Tumors of the optic nerve and optic chiasm

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In a series of 20 cases of glioma of the optic nerve, approximately 25% were
successfully treated surgically. This series also confirms the observations of
Taveras, et al., (1956) that 25% of patients thus treated obtain considerable
additional improvement in vision after radiation therapy, and that another 10%
derive minimal benefit.

Neoplasms of the optic nerves occur
in one of every 100,000 patients, ac-
cording to Arkhangelsky.1 Astrocy-
tomas of the optic nerve are the second most
common orbital tumor in the Mayo Clinic.
In a review2 of 186 cases recorded up to
1960, 51 meningiomas were listed as con-
trasted with 26 astrocytomas. Astrocytomas
most frequently afflicted children 15 years
old or younger. Of a total of 40 orbital
tumors, 20 were gliomas of the optic nerves.
Dandy3 classified gliomas of the optic appa-
tratus in two groups: those primarily in-
volving the optic nerves and those involving
the chiasm diffusely. The presenting symptoms
and signs thus vary, depending on the loca-
tion of the tumor.

Symptoms and Signs
In the series reported by Dodge, et al.,4
in 1958, loss of vision occurred in 50% of
the instances of gliomas of the optic nerves
and in 91% of the chiasmal tumors. Prop-
tosis accompanied all the gliomas of the optic
nerves but only 18% of the diffuse tumors.
Strabismus was caused by 58% of the an-
terior tumors and by 35% of the diffuse
lesions. Unilateral papilledema was present
in 58% of gliomas of the optic nerves, but
papilledema was associated with only 32%
of the diffuse tumors and in that relationship
invariably was bilateral. Pallor of the optic
discs accompanied 25% of the unilateral
gliomas of the optic nerves, but 62% of the
diffuse neoplasms. Headache or pain in the
eyes was relatively common, occurring in
25% of the patients who had tumors of the
optic nerves and 47% of those who had
chiasmal tumors. Motor symptoms did not
afflict patients who had tumors of the optic
nerves, but were observed in 12% of those
who had chiasmal lesions. Convulsions oc-
curred in 9% of the patients with chiasmal
tumors but not in those who had lesions of
the optic nerves. Metabolic disturbance oc-
curred only among those patients who had
the diffuse type of lesion; this complication
was observed in 26%.

Roentgenographic Findings
Martin and Cushing5 described the “pear-
shaped” sella which is formed in the presence
of diffuse gliomas of the optic chiasm. Schu-
ster and Westberg6 found no roentgenological
changes in the sella in half of their cases; the
other half had the abnormal pear-shaped
sella. Two thirds of their cases had enlarged
optic foramina without cortical destruction.

J. Neurosurg. / Volume 33 / October, 1970

439
Fowler and Matson\(^4\) emphasized that normal foramina vary from 4.1 to 4.65 mm in diameter. Cerebral angiography was rarely performed in our series. Pneumoencephalography was necessary in only a few instances. The diagnosis usually was established by the funduscopic and ophthalmologic examinations and studies of the visual fields, considered in association with roentgenograms of the head including tomographic and special orbital and optic foraminal views.\(^5\)

**Therapy**

When the diagnosis has been established, we prefer operative exposure of the tumor. A preliminary steroid preparation is administered routinely. The tumors are approached transcranially. If the diffuse type of lesion is encountered, a biopsy from it is analyzed immediately. If the tumor is producing obstruction of the third ventricle, the lateral ventricle is entered transcorpically and the septum pellucidum opened, thus creating a temporary ventricular stoma for purposes of decompression. If this stoma ceases to function, a ventriculocisternal shunt is carried out by means of Torkildsen’s technique. This maneuver was necessary in three of our 20 patients. Irradiation therapy is administered in the postoperative period.

When the tumor is confined to the optic nerve, the nerve and tumor are removed from the globe to the optic chiasm by excising the roof of the orbit and the roof of the optic foramen. If the chiasmal end of the nerve is microscopically free of tumor tissue, irradiation therapy is unnecessary. In one case we believed we successfully removed an astrocytoma at the junction of the left optic nerve, chiasm, and left optic tract. Because of the uncertainty of total microscopic eradication of the neoplasm, however, the patient received irradiation postoperatively. This patient has shown no evidence of recurrence of the tumor in 12 years.

**Results**

Fowler and Matson\(^4\) reported 13 patients: four had intraorbital gliomas and nine had intracranial gliomas. There were two postoperative deaths. They considered that five were cured by surgical removal of the lesions: the extent of their follow-up studies ranged from 1 month to 8 years. Taveras, *et al.*,\(^8\) emphasized the importance of roentgen therapy; four of their 17 patients achieved improvement in visual acuity after irradiation. None of Fowler and Matson’s patients achieved improvement in visual acuity that could be attributed to irradiation therapy.

**Current Series**

It therefore seemed advisable to review the condition of the 20 patients operated on by one of us (MacCarty)\(^5\) with particular emphasis on the operability of the lesions and the effects of irradiation therapy. Our series consisted of nine male and 11 female patients 2 to 51 years old (Table 1). The stigmata of von Recklinghausen’s disease were present in three males and four females. Five patients had tumors involving only the optic nerve, and 15 had diffuse chiasmal tumors. All the neoplasms were grade 1 or 2 astrocytomas.

**Operative Results.** Surgical removal was accomplished, apparently successfully, in six instances (five tumors of the optic nerve and one situated at the junction of the left optic

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**TABLE 1**

*Summary of 20 cases of astrocytoma of the optic nerve and chiasm*

<table>
<thead>
<tr>
<th>Data</th>
<th>No. of Cases</th>
</tr>
</thead>
<tbody>
<tr>
<td>Optic involvement:</td>
<td></td>
</tr>
<tr>
<td>nerve only</td>
<td>5</td>
</tr>
<tr>
<td>chiasm</td>
<td>15</td>
</tr>
<tr>
<td>Von Recklinghausen’s</td>
<td></td>
</tr>
<tr>
<td>disease:</td>
<td></td>
</tr>
<tr>
<td>male</td>
<td>3</td>
</tr>
<tr>
<td>female</td>
<td>4</td>
</tr>
<tr>
<td>Tumor removal:</td>
<td></td>
</tr>
<tr>
<td>total</td>
<td>6</td>
</tr>
<tr>
<td>subtotal</td>
<td>14</td>
</tr>
<tr>
<td>Operative deaths</td>
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</tr>
<tr>
<td>Cases irradiated</td>
<td>16</td>
</tr>
<tr>
<td>Follow-up (11 months to 12 years):</td>
<td></td>
</tr>
<tr>
<td>alive</td>
<td>17</td>
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<tr>
<td>dead</td>
<td>2</td>
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<tr>
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<tr>
<td>Vision:</td>
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<tr>
<td>improved</td>
<td>5</td>
</tr>
<tr>
<td>stable</td>
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</tr>
<tr>
<td>presumed dead</td>
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</tr>
</tbody>
</table>

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MacCarty, Boyd and Childs

*J. Neurosurg. / Volume 33 / October, 1970*
Optic nerve and chiasm tumors

Fig. 1. Visual fields of 38-year-old man with a diffuse glioma. Improvement in the right visual field and some deterioration in the visual field and central vision of the left eye took place immediately after operation. His visual status has remained stable for 12 years.

Fig. 2. Visual fields of 9-year-old boy with grade 2 astrocytoma. Improved central vision was attributed to irradiation therapy. Complete bitemporal hemianopsia persisted.
Fig. 3. Visual fields of 16-year-old boy with grade 2 astrocytoma. Although central vision was essentially normal pre- and postoperatively, bitemporal hemianopsia cleared after irradiation therapy.

Fig. 4. Visual fields of 17-year-old girl with grade 1 fibrous astrocytoma. The principal improvement in this patient's visual status attributable to irradiation therapy was abatement of the defect in the visual field of the right eye.
Optic nerve and chiasm tumors

**FIG. 5.** Visual fields of 51-year-old man with grade 1 astrocytoma. Improvement in central vision and visual fields was attributable to irradiation therapy.

**FIG. 6.** Visual fields of 29-year-old woman with grade 2 astrocytoma improved after irradiation therapy.
nerve, chiasm, and left optic tract). There were no deaths in the hospital. The survival period as ascertained by recent review ranges from 11 months to more than 12 years. Seventeen patients are living, two have died, and one is presumed to have died.

Visual Status. In five instances an optic nerve was removed; there was normal vision and visual fields in the contralateral eye. One patient, a 38-year-old man, underwent surgical removal of a "diffuse" glioma with resultant immediate improvement of the right visual field and some deterioration of the visual field and central vision of the left eye. His status has remained stable for 12 years (Fig. 1).

Five patients for whom craniotomy and biopsy of the chiasmal tumors were done, followed by irradiation therapy, obtained improvement of vision. The radiation dosage ranged from 3000 to 5000 rads administered through multiple small fields aimed at the site of the lesion. In one case (Fig. 2) preoperative vision was 20/400 on the left and 20/200 on the right and improved to 20/50 on the left and to 20/25 on the right. Complete bitemporal hemianopsia persisted. In another case (Fig. 3) preoperative and postoperative central vision was essentially normal bilaterally, but bitemporal hemianopsia cleared after irradiation therapy. The improvement has persisted for 6 years, but there has been no recent examination. The third patient (Fig. 4) in this group experienced no significant improvement in central vision, which had been 20/20 on the left both preoperatively and after irradiation therapy, while vision on the right was 20/80 preoperatively and 20/70 after irradiation therapy. However, the defect in the visual field of the right eye abated appreciably (Fig. 4). The fourth patient had normal central vision on the left, before and after therapy, impaired vision preoperatively on the right with subsequent improvement from 20/400 to 20/100, and improved visual fields (Fig. 5). The last patient in this series exhibited improvement only in the visual fields after treatment (Fig. 6). In two patients the vision remained stable without improvement or deterioration for 10 years and 8 years.

Seven patients continued to experience deterioration of vision: four are alive, two subsequently have died, and one was in a terminal state at the time of the last inquiry.

Summary

The problem of tumors of the optic nerve and chiasm has been reviewed. In a series of 20 cases of glioma of the optic nerve approximately 25% of the patients were successfully treated surgically, and showed additional improvement in vision after irradiation therapy.

References


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