OPTIC PATHWAY GLIOMAS IN CHILDREN:
EXPERIENCE AT KING KHALID UNIVERSITY HOSPITAL, RIYADH

ABDULHAKIM JAMJOOM, FRCS Ed(SN); ZAIN ALABEDEEN JAMJOOM, MD;
NAIM-UR-RAHMAN, FRCS Ed(SN); MOHAMMED AL-SOHAIBANI, FACP

A review of all 8 cases of childhood optic pathway glioma that were treated at the
neurosurgical unit of King Khalid University Hospital in Riyadh between 1984 and 1991 is
presented. The series is unusual, in that all the tumors were chiasmal in location, and none
of the children had any signs of von Recklinghausen’s neurofibromatosis. Computerized
tomography, which was performed in all children, showed extension of the tumor beyond the
confines of the chiasmal region in all cases. In keeping with the location and the large size
upon presentation, we found a high incidence of associated raised intracranial pressure,
hydrocephalus, and total visual failure. Histological confirmation of a benign astrocytoma
was available for 7 cases (88%); in three children, partial excision of the tumor had been
performed, while in four, a biopsy only had been done. Six children received radiotherapy.
At a mean follow-up period of 23 months, all children were alive, the tumor appeared smaller
on CT-scan in the three children in whom partial excision had been performed, and in two
patients there was some improvement in vision.

Key words  astrocytoma, benign - childhood, brain tumors - computerized tomography -
hydrocephalus - optic chiasmal glioma - visual failure

OPTIC PATHWAY GLIOMAS account for 5% of
childhood brain tumors. In the past, these
lesions were thought to be hamartomatous, or
reflect degenerative gliomatosis. However, there
is little doubt now that they are true gliomas,
pilocytic in nature and occasionally showing
mucinous or cystic changes. Although
histologically all similar, optic gliomas may be
divided into two groups according to location:
 ante rior tumors that involve an optic nerve, the
chiasm, or a combination of optic nerve(s) and
chiasm but do not compromise any structures
beyond the chiasm; and posterior tumors that
involve the optic chiasm and extend beyond it to
invade the regions of adjacent structures. The
differing location causes significant differences in
clinical features, management and prognosis.

The natural history and optimal method of
treatment of optic gliomas remain
troversial. To our knowledge, no reports
regarding the features of this disease in Saudi
Arabia have been published to date. This article
constitutes a review of the modes of presentation
and the management of all children with optic
gliomas, as seen over a seven-year period in a
major neurosurgical unit in Riyadh, capital of
Saudi Arabia. We also give a brief report of the
short-term progress in these children.

Materials and Methods

The clinical records of all patients aged ≤ 13
years, who had been treated for any type of optic
pathway glioma at the neurosurgical division of the Department of Surgery, King Khalid University Hospital, Riyadh, between 1984 and 1991 were reviewed. The 8 cases that were found represented 13% of all childhood brain tumors treated at this institution during that period, and were all found to be chiasmal optic gliomas. Histological confirmation of the diagnosis that had been made on the grounds of clinical features and appearance on computerized tomography (CT-scan) was available for 7 cases. (One further child seen at this hospital, in whom a unilateral intraorbital optic nerve glioma had been treated by surgery and radiotherapy elsewhere, was not included in our study.)

The following data were extracted from the hospital records:

A) At presentation: age, sex, nationality, nature and duration of symptoms, clinical features including the Karnofsky score, and clinical stigmata of von Recklinghausen’s neurofibromatosis (VRN), if any. Karnofsky scored the patients’ degree of disablement according to the following code:

100 = Normal, no evidence of disease
90 = able to carry on normal activity, minor symptoms
80 = normal activity with extra effort
70 = not able to carry on normal activity, but able to live independently of assistance
60 = some assistance required but able to care for most needs
50 = requires considerable assistance and frequent medical care
40 = disabled, requires special assistance and care
30 = severely disabled
20 = very ill, active support treatment required
10 = moribund, fatal processes are rapidly progressing

Raised intracranial pressure was diagnosed clinically on the basis of a history of headache, vomiting, drowsiness, and other features. No lumbar punctures were performed.

Visually evoked potentials (VEP) were recorded, and all children were subjected to CT-scan, both without, and with intravenous contrast enhancement, to determine location and extent of the tumor and presence or absence of hydrocephalus.

B) During progress: any surgery or radiotherapy performed, and duration of follow-up.

C) Status, at the latest recording, regarding: clinical findings and visual acuity, particularly also reassessment of the Karnofsky score, and CT-scan findings.

Blindness was diagnosed if the pupil did not react to light and no VEP could be demonstrated for that eye.

Results

Four boys and four girls, whose ages ranged from 5 months to 13 years (mean 5.8 years) were included in the review. Four patients were Saudis, three were from Yemen, and one was from Kuwait. Symptoms had been noted for 1.5 months to 4 years (median 6 months) prior to presentation. The clinical features evident at the time of presentation are summarized in Table 1. No evidence of VRN could be found in any of our patients.

In 6 children (75%), CT-scan showed a globular (Fig. 1) or multilobulated (Fig. 2) lesion in the region of the optic chiasm, with no radiographically obvious involvement of the optic nerves or tracts. In one child (case 2), a globular chiasmal lesion, extending forwards in the form of tubular thickening of both optic nerves was seen (Fig. 3), while in another, case 7, symmetrical involvement of the optic tracts (Fig. 4) was found in addition to bilateral optic nerve extension of the mainly chiasmal glioma. All tumors were found, on radiological investigation at the time of first presentation of the patient, to be large and extending well beyond the confines of the chiasm to invade the

Table I. Clinical features at the time of presentation, in 8 children with (chiasmal) optic glioma.

<table>
<thead>
<tr>
<th>Clinical feature</th>
<th>Number of patients</th>
<th>(%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Visual failure</td>
<td>7</td>
<td>(88)</td>
</tr>
<tr>
<td>unilateral</td>
<td>3</td>
<td>(38)</td>
</tr>
<tr>
<td>bilateral</td>
<td>4</td>
<td>(50)</td>
</tr>
<tr>
<td>Optic atrophy</td>
<td>10</td>
<td></td>
</tr>
<tr>
<td>unilateral</td>
<td>6</td>
<td>(75)</td>
</tr>
<tr>
<td>bilateral</td>
<td>4</td>
<td>(50)</td>
</tr>
<tr>
<td>Raised intracranial pressure</td>
<td>7</td>
<td>(88)</td>
</tr>
<tr>
<td>Long tract signs</td>
<td>3</td>
<td>(38)</td>
</tr>
<tr>
<td>Papilledema</td>
<td>2</td>
<td>(25)</td>
</tr>
<tr>
<td>Growth retardation</td>
<td>2</td>
<td>(25)</td>
</tr>
<tr>
<td>Epilepsy</td>
<td>1</td>
<td>(13)</td>
</tr>
</tbody>
</table>
CHIASMAL GLIOMAS IN CHILDREN

Figure 1. CT-scan, with contrast enhancement, showing a large, globular optic chiasmal glioma (case 1).

Figure 2. CT-scan, with contrast enhancement, showing an extensive, multilobulated optic chiasmal glioma (case 3).

Figure 3. CT-scan, with contrast enhancement, showing an optic chiasmal glioma with the characteristic tubular thickenings of both optic nerves (arrow heads). The lesion is seen to be causing obstructive hydrocephalus (case 2).

Figure 4. CT-scan, with contrast enhancement, showing symmetrical involvement of both lateral geniculate bodies by the optic chiasmal glioma (arrow heads) (case 7).
region of the third ventricle in 7 (88%), the sella turcica in 4 (50%), the diencephalon in 5 (63%), the temporal lobe in 4 (50%), and the frontal lobe in 3 patients (38%).

There was radiographical evidence of an obstructive hydrocephalus in all patients except case 5 (i.e. in 88% of our patients) (see also Fig. 3) and all these children were managed with construction of a cerebrospinal fluid (CSF) shunt to the peritoneum.

Total surgical excision of the tumor was not attempted in any patient. Partial excision was performed in 3 children (38%), while in 4 (50%) craniotomy with only a biopsy of the tumor was performed. In case 7, where there was bilateral and symmetrical involvement of optic nerves and tracts on CT-scan, diagnostic biopsy was judged to be perfluous. However, the patient's hydrocephalus was treated by construction of a CSF shunt.

Histological examination of the seven tumors that were subjected to surgery demonstrated a well differentiated, benign astrocytoma in each case.

Postoperative radiotherapy, in a dose of between 4,500 and 5,500 cGy, was given to 6 patients. The parents of one child refused radiotherapy, while another patient, whose vision is good in both eyes, is being closely followed.

The duration of follow-up varied from 6 months to 4 years, with a mean of 23 months. Findings at the latest follow-up examination are summarized in Table 2. There was no mortality. Vision improved slightly in two children (25%) and remained unchanged in the other six. Follow-up CT-scans demonstrated a reduction in size of the tumor in only those three children where partial excision had been performed (see also Figs. 5 A and B). Only two patients, however, are able to carry on normal activities, four require some assistance, and two are severely disabled.

Discussion

Alvord and Lofton(5) reviewed the literature up to 1988 and found that of the total of 623 reported cases of optic glioma, only 239 (38%) were posterior. This may reflect a reporting bias in favour of anterior tumors. All our cases (100%) were posteriorly situated, but this is not surprising, as it may be expected that a neurosurgical unit would deal predominantly with such tumors while anterior optic gliomas would be more likely to be treated in ophthalmological units.

There has been considerable debate regarding the natural history of optic gliomas. In adults, these tumors are recognized to sometimes be highly malignant.(8) In children, however, it is now believed that cases with optic gliomas fall into two distinct groups: one with an indolent course and little change in the size of the tumor over time, and another where there is progressive tumor enlargement.(4,5) The large size of the tumors, at presentation, in our patient collective, after a relatively short duration of symptoms (mean: 6 months), would speak for an aggressive nature.
while histological appearance, available in 7 of 8 cases, was judged benign in every case. Our follow-up period has been too short to make it possible to arrive at any conclusion in this matter, although mortality from the glioma itself tends to occur, if at all, within the first few years after diagnosis.\(^{(9)}\)

None of our children demonstrated any clinical stigmata of VRN, while in other countries, 12–50% of cases with optic glioma were accompanied by features of VRN.\(^{(1,2,9)}\) A higher incidence of VRN has been found in patients with optic glioma that exhibit a stable course than in those with the aggressive type of tumor.\(^{(4,9)}\) Similarly, patients with untreated chiasmal optic gliomas who additionally suffer from VRN have a somewhat better prognosis than those without VRN, while in patients that were treated with \(\geq 4,500\) cGy irradiation, the prognosis was found to be identical, regardless of the presence or absence of VRN.\(^{(5)}\)

The large size of the tumors at presentation in our children explains the unusually high incidence of hydrocephalus (88%). The overall incidence of hydrocephalus associated with all cases of posterior optic glioma reported in the literature up to 1988 was merely 23.4%.\(^{(5)}\) The high incidence of raised intracranial pressure, pyramidal signs, and epilepsy, as well as visual impairment and optic atrophy in our group, which has also been seen in other series of chiasmal gliomas\(^{(9)}\) may be explained by the delay in presentation of these children, and also the difficulty in detecting and assessing visual disturbances in young children. Since there is no unequivocally effective treatment that can be offered for this type of optic glioma, this delay is not quite as grave as in optic nerve gliomas where early diagnosis gives the managing physician a better chance of differentiating, according to progress, between the aggressive and the benign type of tumor, possibly making the difference between life and death for certain patients.\(^{(10)}\) Nevertheless, since these patients usually present first to ophthalmologists, it is of utmost importance that the differential diagnosis of optic glioma be kept in mind by practitioners of this specialty, so that appropriate investigations (CT-scan or MRI) will be carried out as soon as possible after presentation. Since we can do relatively little about the delay in presentation to medical attention of these patients, it behoves us to at least keep the time interval between presentation and diagnosis as short as possible.

Our children were investigated by computerized tomography (CT-scan) only. Fletcher and coworkers\(^{(11)}\) stated that there are three diagnostic
patterns of chiasmal glioma on CT-scan: tubular thickening of the optic nerve(s) and chiasm, a suprasellar tumor with contiguous optic nerve extension, and a suprasellar tumor with optic tract involvement. We found these features in only 2 of our 8 cases; the other six displayed a large suprasellar mass on CT-scan, which could equally well have been of hypothalamic origin. The diagnosis was, however, confirmed at surgery when the abnormal chiasm was inspected and biopsied. Magnetic resonance imaging (MRI), which is very useful in such cases, is unfortunately not available in our hospital.

The role of surgery for posterior optic gliomas is not clear. Total excision of such tumors is not recommended as it is technically difficult, being associated with a high mortality, and may sacrifice years of good vision. In fact, total excision was documented for only one of the 97 surgically treated cases of posterior tumor documented in the literature up to 1988. While a biopsy is very useful in confirming the diagnosis, occasionally, in the presence of VRN and/or typical radiological features, it may not be necessary.

The efficacy of radiotherapy in the treatment of chiasmal optic glioma is not well established. Most clinicians dealing with these cases agree that once there is clinical and/or radiological evidence of tumor growth, radiotherapy should be given. It must be remembered that this treatment is not without serious side effects, particularly in children.

For reasons already mentioned, two of our children (cases 2 and 8) have not received radiotherapy yet. As far as the brief follow-up allows for any conclusion, this does not appear to have affected their prognosis as in both cases the tumors have remained unchanged at 12, and 18 months, respectively. In those children that did not undergo any surgical resection apart from a small biopsy but who were treated with radiotherapy (cases 1 and 4), the tumors similarly appear unchanged at 12, and 24 months follow-up, respectively. In cases 4 and 6, however, there has been some improvement in the Karnofsky score when compared to pretreatment findings. Recently, some success was reported after use of interstitial radiotherapy, but further evaluation is needed.

None of our cases were treated with chemotherapy, and this has been employed for too short a time, and on too varied a population, to give assurance of any efficacy in the treatment of optic glioma.

In our series of children, the assessment of visual evoked potentials (VEP) was limited to the confirmation of blindness. It is recognized that VEPs can be useful in the monitoring of patients, since stability of serial VEPs confirms absence of extension of a tumor over time, and thus a benign course, encouraging the managing physician to refrain from aggressive intervention.

Although the indifferent progress that we recorded is not unexpected with posteriorly located tumors, earlier diagnosis would be desirable in order to permit monitored supervision, and leave room for possible intervention at a stage when neurological deficits have not yet become too crippling. This is particularly true for optic nerve gliomas, i.e. optic gliomas confined to an optic nerve, where excision, in those relatively uncommon cases exhibiting aggressive growth, may give a chance of a cure. The short follow-up periods of our patients, and the small number of cases in our series make it impossible to reach any valid conclusions regarding the efficacy of the various treatment modalities used by us, and we give the data merely for the sake of completeness.

Conclusions

We are aware that the reported series is small and not unbiased. There are many neurosurgical units in our city, and there is no organized referral pattern. Our data do, however, demonstrate that both the diagnosis, and the referral to a tertiary care center are much delayed for children suffering from chiasmal optic gliomas in Saudi Arabia; large lesions at presentation, with considerable visual and other neurological impairment are therefore the rule. We strongly recommend that specialized radiographic investigation be performed as soon as possible in children that exhibit visual impairment and defects in pupillary reaction, so that diagnosis may be facilitated at a stage when irretrievable optic and cerebral damage has not yet progressed to a debilitating level.
References


Editorial Comment: Dr. Abdulhakim Jamjoom and his colleagues contribute interesting observations concerning relatively large intracranial gliomas presenting with significant findings of neurosurgical importance, seen at a neurosurgical unit. It is very important clinically to separate the cases presented by Jamjoom et al. from the much more frequently encountered, relatively small and often nonprogressive gliomas of the anterior optic nerve, which present to ophthalmologists because of proptosis or decrease in vision. Conclusions about appropriate management which one might draw from the former patients (with neurosurgical presentation) should not be applied to the latter group (with ophthalmological presentation). In fact, it is important to try to assess the biological activity of the more common latter group, reserving removal of the optic nerve (which immediately blinds the eye, obviously) for those cases in which either history or physical examination findings indicate likelihood of vigorous biologic activity.

The far more common indolent forms of anterior optic nerve glioma presenting to the ophthalmologist and not to the neurosurgeon should clearly be managed with heavy emphasis on follow-up examination and periodic neuroimaging.

The authors’ conclusions could be interpreted as suggesting that optic nerve gliomas should be diagnosed as early as possible and removed quickly. This advice, which may be applicable for larger, posterior, aggressive lesions presenting with hydrocephalus to the neurosurgeon, is harmful if applied to the more common indolent ophthalmologic-presenting cases.

Robert S. Hepler, MD
Medical Director
King Khaled Eye Specialist Hospital
Member of the Editorial Board
Saudi Journal of Ophthalmology
Riyadh, Saudi Arabia.