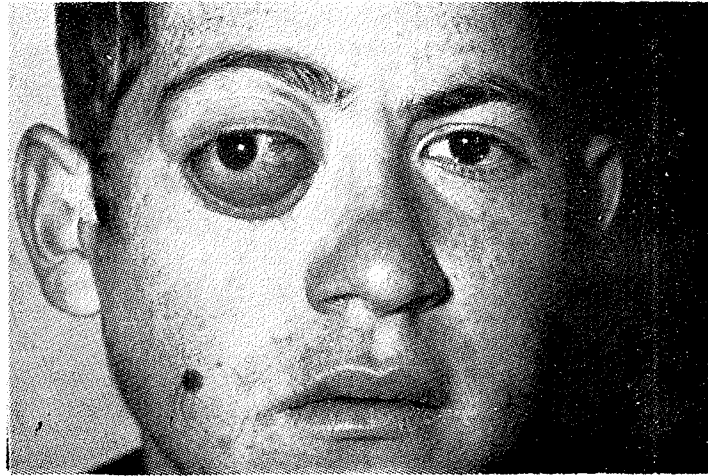


Fig. 1) Direct Exophthalmos of the right eye at the time of admission.

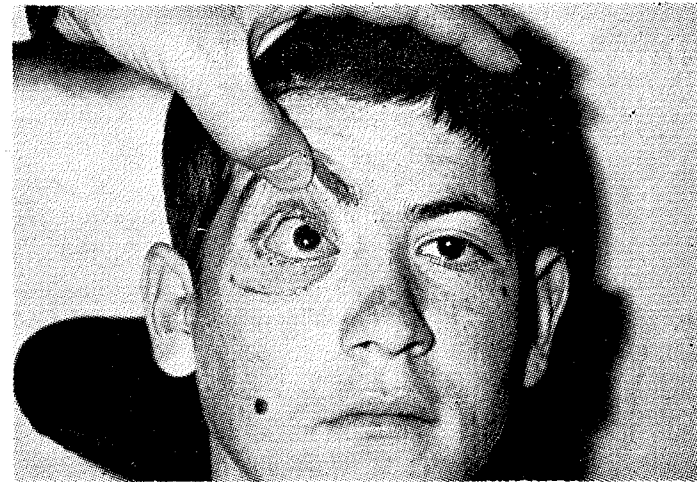


Fig. 2) Same patient after operation, exophthalmos has disappeared completely, but the eye is opened passively. Ptosis is present.

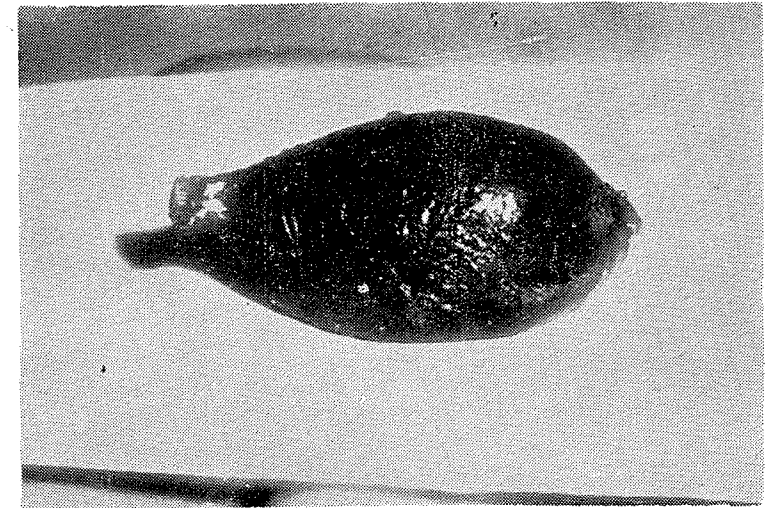


Fig. 3) Gross appearance of the tumour.

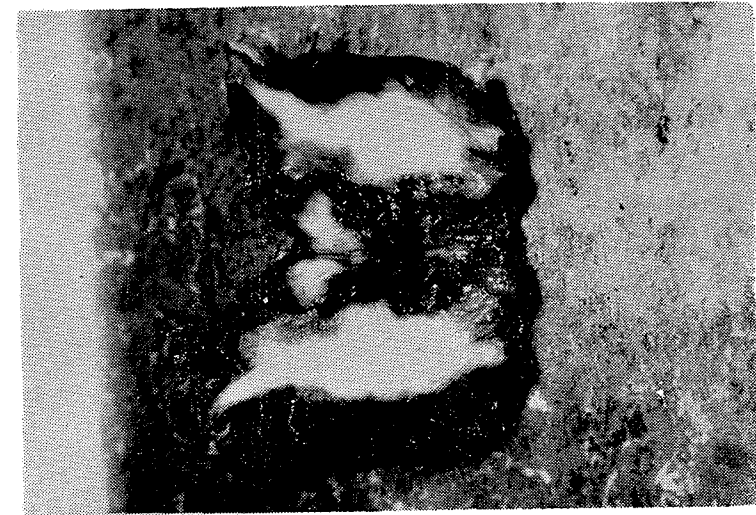


Fig. 4) Cut surface of the tumour showing the direction of optic nerve in its sheath and the fusiform appearance of the tumour.

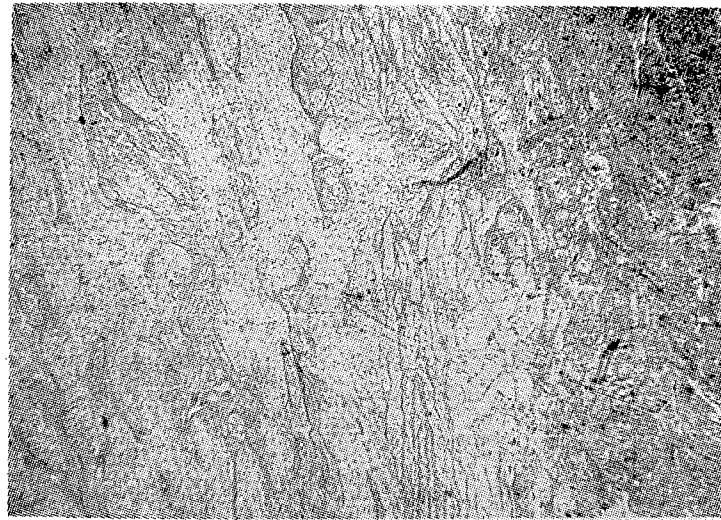


Fig. 5) Section of the tumour at peripheral region showing hyperplasia of nerve bundles on the left and the thickening of the pia-mater on the right.

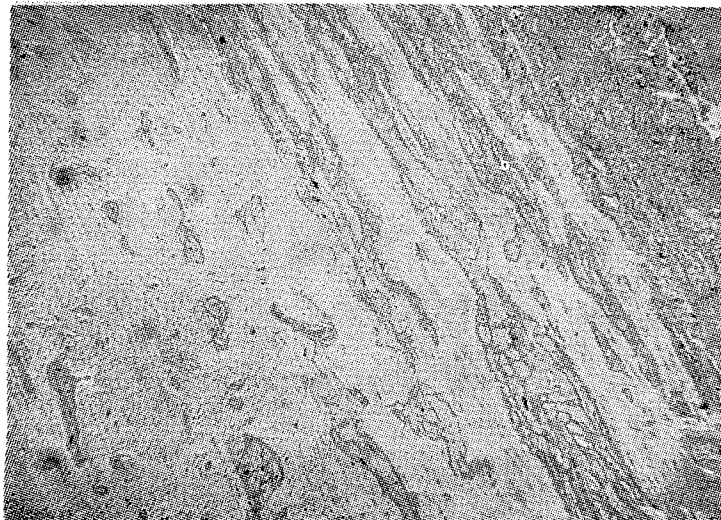


Fig. 6) Similar to figure 5, hypertrophy of the sheath being more obvious.

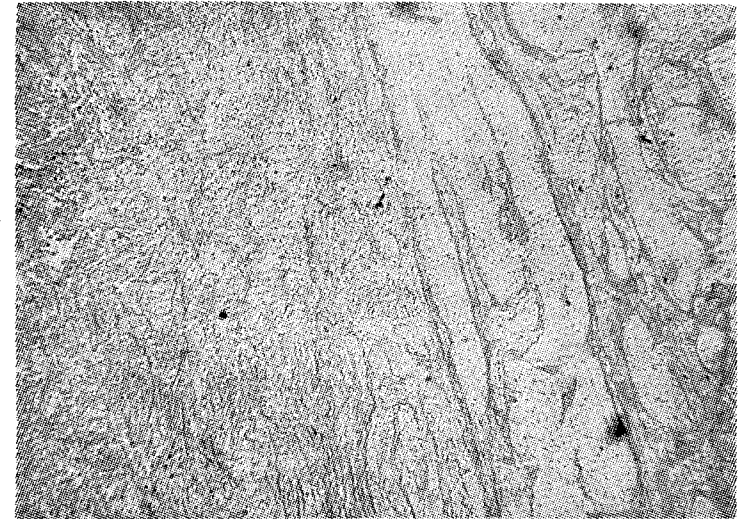


Fig. 7) Margin of the tumour showing thickening of the pia-mater (on the left side of picture).

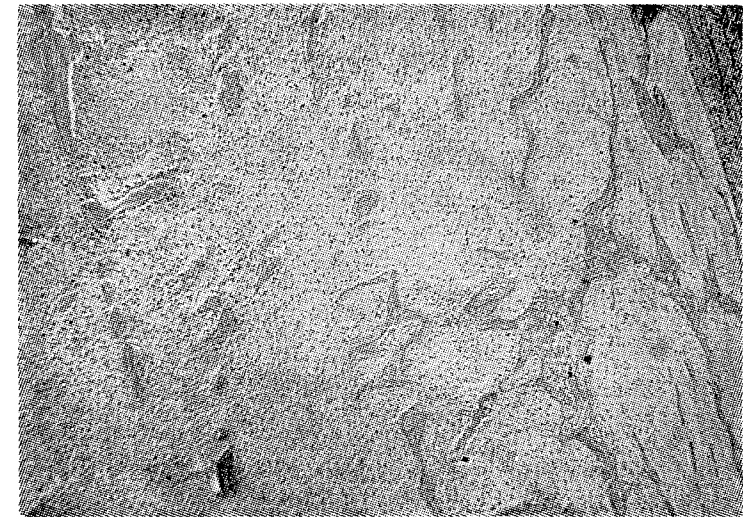


Fig. 8) Excessive encroachment of glial tissue into pia-mater leading to disintegration of the latter.

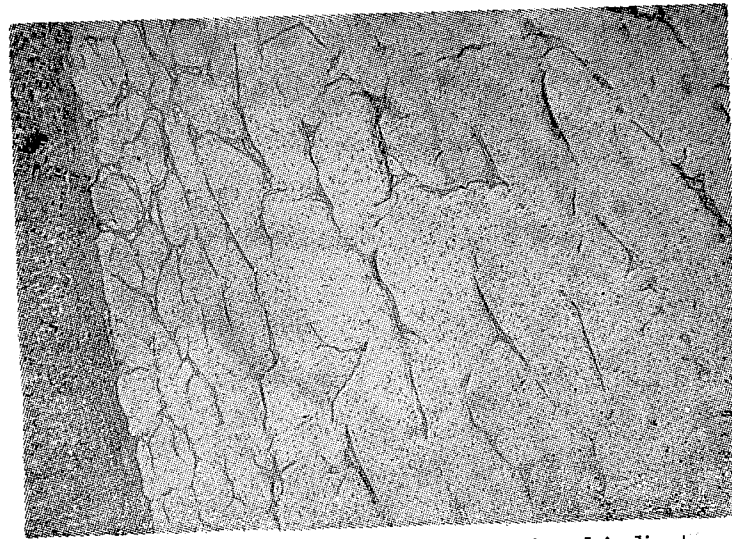


Fig.9) On the left side of the picture the dark band indicates pia-mater. On the right side, the nerve bundles are seen separated by fibres of pia-mater. Approaching the center the nerve bundles become wider and the pia-mater remnants are broken off.



Fig. 10) The state of nerve bundles is significant on the left side of picture while on the right it is quite obscure.

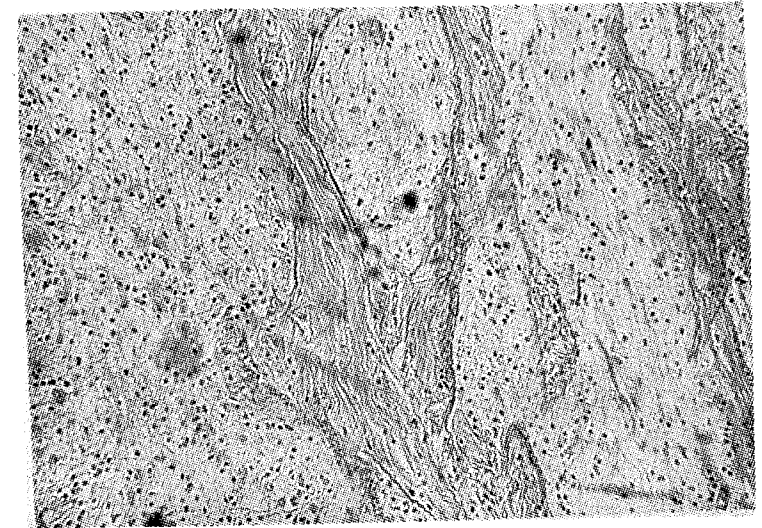


Fig. 11) Hyperplasia of glial tissue and the thickening of the pia-mater.

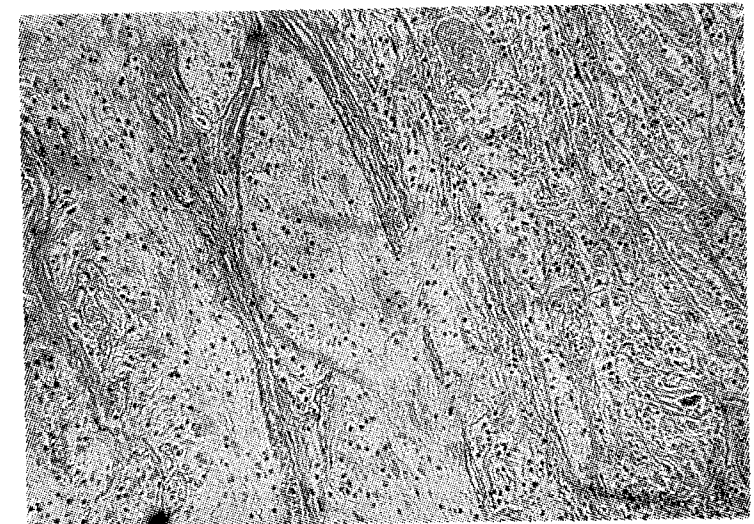


Fig. 12) Invasion by the glial tissue of the hypertrophied pia-mater (left part of picture).

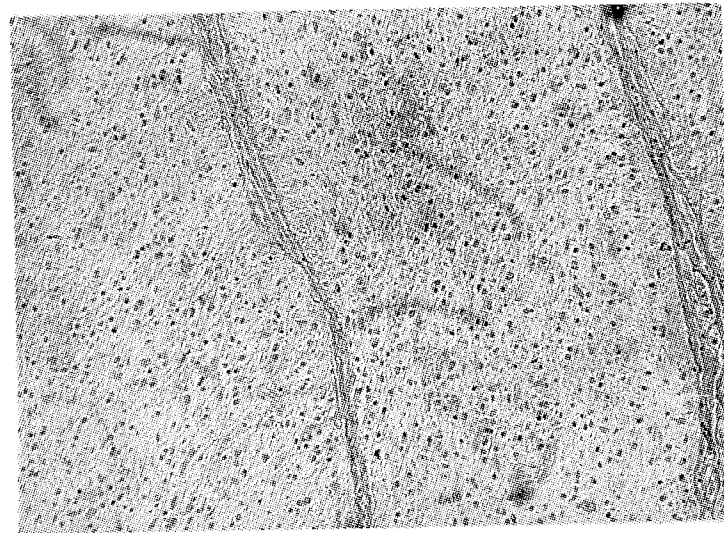


Fig. 13) Section of a nerve bundle displaying hyperplasia of glial tissue using moderate magnification. One can still recognize pia-mater process.

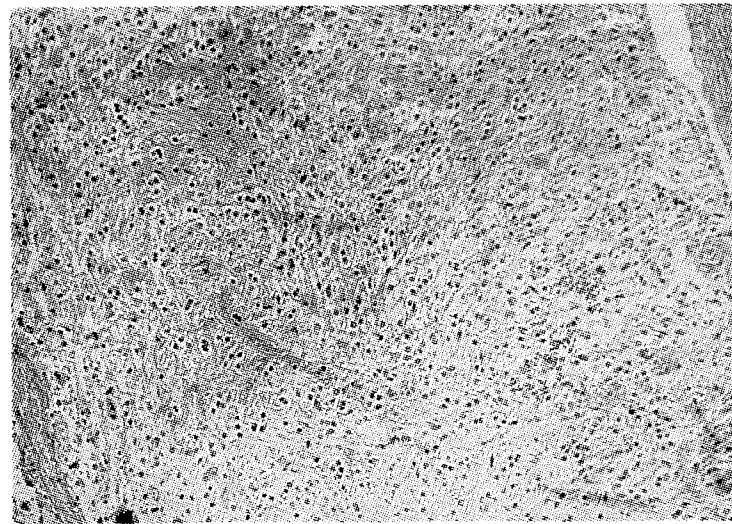


Fig. 14) Proliferation of oligodendroglial cells giving a honeycombed aspect to the tissue.

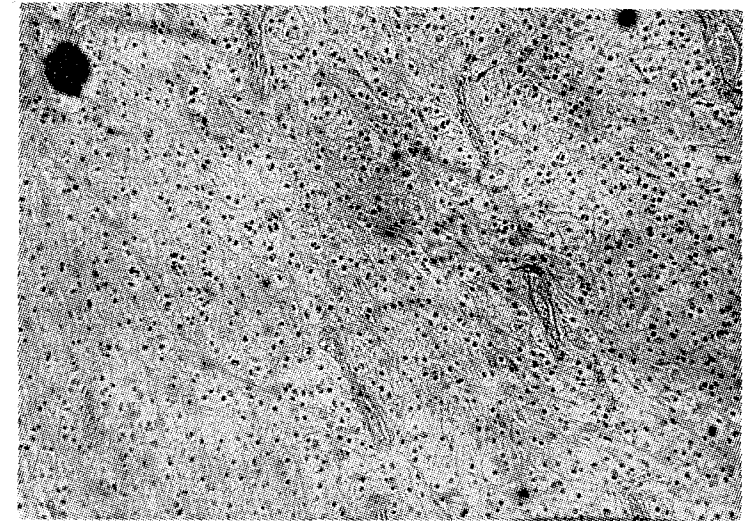


Fig. 15) Similar to figure 14, remnants of pia-mater process still recognizable.

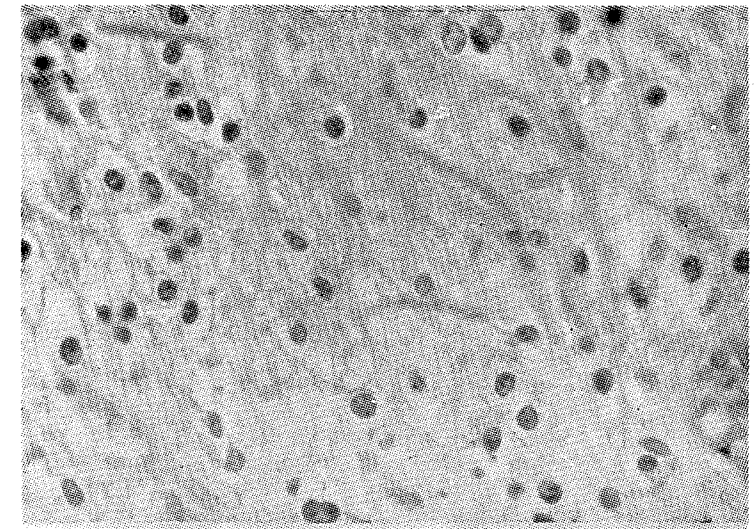


Fig. 16) Glial cells of oligodendroglial type are displayed on either side.

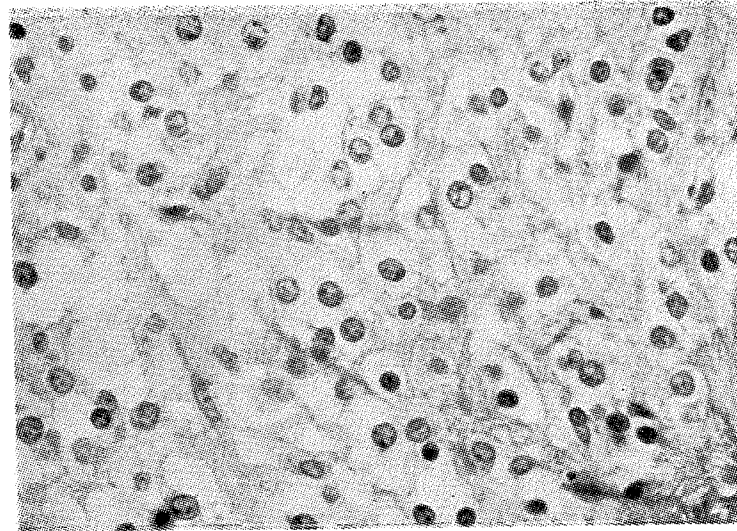


Fig. 17) A high magnification of figure 15. The characteristic honeycomb of oligodendroglia cells is clearly shown. The protoplasm just around the nuclei is clear. Between white and the darker peripheral protoplasm a distinct boundary is perceptible.

#### Diagnosis:

Oligodendrocytoma, a type of glioma of the optic nerve.

#### Summary

The case under consideration was a glioma of the optic nerve, one of the rarer tumors. Microscopically it was diagnosed as a oligodendrocytoma.

The patient, whose disease began before the age of puberty, complained of poor vision which in the course of five years resulted in blindness.

#### Résumé

Un cas très rare de gliome de nerf optique, avec le diagnostic microscopique d'oligodendrocytome est décrit.

La maladie avait commencé bien avant l'âge de puberté et le malade souffrait d'abord d'une vision pauvre qui s'est développée vers une cécité complète en cinq ans.

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