OPTIC NERVE GLIOMA*†

BY

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ONE of the most characteristic features of optic nerve glioma is that it "is a disease almost entirely of early age and adolescence" (Spaulding, 1958). 75 per cent. occur in patients under the age of 10 years, the peak incidence is 2 to 6 years (Chutorian, Schwartz, Evans, and Carter, 1964), and the eldest so far reported is 60 years of age (Bucy, Russell, and Whitsell, 1950).

The following case is reported to demonstrate the difficulties in diagnosis of optic nerve glioma presenting in the older age groups.

Case Report

A 79-year-old woman, who was known to have hypertension, had complained of intermittent episodes of vertigo for 2 years, and 3 days before admission she complained of black and white patterns in the right half of the visual fields which lasted 5 minutes and were followed by mild scalp pain and progressive deterioration of vision.

Examination.—There was slight bilateral exophthalmos, right 23 mm. and left 21 mm. The pupils were large (4.5 to 5 mm.) and reacted sluggishly to direct and consensual light. A right temporal hemianopia was noted. The visual acuity was 3/60 in the right eye and 6/36 in the left. The fundi were normal apart from mild arteriosclerotic changes.

Investigations.—Hb 86 per cent., white blood count 5,500 cells/cm., polymorphs 57 per cent., lymphocytes 35 per cent., monocytes 8 per cent., erythrocyte sedimentation rate 40 mm./1st hr corrected to 33 mm. (Wintrobe.)

X rays of the skull, optic foramina, and pituitary fossa were normal.

The cerebrospinal fluid was under normal pressure and its constituents were normal. Biopsy of the temporal artery showed no evidence of arteritis.

Progression to almost complete blindness was rapid over 2 weeks, during which time measurement of the degree of exophthalmos was unchanged but the optic discs, especially the right, became pale and atrophic. In spite of the negative biopsy, treatment with 20 mg. prednisolone three times daily was instituted and the erythrocyte sedimentation rate fell to 10 mm.

After the sudden development of right hemiplegia the patient died. *Post mortem* examination revealed a large left-sided cerebral haemorrhage together with a right optic nerve glioma.

Microscopical Examination of the Eyes (Prof. N. Ashton).—Apart from *post mortem* degenerative changes and senile peripheral cystic degeneration of the retina, both globes are histologically unremarkable. Both optic nerve heads, however, show a moderately dense infiltration with glial cells which appear to have accumulated at the laminae cribrosae,

* Received for publication June 12, 1966.

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and longitudinal and transverse sections of the optic nerve show invasion by an astrocytoma. In the right nerve the growth occupies one half its circumference in a localized area near the centre of the nerve segment and is associated with necrosis at the anterior and posterior margins of the growth. The cell type is a small spindle astrocyte which extends diffusely into the nerve. The left nerve is also involved, but much more diffusely, and there is no clear line of demarcation between the growth and normal nerve. Areas of necrosis are also present.

Pathological Diagnosis.—Astrocytomata invading both optic nerves.

Discussion

The sudden onset of poor vision in an old person, together with a raised erythrocyte sedimentation rate, suggests a diagnosis of giant cell arteritis, but the absence of headache and the progressive visual deterioration, the negative arterial biopsy, and the failure to respond to steroid therapy made this unlikely.

Because of the right temporal field defect, optic nerve compression was considered to be the most likely diagnosis.

The commonest causes of optic nerve compression have been listed by Richardson and Rose (1965) as:

- (1) Pituitary tumour;
- (2) Craniopharyngioma;
- (3) Optic nerve meningioma;
- (4) Optic nerve glioma;
- (5) Metastases;
- (6) Ophthalmic artery aneurysms.

The normal skull *x* ray was against a diagnosis of pituitary tumour (which produces an abnormal sella in over 90 per cent. of cases) or craniopharyngioma (in over 50 per cent. of these cases calcification of the tumour is noted on *x* ray). The rapidity of onset of visual deterioration in the absence of *x*-ray changes suggested metastases. In a younger patient, further neurological investigation, such as carotid arteriography and encephalography, would almost certainly have been carried out. Even when these are negative, exploratory craniotomy may be necessary (Rose and Richardson, 1966). Failing vision occurs earlier in glioma than meningioma (Hope-Robertson, 1949) but, although optic nerve gliomata are five times more common than meningiomata in children (Duke-Elder, 1940), in adults meningioma is commoner (Hudson, 1912). Extra-ocular muscles tend to be involved early with meningioma; the limitation of movement of the globe which occurs with a glioma is presumed to be mechanical and due to the bulk of the tumour, there being no case recorded where glioma of the nerve has invaded or enveloped the ocular muscles or nerves to produce ocular palsies (Tym, 1961).

Optic nerve glioma commonly presents as a painless, non-pulsatile progressive proptosis (Duke-Elder, 1940) which may be asymmetrical producing, for example, a displacement outwards and downwards of the globe which tends to retain mobility. There is only slight limitation of movement. A reduction of visual acuity is noticed at a later date than proptosis, and this is especially true of children, in whom the acuity is difficult to assess; loss of vision in the affected eye can occur so early in life that binocular vision does not develop and squint results.

The pupil may be dilated, reacting slowly to direct and briskly to consensual light (if the opposite optic pathway is unaffected).

Ophthalmoscopic examination may reveal

- (1) Optic atrophy;
- (2) The tumour itself (Wilson and Farmer, 1940);
- (3) Unilateral papilloedema usually due to venous obstruction;
- (4) Bilateral papilloedema usually due to raised intracranial pressure from ventricular obstruction (in cases of chiasmal glioma).

Radiology.—When a glioma involves the intracanalicular portion of the optic nerve, x rays of the skull show enlargement of the optic canal (van der Hoeve, 1925). An optic foramen of 6.5–7 mm. diameter or 1 mm. larger than the contralateral foramen is considered abnormal. (Unlike gliomata, meningiomata usually show hyperostosis around the optic foramen.) Decalcification of the posterior wall of the orbit and lesser wing of the sphenoid has been recorded with glioma as has calcification in the tumour itself (Cushing, 1930).

If the tumour extends into the chiasm there may be:

- (1) Flattening of the sella
and/or
- (2) Erosion of the anterior wall of the sella and under-surfaces of the anterior clinoid processes.
- (3) A “J” shaped, “slipper”, or “shoe” sign produced by pressure of the tumour at the lateral aspect of the chiasmal sulcus beneath the anterior clinoid process, the sulcus becoming obvious and hollowed out.

This last appearance is frequent in optic nerve glioma and may be so marked that it becomes “gourd-shaped”, extending from the body of the sella under the anterior clinoid process. (Gourd-shaped sellae are also found in mongolism and Hurler’s syndrome.) If the ventricular system is blocked, there may be radiological signs of raised intracranial pressure, such as a silver-beaten appearance of the skull and widening of sutures. Carotid angiography may show displacement of the ophthalmic artery in the orbit (Grino and Billet, 1949), while orbital pneumatography may outline the intra-orbital tumour (Dubilier, von Gal, Freemond, and Evans, 1956). Air encephalography and air or Myodil ventriculography may demonstrate filling defects in the third ventricle due to glioma.

Pathology.—The commonest site of glioma of the optic nerve is the optic foramen 10 mm. behind the globe (Duke-Elder, 1940) at the point where the central retinal artery enters the nerve (Wolff, 1940). Histologically, the glioma is composed of astrocytes and oligodendrocytes, one cell type usually predominating. In the case reported here the predominant type was the small spindle astrocyte.

Whereas optic nerve glioma is usually slow growing and associated in at least half the cases with generalized neurofibromatosis (Tym, 1961), the tumour in our case grew rapidly and there was no evidence of von Recklinghausen’s disease.

Summary

An optic glioma in a woman aged 79 years is reported. The rarity of this type of tumour at this age is emphasized and the clinical and radiological features discussed.

It is a pleasure to thank Dr. Alexander Kahan of St. James's Hospital, Balham, for allowing us to publish this case. Our thanks are also due to Prof. N. Ashton for the pathological report.

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