CASE NOTES

GLIOMA OF CHIASMA AND OPTIC NERVE*

BY

ALY MORTADA

Department of Ophthalmology, Faculty of Medicine, Cairo University, Cairo, Egypt

IN a case of orbital optic nerve glioma without intracranial extension, Davis (1940) removed the tumour through a skin incision at the lower orbital margin, cutting the optic nerve near the apex of the orbit and behind the globe and thus retaining the eye. Verhoeff (1922) found that, even when the tumour was partially removed by the orbital route leaving some intracranial portion, no orbital recurrence had been reported. When there was an intracranial prechiasmal extension, Dandy (1941), using a transfrontal approach, divided the optic nerve just anterior to the chiasma and immediately behind the globe to remove the tumour. If the chiasma was involved, Martin and Cushing (1923) avoided surgical intervention as the patient usually died within 48 hours in hyperpyrexia after the operation. In such cases, Katzin (1945), Taveras, Mount, and Wood (1956), and Walsh (1956) treated the tumour by radiation with good results.

Case Report

An 8-year-old boy (Fig. 1) had had gradual painless proptosis and diminution of vision of the left eye of one year's duration. There was no history of trauma. The parents were not consanguineous and did not show skin pigmentations or nodules.

FIG. 1.—Glioma of left optic nerve and chiasma in an 8-year-old boy, showing left proptosis.

Examination.—The general health was good. There were no cerebral symptoms. The skin of the back showed three small brownish patches, the largest 1 cm. in diameter, but no subcutaneous nodules. The pre-auricular, submaxillary, cervical, and other lymph glands in the body were not enlarged. The chest and abdomen showed no abnormality. The blood pressure, urine, and blood count were normal. The Wassermann reaction and tuberculin tests were negative. The cerebrospinal fluid pressure, cytology, and chemistry were normal.

The left eye showed a proptosis of 30 mm. (right eye 14 mm.). The eye deviated downwards and inwards with limitation of ocular movements in all directions. The lids and

* Received for publication August 3, 1961.

conjunctiva were normal. The pupil was dilated and inactive to direct light. The fundus showed post-papilloedemic optic atrophy. There was no perception of light. The ocular tension was normal. A mass was felt deep in the orbit above and below the globe.

Externally the right eye was normal, but the fundus showed primary optic atrophy. The visual acuity was 3/60, but there was no error of refraction. The temporal field loss denoted a chiasmal affection.

A postero-anterior skull *x* ray (Fig. 2) showed a very wide left optic canal and orbit. The left oblique skull *x* ray (Fig. 3) showed a very wide left optic canal denoting an intracranial tumour extension. The right oblique skull *x* ray (Fig. 4) showed the normal right optic canal. The lateral skull *x* ray showed normal sella turcica and clinoid processes and absence of suprasellar calcification.

FIG. 2.—Postero-anterior skull *x* ray, showing wider left orbit and left optic canal.

FIG. 3.—Left oblique skull *x* ray, showing wide left optic canal.

FIG. 4.—Right oblique skull *x* ray, showing normal right optic canal.

Treatment.—The child's parents refused transfrontal exploration, but as the chiasma was affected the orbital part of the tumour was removed for biopsy and treatment of the proptosis. The intracranial part of the tumour was treated by irradiation.

Through a lower fornix conjunctival incision, the posterior surface of the globe was found to be smooth and not infiltrated by the tumour. The optic nerve for 7 mm. behind the globe was not affected, but the rest of it was involved by the tumour which occupied the posterior half of the orbit. By blunt dissection with the little finger, the tumour was

easily separated from the four orbital walls. The optic nerve was cut by scissors 2 mm. behind the globe, and the tumour was pulled forwards and cut where it entered the optic foramen. Bleeding was not troublesome, and the eye was not removed. The wound healed quickly without complications.

The removed orbital part of the tumour (Fig. 5), which measured 2×3 cm., was covered with intact dura with a smooth surface; it was fusiform in shape, grey in colour, and of elastic consistency. Small cystic areas were seen in the middle of the tumour on section.

FIG. 5.—Orbital part of glioma of optic nerve and chiasma. The optic nerve portion is continuous with the tumour.

HISTOPATHOLOGICAL EXAMINATION.—This is a moderately cellular tumour. The cells are orientated in streams but there are no true palisades (Fig. 6).

FIG. 6.—Section of glioma of optic nerve, showing orientation of tumour cells in rows and parallel arrangement of glial fibres. $\times 80$.

The tumour cells are elongated and spindle-shaped, uni-polar or bi-polar, forming the so-called "spongioblasts" with processes, which are sometimes straight and sometimes corkscrewed, arising from each end (Fig. 7, opposite). The nuclei are oval or elongated with a moderate amount of chromatin. There are no mitotic figures. The tumour is rich in glial fibres which are arranged in parallel rows. Degenerated glial fibres taking the deep red eosin stain constitute the so-called Rosenthal fibres (Fig. 8, opposite). In parts the tumour is undergoing mucoid degeneration. The nuclei of degenerating uni- and bi-polar cells are round, simulating oligodendroglia nuclei; they are usually seen at the glial network intersections and not in the middle of the small compartments. There are no ganglion cells, rosette formations, multi-nuclear giant cells, haemorrhages, calcification, or fatty degeneration. The histopathological picture is consistent with spongioblastoma polare of the optic nerve.

The treatment of the intracranial part of the tumour was continued by radiation. After 3 years there was no recurrence of the proptosis (Fig. 9), development of cerebral signs, or deterioration of the vision or field of vision of the right eye.

FIG. 7.—Section of glioma of optic nerve. The tumour cells are elongated spindle-shaped uni- and bi-polar cells, the so-called "spongioblasts". The nuclei are oval or elongated with a moderate amount of chromatin. $\times 360$.

FIG. 8.—Section of glioma of optic nerve, showing degenerated glial fibres giving cone-shaped Rosenthal fibres. "Cytooid bodies of Verhoeff." $\times 360$.

FIG. 9.—Appearance 3 years after removing orbital part of tumour and treating intracranial part by irradiation. Note intact left eye and absence of proptosis.

Summary

The transfrontal approach to remove gliomata of the chiasma and optic nerve has a high mortality rate. In such cases the orbital part of the tumour may be removed through a lower fornix conjunctival incision, retaining the eye. In the case here reported the intracranial part of the tumour was treated by irradiation with good results.

I thank Prof. M. Hashim, Senior Professor of Pathology in the Faculty of Medicine, Cairo University, for his histopathological diagnosis.

REFERENCES

- DANDY, W. E. (1941). *Arch. Ophthalm. (Chicago)*, **25**, 191.
 DAVIS, F. A. (1940). *Ibid.*, **23**, 735.
 KATZIN, H. M. (1945). *J. Mt Sinai Hosp.*, **11**, 332.
 MARTIN, P., and CUSHING, H. W. (1923). *Arch. Ophthalm. (N.Y.)*, **52**, 209.
 TAVERAS, J. M., MOUNT, L. A., and WOOD, E. W. (1956). *Radiology*, **66**, 518.
 VERHOEFF, F. H. (1922). *Arch. Ophthalm. (N.Y.)*, **51**, 120.
 WALSH, F. B. (1956). *Amer. J. Ophthalm.*, **42**, 347.