Management of optic nerve gliomas

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SUMMARY Seventeen patients thought to have orbital optic nerve gliomas when first seen have been reviewed after up to 12 years. Enlargement of the optic canal was present in 15 of the 16 patients examined, but this finding was unreliable as an indicator of the posterior extent of the tumour. Nine patients had a stable course with little change over a period of up to 8 years; there was optic atrophy in all and neurofibromatosis was relatively common (7/9). Eight patients showed progressive enlargement of the tumour; 6 had swollen discs, and the incidence of neurofibromatosis was relatively low (3/8). The optic nerve was excised in 7 of the latter group. Biopsies of the optic nerve taken from the region of maximal enlargement were difficult to interpret and unhelpful in planning management. Radical surgery should be reserved for the minority of patients in whom there is progressively enlarging tumour without evidence of chiasmal involvement.

Gliomas of the optic nerve are relatively uncommon lesions, and their natural history and management are the subject of considerable debate. In 1969 Hoyt and Baghdassarian¹ suggested that these tumours behaved as hamartomas and concluded that the management of patients with them should be conservative. We have seen 17 patients with optic nerve glioma in the past 12 years, and it has become clear that an appreciable proportion of them have had a progressive course. As a result our attitude towards treatment has changed. In this paper we record our experience of these patients and outline our present approach to their management.

Patients and methods

The diagnosis of optic nerve glioma was made in 17 patients out of more than a thousand with orbital disease seen at the Orbital Clinic at Moorfields Eye Hospital between 1968 and 1980. The diagnosis was initially made clinically on the basis of poor vision in association with proptosis and optic atrophy or swelling of the optic disc, and radiographic evidence of enlargement of the optic foramen. In 1 patient the optic foramen was normal; 1 child was too young to co-operate for optic foramen views; and in 1 patient there was no proptosis during the period of observation. Enlargement of the optic nerve was,

however, demonstrated in these 3 patients. Further details are given in the section on 'Results'. In patients seen later in the series enlargement of the optic nerve on CT scans was required for the diagnosis. The clinical diagnosis was confirmed histologically in 7 cases.

In all patients a detailed clinical history was obtained and a full ocular and neurological examination was carried out. If the patient was old enough to co-operate, the visual fields were charted; in some patients pattern visual evoked potentials (VEP) were recorded. The neurologist, in addition to assessing the state of the central nervous system, checked for stigmata of Von Recklinghausen's neurofibromatosis, using the criteria of Crowe and Schull,² and interviewed the family to see if there was evidence of neurofibromatosis in the relatives. Radiographic examination of the patients included routine skull films together with optic canal views in each case. In many patients axial hypocyclusoidal tomograms were obtained.

Patients with a slowly progressive or static lesion (as monitored by visual acuity, field charts, and degree of proptosis) were seen at least yearly by the ophthalmologist and the neurologist. Patients with a more progressive lesion were usually seen every 4 weeks.

Initially a biopsy of the optic nerve was performed in 4 patients in whom there was evidence of recent tumour enlargement. This was done to differentiate optic nerve gliomas from optic nerve meningiomas. However, without complete excision of the optic nerve the histological interpretation of the excised

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tissue proved extremely difficult. Biopsies were not therefore performed on the later cases.

If the clinical course indicated progressive enlargement of the tumour, the patient was referred to our neurosurgical colleague so that the lesion could be approached transcranially and the whole of the optic nerve from the globe to chiasm explored. In most of these cases the optic nerve from the globe to the chiasm was excised. If the chiasm was involved, surgical resection was not performed, but a shunt was inserted if hydrocephalus was present.

Results

Seventeen patients with an orbital optic nerve glioma diagnosed by the criteria defined above were included in the study. The details of their age and sex distribution are shown in Table 1. Ten of the 17 patients had Von Recklinghausen's neurofibromatosis. In 9 of these 10 café-au-lait spots were the only stigmata of the disease. The exception was a patient who presented at 38 years of age having had poor vision in one eye since at least the age of 11; she had multiple cutaneous mollusca fibrosa.

The presenting symptoms were particularly interesting (Table 2). Fourteen patients had a proptosed eye which was noticed by parents. Eight of these 14 patients had no complaint other than proptosis; only 4 patients had complained of poor vision in the affected eye, while 2 had a convergent squint in addition to proptosis. Of the remaining 3 patients 2 were noted to have a squint and diminished vision when examined in a school clinic. One patient, the oldest in the series, had noticed a pro-

Table 1 *Orbital optic nerve glioma, clinical data*

Total patients	17	
Sex	Male 4	Female 13
Involved orbit	Right 9	Left 8
Age at presentation	Mean	20.4 years
	Median	5.5 years
	Range	2-38 years
Age at onset of symptoms	Mean	4.5 years of age
	Range	0.5-18 years of age

Table 2 *Initial complaint*

Proptosis	8
Proptosis and poor visual acuity	4
Proptosis and squint	2
Squint and poor visual acuity	2
Poor vision	1

Table 3 *Clinical presentation*

	Present	Absent
Proptosis	15	2
Poor vision	17	—
Relative afferent pupillary defect	17	—
Optic disc swelling	6	—
Optic atrophy	11	—
Enlarged optic canal	15	1
Neurofibromatosis	10	7

Table 4 *Visual acuity on presentation*

Visual acuity	Number of patients
6/18	1
6/24	4
6/36	4
C.F.	1
PL	3
NPL	4

PL=perception of light. NPL=no perception of light.

gressive deterioration in the vision of the affected eye but did not notice that the eye was very slightly proptosed.

All 17 patients had decreased visual acuity at the time of presentation (Table 3), ranging from 6/18 to no perception of light (Table 4), with a relative afferent pupillary defect on the affected side. Eleven patients had a pale atrophic optic disc; 1 of these patients had opticociliary shunt vessels. Four patients had a swollen optic disc, and 1 patient had a mass extending forwards into the globe and involving the whole of the optic disc. One patient with 6/24 vision in the affected eye had normal discs on presentation, but optic atrophy was noted shortly thereafter.

Fifteen of the 17 patients had proptosis ranging from 2 mm to 8 mm (mean 4 mm). Of the 2 patients who had no proptosis initially 1 developed it later. The other remained nonproptosed (4 years follow-up); enlargement of the optic nerve was confirmed by CT scan.

On initial x-ray examination 15 of 16 patients had an enlarged optic foramen on the affected side (Fig. 1).

One patient, who presented at age 5 with a 3-year history of squint and was seen on routine school visual examination to have poor vision in that eye (perception of light), had an enlarged optic nerve on B-scan ultrasonography but no proptosis or enlargement of the optic foramen. During the next 2 years she developed a proptosis of 3 mm, which has remained static for 12 months; radiographs of

Fig. 1 Oblique radiograph of skull showing rounded enlargement of the optic canal typical of optic nerve glioma.

her optic foramen remain normal. One other patient was too young to co-operate for optic foramen x-ray views but had an optic nerve glioma verified histologically. CT scans always showed the position of the mass, but it was impossible to differentiate between the tumour and any adjacent arachnoid hyperplasia. In several cases thickening of the contralateral clinically normal nerve could be seen (Fig. 2).

CLINICAL COURSE

The clinical course of the patients was variable. Nine patients had a slow indolent course during several years' follow-up (mean 4.2 years, range 1–8 years). Three of these patients were eventually lost to follow-up, but in each case there had been no significant change in the symptomatology prior to the child and parents defaulting. All but 3 patients had symptoms for longer than 1 year before presentation (mean 2.5 years, range 4 months–9 years). The mean age at presentation of this group was 6.1 years (range 2–15 years). Vision when first seen ranged from 6/18 to no perception of light. All patients but 1 had proptosis. In all cases the optic nerve head was atrophic and flat, with shunt vessels present in 1 case. Seven of the 9 patients had neurofibromatosis.

Thus the patients in the above group showed little change in the level of visual acuity and field of vision. Two patients improved. In 1 child the

vision in the affected eye improved slightly with patching, and in another case the proptosis resolved during a 3-year period.

Eight patients showed evidence of growth of the tumour and 7 had the affected nerve excised. The mean age of presentation of this group was 5.8 years (range 2.5–20 years), with a relatively brief duration of symptoms (mean 1.6 years—range 2 months to 8.5 years). Vision on presentation was poor, ranging from 6/24 to no perception of light. The optic disc in 6 patients was swollen; in another 2 the disc was atrophic. One of these patients had shunt vessels on the disc. Three patients had neurofibromatosis. The individual clinical details of patients in this group are worth recording.

CASE REPORTS

Patient 1. A 3½-year-old girl with neurofibromatosis developed a progressive left proptosis for 4 months before attending the Orbital Clinic. The left eye was blind and proptosed 4 mm. The optic disc was swollen and invaded by a cream coloured mass which extended forwards into the vitreous. Radiographs showed an enlarged left optic foramen, and B-scan ultrasound showed a solid retrobulbar mass with extension into the optic nerve head. Because of progressive proptosis the optic nerve was explored through a medial conjunctival incision 1 year after the child first attended the clinic. Incision of the dura released a large quantity of mucinous material.

The biopsy showed an intact dural sheath with an underlying area of meningeal hyperplasia. CT scans showed enlargement of the whole of the optic nerve from the globe to the chiasm. The child was referred to our neurosurgical colleague and the affected optic nerve removed from behind the globe to the chiasm, which was not involved, through a frontal craniotomy. Histologically the tumour was a grade 1 astrocytoma. No tumour was present at the cut surface of the intracranial portion of the nerve. Intractable glaucoma developed, and the globe was enucleated 5 months later. Nine months after the craniotomy the child's prosthesis became displaced by an enlarging orbital mass; biopsy of this lesion showed recurrence of the glioma. The mass continued to enlarge and the orbit was eventually partially exenterated. She is alive with no sign of a recurrence of the tumour 2 years after her last operation.

Patient 2. An 8-year-old girl with a 4-month history of progressive left proptosis was referred to this clinic. The affected eye was proptosed 3 mm and convergent 40°. Vision was reduced to 6/36, the left optic disc appeared hypoplastic, with swelling of the nerve head in the upper nasal quadrant. There were no stigmata of neurofibromatosis. Radiographs showed enlargement of the left optic canal. The pattern VEP showed a normal response from the right eye. The optic nerve was explored through a medial transconjunctival approach; incision of the dural sheath released mucinous material. A portion of the sheath together with the underlying tissue was excised and was reported as showing meningotheelial cells, fibrous tissue, and a few well differentiated glial cells. During the next 3 years the visual acuity in the left eye further decreased to 6/60. The proptosis increased to 10 mm. CT scans showed an ill-defined expansion of the left optic nerve, but the intracranial scan was normal, as was an air encephalogram. The intracranial and intracanalicular portion of the optic nerve were removed through a frontal craniotomy. Histopathological examination showed a normal optic nerve with an extensive collar of meningotheelial cells and intermingled psammoma bodies. No evidence of glial hyperplasia or astrocytic glioma was seen throughout the intracanalicular or intracranial portions of the optic nerve. She was diagnosed as having a primary optic nerve meningioma. Three months later a large tumour mass continuous with the optic nerve was excised through an anterior orbitotomy, and the optic nerve was divided flush with the globe. Histological examination showed a typical grade 1 astrocytic glioma of the optic nerve with extensive meningotheelial cell proliferation and numerous psammoma bodies. The child is alive and well with

Fig. 2 Axial CT scan showing left optic nerve glioma (arrowed). The right optic nerve is diffusely enlarged; the right visual field and acuity were normal.

no sign of a recurrence after 2 years.

Patient 3. A girl presented at the age of 5½ years with a 4-month history of a left divergent squint and a 2-month history of proptosis. Her blind left eye was proptosed 3 mm, divergent 25°, and the optic disc was swollen. The right eye was normal in all respects with an unaided vision of 6/6; the VEP was normal from this side. There were no signs of neurofibromatosis. Radiographs showed an enlarged left optic foramen. Eight months later the divergent squint was surgically corrected and the optic nerve explored. A biopsy of the nerve showed hyperplastic meningeal tissue. Four months later the VEP showed evidence of involvement of the nasal fibres from the right eye, though a field defect could not be detected by perimetry. Twelve months later she experienced severe left hemicranial headaches with nausea and vomiting, which after several months subsided spontaneously. Three years after her initial presentation perimetry showed a right superior temporal quadrantic field defect. By this time the left eye was proptosed 9 mm. CT scans showed a large left optic nerve mass with involvement of the chiasm. An air encephalogram showed a moderate degree of symmetrical hydrocephalus, with the third ventricle displaced upwards and almost obliterated by a large suprasellar mass. A left frontal craniotomy was performed, during which it was found that the tumour involved both optic nerves together with the chiasm. Posteriorly the tumour surrounded the left internal carotid and anterior cerebral arteries. Portions of the tumour were removed for histological examination. Examination showed the tumour to have the features of a grade 1 astrocytoma. 5500 rads of radiotherapy

were given to the chiasm and the left orbit. Subsequently the shunt for hydrocephalus was removed. She remains alive and well without progression of the proptosis or visual field deficit for the past 4 years.

Patient 4. A girl was seen at the age of 2½ years with a 2-month history of proptosis and poor vision in the left eye. Vision in the affected eye was reduced to 6/36; the eye was proptosed 3 mm, and the optic disc was swollen. No signs of neurofibromatosis were seen. X-rays showed an enlarged left optic foramen and CT scans enlargement of the intra-orbital part of the left optic nerve. A biopsy of the optic nerve through a medial conjunctival approach was performed and was reported as showing astrocytic proliferation with occasional Rosenthal fibres. It was not possible to tell whether the biopsy was of the nerve itself or of an extension of tumour tissue into the sheath; meningeothelial cells were not present. During the ensuing 6 months the proptosis increased 6 mm, and the child was transferred to the care of our neurosurgical colleague. An air encephalogram showed no evidence of intracranial extension. The whole of the optic nerve from the globe to the chiasm was removed through a transfrontal craniotomy. Histologically the tumour was an astrocytoma grade 1. At operation it was thought that the tumour did not involve the chiasm, but microscopically the cut end of the intracranial portion of the nerve showed gliomatous tissue. Apart from postoperative ptosis the child is well 6 months later.

Patient 5. A girl aged 7½ years was brought to the clinic having noticed poor vision and progressive protrusion of the left eye for the previous 2 months. Her left eye was proptosed 4 mm, the vision was reduced to 6/36, and the optic disc was swollen. Radiographs showed enlargement of the left optic canal and CT scans enlargement of the optic nerve to the orbital apex. VEPs were obtained from the right eye and were reported as being within normal limits. The patient was referred to our neurosurgical colleague for removal of the optic nerve because of the rapidity of the growth of the tumour. The nerve was totally excised, but there was histological evidence of glioma at the cut end of the intracranial part of the optic nerve. Histologically the tumour was a grade 1 astrocytoma. The child is well with no signs of a recurrence of the tumour 9 months after surgery.

Patient 6. The father of a 6-year-old girl had noticed that his daughter's left eye had become progressively prominent for 6 months prior to consulting an ophthalmologist. The eye was blind and proptosed 8 mm. Ocular movements were full. However, the left optic disc was swollen, with

associated peripapillary haemorrhages and exudates. A VEP from the right eye and the visual fields were normal. The child was referred to our neurosurgical colleague, who explored the chiasm but found that the glioma extended into the chiasm. Surgery was confined to obtaining a biopsy, which confirmed the diagnosis of glioma, astrocytoma grade 1. During the ensuing 15 months the amount of proptosis has increased slightly, and it is probable that a portion of the orbital part of the glioma will have to be resected in order to preserve the globe.

Patient 7. A 9-year-old girl was referred to an ophthalmologist. Her parents had noticed a slight right divergent squint since she was 6 months old. At a routine school examination it was found that the vision in her right eye was poor. At her initial examination the right visual acuity was 6/24, with an associated slight right divergent squint. She was initially treated with partial occlusion of the left eye, but after 18 months it was noticed that the vision in her right eye had deteriorated, and she could only count fingers in the upper half of the visual field. At the same time she was found to have a pale right optic disc, and the eye was proptosed 3 mm. She was subsequently referred to the Orbital Clinic, where x-rays of her skull showed enlargement of the right optic canal. There was no evidence of neurofibromatosis. During the next 5 years vision in her right eye deteriorated to the appreciation of hand movements at 0.25 metre and the proptosis increased to 6 mm. The visual field in the left eye was examined at regular intervals and the VEPs were obtained. Four years after her initial presentation the VEP showed evidence of a temporal field defect. Twelve months later there was definite evidence of a central temporal half field defect in the right eye by conventional perimetry. CT scans showed enlargement of the right optic nerve with evidence of chiasmal involvement. The child was transferred to Maida Vale Hospital for a pneumo-encephalogram, which revealed a suprasellar mass and hydrocephalus. The patient died after the procedure. Permission for a necropsy was declined.

Patient 8. A 20-year-old female student had noticed blurring of right vision for 18 months with prominence of the eye for 12 months before consulting an ophthalmic surgeon. During the ensuing year vision in the affected eye deteriorated, and the degree of proptosis increased. A neurosurgeon from another centre performed a transfrontal approach to the right orbit but was unable to find any abnormality. After an ineffective 6-week course of systemic steroids the patient was referred to this hospital. Her right eye was proptosed 5 mm and displaced downwards 3 mm; upward movement of the globe was selectively restricted. The optic disc was pale

and flat, though a year previously definite swelling had been noted. There were numerous opticociliary shunt vessels. A CT scan showed gross enlargement of the optic nerve immediately behind the globe. Subsequently the orbital portion of the optic nerve containing a large grade 1 astrocytoma was excised through a lateral orbitotomy approach. During a 4-year follow-up there has been no recurrence of the tumour.

Discussion

The outstanding finding of the present investigation is that our patients fell into 2 fairly distinct groups with differing clinical course. Nine patients had an indolent course with little change in the size and extent of the tumour. In the remaining 8 patients the tumours progressively enlarged.

The age distribution of the patients at presentation was similar in the 2 groups, but the progressive group had a shorter history and a relatively rapid increase in proptosis during the period before and soon after the initial consultation. Other evidence of rapid tumour enlargement was found in the ocular fundus. In 6 patients the optic disc was swollen when the patient was first seen. This was in contrast to the more stable group of patients in whom none had evidence of optic disc swelling, though there was optic atrophy in all cases. Of particular interest was patient 7, in whom there was evidence of a quiescent glioma for 9 years before the eye became proptosed. Five years later she had developed a field defect in the contralateral eye and the proptosis had increased considerably. This case was exceptional, but it does illustrate that enlargement of a glioma of the optic nerve can occur in patients who have been clinically quiescent for some years.

There was a significant difference in the incidence of neurofibromatosis in the 2 groups. The overall incidence was rather higher (10/17, 63%) than in previous reports,^{1,3-8} as low as 10%⁶ and as high as 50%.⁸ Only 3 of 8 patients in the progressive group had neurofibromatosis compared with 7 out of 9 with quiescent tumours. This observation accords with the views of Martin and Cushing⁹ and Lloyd⁸ and Klug,¹⁰ who reported that patients with optic nerve glioma in association with neurofibromatosis had a more benign course than patients without this association. The explanation for the wide divergence in figures for the overall incidence of neurofibromatosis is uncertain, but 3 factors are likely to contribute: (1) the relatively small number of patients in most series; (2) the difficulty of recognising the minimal clinical signs of neurofibromatosis; and (3) the delay in the development of the

stigmata of the disease in some children.

The recognition that there exist 2 groups of patients with orbital optic nerve glioma has important implications for management. Many surgeons have adopted a conservative policy following Hoyt and Baghdassarian's conclusion that gliomas of the optic pathways are indolent and slow growing and that for practical purposes they can be regarded as hamartomas, most of them requiring no active intervention. This view has been supported by a number of authors subsequently.¹¹⁻¹⁴ There is however good neuropathological evidence that at least some optic nerve gliomas are true tumours with a potential for local invasion.¹⁵ This observation, together with our experience that nearly half our patients showed clear evidence of progression, has led us to the conclusion that the potential for enlargement of optic nerve glioma in childhood is not infrequently realised, and that radical surgery should be considered for patients in which it is.

It is clear that if the chiasm is already involved when the patient presents a radical cure is not possible. In these circumstances surgery should be confined to the treatment of complications such as disfiguring proptosis or hydrocephalus, as Hoyt and Baghdassarian suggest.¹ The first step in the assessment of the patient must therefore be to try to determine whether or not the tumour is confined to the orbit. We have used 3 approaches to this problem.

(1) *Radiological examination of the optic canals.* There was evidence of enlargement in 15 out of 16 of our patients. In several of these patients, however, surgical exploration showed that the glioma did not extend into the enlarged canal. As others have noted,¹² the widened canal could be associated with dural thickening or arachnoid hyperplasia. The size of the optic canal thus gives little indication of the posterior extent of an optic nerve glioma.

(2) *CT scanning.* The advent of high resolution CT scanning in both axial and coronal planes has enabled tumours arising from the optic nerves to be recognised much more readily than was possible previously. Again, however, it is not possible to distinguish between gliomatous tissue and meningeal hyperplasia. Moreover, enlargement of the chiasm by tumour may still be difficult to detect even with the very high resolution scanners now available. Pneumoencephalography is more reliable, but as Anderson and Spencer¹⁶ have reported, and our cases 4 and 5 confirm, the absence of macroscopic enlargement of the chiasm does not exclude involvement by tumour.

(3) *Visual field examination.* The most sensitive methods for detecting involvement of the nerve fibres from the other eye are perimetry and VEP

examination. Halliday *et al.*¹⁷ have shown that it is possible to detect abnormalities in the pattern VEP before a field defect is demonstrable clinically. In 2 of our patients (patients 6 and 7) there was electrophysiological evidence of a temporal field defect 6 and 12 months before it could be demonstrated by conventional perimetry. However, even this sensitive technique failed to show chiasmal involvement in 2 of the cases (4 and 5), in whom the face of the optic nerve severed at its junction with the chiasm showed histological evidence of tumour. These observations indicate that abnormal tissue may be present in the visual pathways without the clinician being aware of its true extent.

Having demonstrated the presence of an optic nerve tumour and obtained some idea of its extent, the ophthalmologist must then decide whether it is appropriate to biopsy the tumour in order to establish the diagnosis. Our initial policy was not to biopsy small, apparently indolent lesions. However, when there was evidence of recent tumour enlargement and there was severe visual impairment, we obtained tissue from the nerve in the region of maximal swelling in an attempt to distinguish between optic nerve meningioma, which has a reputation in children for aggressive growth and local invasion¹⁸ and therefore requires early radical excision, and glioma. In the event, as others have found,⁶ it was difficult to interpret the biopsy findings. In 4 of the 5 biopsies there was arachnoid hyperplasia compatible with either glioma or primary optic nerve meningioma. In the fifth biopsy it was impossible to tell whether the tissue came from the nerve itself or from an extension of the tumour into the sheath. Because we have not found biopsy helpful in planning management we have abandoned it as a routine, and we restrict surgical intervention to total excision of tumours showing evidence of progressive enlargement or to the symptomatic relief of complications.

Finally there is the question of surgical approach to the expanding tumour. In our own material, as in that of others, enlargement was produced by meningeal hyperplasia and the production of mucoid material containing tumour astrocytes as well as by glial proliferation. Since it is impossible clinically or radiologically to distinguish the contribution of each to any particular tumour, we have managed all patients in the same way. If there is no evidence of chiasmal involvement, the patient is referred to our neurosurgical colleague, who explores the chiasm through a frontal approach. If inspection confirms the absence of chiasmal swelling, the affected optic nerve is sectioned at its junction with the chiasm. The optic canal is unroofed and the nerve excised to the posterior surface of the

globe. A number of authors have stated that partial removal of these lesions is all that is needed and that the chances of enlargement of the residual tumour are extremely low.^{11,19} The importance of complete excision of the nerve, however, is emphasised by the course of case 1, in which a partial removal was followed by rapid expansion of the orbital stump.

It is inevitable that in some patients tumour will be present at the chiasm despite the absence of swelling or of preoperative evidence of damage to fibres from the other eye. This was the case in 2 patients (4 and 5) of our 4 patients in the progressive group (cases 1, 2, 4, and 5), who are known to have had no macroscopic evidence of enlargement of the chiasm. The state of the chiasm in case 8 is not known. Anderson and Spencer¹⁶ have suggested that in such patients, as in those with preoperative evidence of chiasmal enlargement, the abnormal tissue may be present in the chiasm from the start, rather than having extended there from a primary orbital tumour. Our observations shed no light on this suggestion. However, given that there is unequivocal clinical and radiological evidence of tumour enlargement in some patients, it is reasonable to suppose that the chiasm might become secondarily involved by tumour tissue extending along the nerve in the same way that tumours of similar histological type, such as the cerebellar astrocytomas of childhood, extend in white matter elsewhere in the central nervous system.¹⁵ Cases 1 and 2 of the 4 patients in whom there was no macroscopic evidence of chiasmal involvement had no tumour tissue at the cut surface of the intracranial optic nerve. In these 2 patients total excision of the tumour will have removed the possibility of further involvement of the visual pathways from the orbit. They are being closely followed up to determine whether tumour subsequently develops in the chiasm. If it does, the case for an attempt at radical surgical cure will be greatly weakened.

The correct management of the child with optic nerve glioma remains a difficult problem. Our experience of the past 12 years has led us to move from a conservative to a more radical position, though as a result of our present review we think it likely that only a minority of patients should have surgery. We now believe that parents should be told that their child has a tumour of the optic nerve and that the appropriate method of treatment will be determined by the progress of the condition. In about half the patients there will be no deterioration, and management will be conservative. In the remainder progressive deterioration will suggest that the tumour is extending, and for these patients we will advise radical surgery provided there is no radiological, clinical, or VEP evidence of chiasmal

involvement. When the chiasm is involved, we will advise against surgery unless orbital or intracranial complications develop, which should then be managed on their merits.

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