Astrocytomas of the spinal cord in children and adolescents

RONALD REIMER, M.D., AND BURTON M. ONOFRIO, M.D.

St. Mary's Hospital, Rochester, Minnesota

The authors review 32 cases of spinal cord astrocytoma in patients under 20 years of age who were treated at the Mayo Clinic between 1955 and 1980. There was a 1.3:1 male to female ratio. Twenty patients were between 6 and 15 years of age at the time of diagnosis. The duration of symptoms prior to definitive diagnosis varied from 5 days to 9 years, with an average of 24 months. The most common symptoms were pain (62.5%), gait disturbance (43.7%), numbness (18.8%), and sphincteric dysfunction (18.8%). The most common neurological findings were a Babinski response (50.0%), posterior column sensory dysfunction (40.6%), and paraparesis (37.5%). A median follow-up period of 8.6 years (range 0.8 to 25.5 years) revealed that the survival time diminished with increased histological grade of the astrocytoma (p < 0.001).

The development of postlaminectomy spinal deformities represented a serious postoperative complication. This occurred in 13 patients and was first recognized between 8 and 90 months postoperatively. Six deformities occurred following cervical laminectomy, and eight patients required at least one orthopedic procedure. It is crucial to follow these patients for an extended period of time to watch for postoperative spinal deformities.

KEY WORDS • astrocytoma • spinal cord tumor • laminectomy • postlaminectomy deformity • children

The first intradural extramedullary spinal cord tumor was removed successfully by Victor Horsley after the diagnosis was established in 1887 by Gowers. The first successful removal of an intramedullary cord tumor, a neurofibrosarcoma, was carried out by von Eiselsberg in 1907 with useful neurological recovery, and this result was duplicated by Elsberg in his very first intramedullary tumor operation performed in 1910. After nicking the cord while opening the dura that overlay the tumor, Elsberg noted some tumor extruding from the cord. He closed the wound, then undertook reexploration 1 week later to find the lesion more completely extruded and more easily removed. Subsequently, he advocated this two-stage procedure.

Greenwood introduced the bipolar coagulating forceps in 1940 and later pioneered a microsurgical approach to the removal of intramedullary tumors. In 1976, Yaşargil, et al., stated that, with the aid of the operating microscope, preservation of small spinal cord vessels around the tumor was possible. In 1982, Epstein and Epstein reported the use of the Cavitron ultrasonic surgical aspirator permitting nontraumatic aspiration of all visible tumor without movement of adjacent normal spinal cord.

Astrocytomas in Children

Incidence and Categorization

Neoplasms of the central nervous system are less common among children than among adults. The accepted ratio of intracranial to intraspinal neoplasms in adults is 5:1. In the pediatric age group, the ratio varies from 20:1 to 5:1. More specifically, Kopelson, et al., stated that the ratio is 10:1 for astrocytomas and from 20:1 to 3:1 for ependymomas. Okazaki emphasized that embryonal tumors have a far greater incidence in childhood, that sarcomas are more frequent, and that schwannomas and meningiomas are unusual in childhood (Table 1). However, the true incidence of these tumors in children is unknown because there is a lack of uniformity in study technique.

Astrocytomas and ependymomas comprise the largest group of primary intramedullary lesions. Unlike the adult population, in which ependymomas account for 50% of intramedullary tumors, a compilation of 13 series reveals that approximately 59% of pediatric intramedullary tumors are astrocytomas, and only about 28% are pure intramedullary ependymomas. Intramedullary spinal nevirinomas, neuro-
fibromas, metastases, lipomas, hemangioblastomas, or arteriovenous malformations comprise the other common tumor types.

**Distribution of Astrocytomas**

Among intramedullary spinal cord tumors, astrocytomas are more frequently located in the cervical and thoracic regions while ependymomas have a higher incidence in the conus medullaris and filum terminale. There is a definite predisposition for pediatric tumors to be rostrally located as distinct from the adult type. Forty-six percent of pediatric intramedullary tumors occur in the cervical and cervicothoracic regions compared with only 28% of adult intramedullary tumors. These tumors usually extend over several segments of the cord (average six segments), and on occasion they may be holocord. Cysts have been reported at the rostral and caudal end of the lesion in 40% of astrocytomas, further extending the total intramedullary length. Identification of the solid component by magnetic resonance imaging, contrast metrizamide myelography with computerized tomographic (CT) scanning, and/or angiography is mandatory in limiting the extent of the laminectomy and minimizing the risk of spinal instability.

**Symptomatology**

Symptoms usually develop in a slow subtle fashion, and are commonly present for several months or even years prior to accurate diagnosis. In a series of 115 patients reported by Matson and Tachdjian, the initial diagnosis was erroneous in 70% of cases. Children with astrocytomas are distinguished, as a group, by a long history of complaints before diagnosis is established. The most common symptoms include back pain, lower extremity weakness, and sphincteric dysfunction. Pain is the initial symptom in more than half of children with intramedullary spinal cord tumors, and is characterized as either root, spinal, or tract pain. Spinal pain is most frequently dull and aching in quality, and is localized to dermatomes adjacent to the tumor. Night pain is often a prominent complaint. Root pain, which is less frequent, is usually projected over a single dermatome, and it is unclear whether this is caused by swelling of the spinal cord with distention of the root or if the root entry zone is infiltrated with tumor. The location of the symptoms may bear no anatomic relationship to the spinal segments that are infiltrated by tumor, and pain may suggest other more common clinical entities.

Upper motor neuron findings may be less well defined than lower motor neuron findings, the latter group offering a better indication of the level of the lesion.
Spinal cord astrocytomas in children

### TABLE 1
Comparative incidence of spinal tumors in patients of all ages and in children

<table>
<thead>
<tr>
<th>Primary Spinal Tumors</th>
<th>Percent of Cases</th>
</tr>
</thead>
<tbody>
<tr>
<td>glioma</td>
<td>20-25</td>
</tr>
<tr>
<td>astrocytoma (30%), oligodendroglioma (3%), ependymoma (65%), other (2%)</td>
<td></td>
</tr>
<tr>
<td>schwannoma</td>
<td>30</td>
</tr>
<tr>
<td>meningioma</td>
<td>25</td>
</tr>
<tr>
<td>other</td>
<td>20-25</td>
</tr>
<tr>
<td>total</td>
<td>100</td>
</tr>
<tr>
<td>children less than 15 years old</td>
<td></td>
</tr>
<tr>
<td>glioma (range 15% to 30%)</td>
<td>20</td>
</tr>
<tr>
<td>schwannoma</td>
<td>10</td>
</tr>
<tr>
<td>meningioma</td>
<td>3-5</td>
</tr>
<tr>
<td>congenital tumors</td>
<td>20-40</td>
</tr>
<tr>
<td>sarcoma (including extension from spinal metastasis)</td>
<td>15-25</td>
</tr>
<tr>
<td>total</td>
<td>100</td>
</tr>
</tbody>
</table>

* Primary spinal tumors are one-tenth as frequent as intracranial tumors in all ages and one-fifth to one-tenth as frequent as intracranial tumors in children. Data obtained from Okazaki.†

† About 70% of tumors are extramedullary and 30% are intramedullary.

In cervical lesions, motor examination may reveal only mild upper extremity paresis without associated paraparesis. The presence of kyphosis and/or scoliosis is of little help in diagnosis, although these conditions can be the result of spinal cord dysfunction.

Increased intracranial pressure, with papilledema and/or hydrocephalus, may complicate the clinical manifestations of intramedullary tumors in as many as 12.5% of patients. This may be caused by intracranial seeding of spinal intramedullary glioblastoma, or it may be secondary to elevation of cerebrospinal fluid (CSF) protein levels. In the latter condition, the papilledema rapidly resolves with surgical removal of the spinal lesion.

Radiographic Findings

Computerized tomographic scanning of the spine with intravenous administration of contrast material as well as intrathecal metrizamide instillation may distinguish not only cord enlargement but also differentiation between cystic and solid components. Myelography may also be performed via a percutaneous spinal cord puncture. A contrast-enhanced CT scan of the head with cuts made through the foramen magnum and C-1 should be done prior to myelography to determine if there is a tumor involving the brain stem or if there are congenital variations such as an Arnold-Chiari malformation, with or without aqueductal stenosis. When contrast material is introduced via the lumbar approach, a complete block to the cephalad flow of dye may require instillation of contrast medium at C1-2 to adequately evaluate the cephalad extent of the tumor.

Newer diagnostic modalities include ultrasonography and magnetic resonance imaging. Ultrasonic visualization of spinal cord tumors intraoperatively after exposure of the dura helps to delineate the precise extent of an intramedullary lesion and any associated cystic component. Magnetic resonance imaging delineates the cord directly rather than indirectly as in myelography, eliminates bone artifact of CT, and possesses multidplanar capabilities.

Summary of Cases

Preoperative Findings

Thirty-two patients under 20 years of age with spinal cord astrocytomas were studied at the Mayo Clinic between 1955 and 1980. During this period, an 8.2:1 ratio of brain to spinal cord astrocytomas was recorded. Twenty patients (62.5%) were between 6 and 15 years of age at the time of diagnosis. There was a 1.3:1 male to female ratio. The duration of initial symptoms prior to definitive diagnosis varied from 5 days to 9 years, with an average of 24 months (Fig. 1). The most common symptoms were pain (62.5%), gait disturbance (43.7%), numbness (18.8%), and sphincter dysfunction (18.8%) (Fig. 2). The most common physical findings included a Babinski response in 16 patients (50%), posterior column sensory dysfunction in 13 (40.6%), and paraparesis in 12 (37.5%) (Fig. 3).

Spine x-ray films were abnormal in 19 patients (59.4%). The most common findings were increased interpedicular distance at the level of the tumor in nine patients (28.1%) and kyphosis or kyphoscoliosis in nine (28.1%). Myelography, performed in 27 patients, was abnormal in all. Fifteen patients (55.6%) displayed a complete obstruction with an average CSF total protein level of 1964 mg%, and 12 (44.4%) displayed a subtotal obstruction or free flow of dye with an average CSF total protein level of 250 mg%. The segments most commonly involved were the cervical cord with extension into the medulla in seven patients (21.9%) and the thoracic cord only in seven (21.9%) (Fig. 4).

J. Neurosurg. / Volume 63 / November, 1985
Operative Results

Decompression and biopsy alone was performed in 22 cases (68.7%), subtotal or partial removal in eight (25%), and radical removal in two (6.3%). Two patients (6.3%) died within the first 5 days postoperatively, and three (9.4%) were lost to follow-up evaluation. Of the remaining patients, 96.3% underwent postoperative radiation therapy with an average total dosage of 4100 rads.

The mean survival period after radical removal was 173.5 months, compared to 66.6 months for decompression and biopsy. For patients undergoing decompression and biopsy only, the 60-month and 84-month survival rates were 53.3% and 42.7%, respectively. This is in contrast to patients who underwent a subtotal removal, whose 84-month survival rate was 100% (p = 0.02). Only two patients had radical resection; they were alive at 3.9 years and 25.5 years postoperatively, precluding effective statistical analysis utilizing Kaplan-Meier actuarial estimates. However, we suggest that a more radical resection may be associated with increased survival times. Pulmonary complications represented the most common cause of death among the patients in this series. Our median follow-up period (for those patients currently alive) was 8.6 years, with a range of 0.8 to 25.5 years.

Upon evaluation of our long-term data, among 27 patients with low-grade astrocytomas, including 17 grade 1 and 10 grade 2 lesions, we found average overall 1-year, 5-year, 7-year, and 10-year survival rates of 88.8%, 80.5%, 71.5%, and 55.3%, respectively. The remaining five patients had grade 3 lesions and all died within the 1st postoperative year. The length of survival diminished with increased histological grade of astrocytoma (p < 0.001).

A total of 13 patients developed a postlaminectomy spinal deformity. Six of these patients had undergone a cervical laminectomy at an average age of 10 years. The spinal deformities included kyphosis or kyphoscoliosis in five patients (15.6%), scoliosis in three (9.4%), subluxation in two (6.3%), spondylolisthesis in one (3.1%), a swan-neck deformity in one (3.1%), and an anterior angulation in one (3.1%). The deformities were first noted roentgenographically between 8 and 90 months postoperatively, with an average of 33 months. Among this group of patients, orthopedic intervention was necessary in eight patients (62%) between 8 months and 12.3 years postlaminectomy. These procedures included Harrington rod placement in four patients, an anterior interbody fusion in four patients, a lateral mass fusion in two patients, and a body cast in one patient (Table 2).

Discussion

The benefits of radiation therapy after surgery for spinal cord astrocytomas are controversial. The biological behavior of intraspinal low-grade astrocytomas with or without radiation therapy is unpredictable. There are reported cases of long-term survivors with myelotomy and biopsy as well as biopsy and radiation therapy. Some authors believe that a radical excision of the tumor makes postoperative radiation unnecessary, while it may be more reasonable following a less complete removal or a simple biopsy.\textsuperscript{28,46,77,78,90}

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Tumor Level</th>
<th>Level of Tumor</th>
<th>Nature of Postlaminectomy Deformity</th>
<th>Time to Development of Deformity*</th>
<th>Orthopedic Stabilization Necessary</th>
<th>Time to Stabilization Procedure*</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>2 thoracolumbar</td>
<td>T9-12 suboccipital craniectomy-C-2</td>
<td>severe kyphoscoliosis</td>
<td>28</td>
<td>T8-12 anterior interbody fusion</td>
<td>132</td>
</tr>
<tr>
<td>2</td>
<td>1 T9-12</td>
<td>suboccipital craniectomy-C-2</td>
<td>severe kyphoscoliosis</td>
<td>15</td>
<td>T9-12 anterior interbody fusion</td>
<td>135</td>
</tr>
<tr>
<td>3</td>
<td>2 T10-12</td>
<td>C1-3 suboccipital craniectomy-C-2</td>
<td>progressive kyphoscoliosis</td>
<td>54</td>
<td>C2-3 &amp; C3-4 subluxation</td>
<td>54</td>
</tr>
<tr>
<td>4</td>
<td>1 T6-10</td>
<td>C1-3 suboccipital craniectomy-C-2</td>
<td>progressive kyphoscoliosis</td>
<td>12</td>
<td>C2-3 &amp; C3-4 subluxation</td>
<td>102</td>
</tr>
<tr>
<td>5</td>
<td>1 T8-11</td>
<td>T10-12 suboccipital craniectomy-C-2</td>
<td>severe kyphoscoliosis</td>
<td>8</td>
<td>C2-3 &amp; C3-4 subluxation</td>
<td>8</td>
</tr>
<tr>
<td>6</td>
<td>2 thoracolumbar</td>
<td>T6-10 thoracolumbar</td>
<td>progressive kyphoscoliosis</td>
<td>41</td>
<td>C2-3 &amp; C3-4 subluxation</td>
<td>8</td>
</tr>
<tr>
<td>7</td>
<td>1 T9-12</td>
<td>T6-10 thoracolumbar</td>
<td>progressive kyphoscoliosis</td>
<td>65</td>
<td>C2-3 &amp; C3-4 subluxation</td>
<td>41</td>
</tr>
<tr>
<td>8</td>
<td>1 T10-12</td>
<td>T6-10 thoracolumbar</td>
<td>progressive kyphoscoliosis</td>
<td>91</td>
<td>C2-3 &amp; C3-4 subluxation</td>
<td>65</td>
</tr>
<tr>
<td>9</td>
<td>1 T10-12</td>
<td>T6-10 thoracolumbar</td>
<td>severe kyphoscoliosis</td>
<td>148</td>
<td>C2-3 &amp; C3-4 subluxation</td>
<td>91</td>
</tr>
<tr>
<td>10</td>
<td>2 suboccipital craniectomy-C-2</td>
<td>T10-12 suboccipital craniectomy-C-2</td>
<td>severe kyphoscoliosis</td>
<td>73</td>
<td>C2-3 &amp; C3-4 subluxation</td>
<td>90</td>
</tr>
<tr>
<td>11</td>
<td>1 suboccipital craniectomy-C-1</td>
<td>T10-12 suboccipital craniectomy-C-2</td>
<td>severe kyphoscoliosis</td>
<td>49</td>
<td>C2-3 &amp; C3-4 subluxation</td>
<td>148</td>
</tr>
<tr>
<td>12</td>
<td>1 L3-sacrum</td>
<td>T10-12 suboccipital craniectomy-C-2</td>
<td>grade 1 spondylolisthesis</td>
<td>17</td>
<td>C2-3 &amp; C3-4 subluxation</td>
<td>73</td>
</tr>
<tr>
<td>13</td>
<td>1 C3-T2</td>
<td>T10-12 suboccipital craniectomy-C-2</td>
<td>severe swan-neck deformity</td>
<td>9</td>
<td>C2-3 &amp; C3-4 subluxation</td>
<td>73</td>
</tr>
</tbody>
</table>

* Interval (mos) after initial surgery.

Table 2

Clinical course in 13 patients with postlaminectomy spinal deformities
Spinal cord astrocytomas in children

Data on radiation therapy for spinal cord astrocytomas are difficult to interpret and are often based on inference from brain lesions of a similar histological type.64 Such data suggest that radiation therapy may increase the recurrence-free survival time in patients with incompletely resected ependymomas.17,34,67,93 In all cases of non-resectable tumors, regardless of histological type, Hendrick42 recommends postoperative radiation therapy.

Risk of radiation therapy to the spinal cord is not well quantitated.74,75 It is accepted, however, that radiation sensitivity increases with the length of cord irradiated, the size of the individual daily dose, and the total dose given, and that it may be diminished with alternate fractionation regimens.5,50,69 The most commonly used dose is 5000 rads in 25 treatments. Kramer, et al.,53-55 recommended that this dose be decreased by 10% for thoracic lesions due to an increased radiosensitivity at this level. They also believed that the functional tolerance of the spinal cord is 10% to 15% lower in children than in adults. A tumor-containing cord may be more sensitive to injury than is a normal cord.62 and one must always be cautious of rapid fractionation regimens.13,23,42,102-104

It is most crucial to observe these patients for the development of a postlaminectomy deformity of spinal curvature, especially in children.11,14,33,57,58,85,98,99,106,107 Experimental spinal deformities have been created with surgery, radiation, and metabolic techniques, and by causing injury to neural, bony, ligamentous, and muscular structures of the growing spine of laboratory animals. The combination of neurological deficit, loss of support by the posterior elements, and radiation-induced arrest of growth of the vertebral bodies represents a triple threat to spinal stability.10,16,31,44,45,63,81,82,93

Conclusions

It is apparent that spinal cord astrocytomas represent a difficult clinical problem and their optimal treatment remains controversial. These patients usually have symptoms for a long period prior to accurate diagnosis. The resolution and accuracy of neuroradiological procedures continue to improve with the advent of magnetic resonance imaging, and we may look forward to establishing an earlier diagnosis in these patients. Our experience suggests that one should consider a radical removal of spinal cord astrocytoma when feasible. It remains difficult to evaluate the use of postoperative radiotherapy in astrocytomas, but we believe it should continue to be utilized as a postoperative adjunct. We cannot overstress the importance of closely observing these patients for the development of a postlaminectomy spinal deformity which may occur from weeks to years postoperatively. Raimondi, et al.,81 stated that osteoplastoc laminotomy prevented spine instability in their patients undergoing multisegmental exposure for spinal cord tumor. This technique, in conjunction with the use of postoperative external immobilization, should be considered in an effort to prevent postoperative instability. Lastly, it appears that long-term survival is related to tumor grade.51

Acknowledgments

We thank Julie Wollschlager and Bernita Bruns for their excellent secretarial assistance in the preparation of this manuscript.

References

Assist Tomogr 6:655–670, 1982


674

J. Neurosurg. / Volume 63 / November, 1985
Spinal cord astrocytomas in children

1976, pp 201–220


Manuscript received October 29, 1984. Accepted in final form February 25, 1985. Address reprint requests to: Ronald Reimer, M.D., Department of Neurologic Surgery, Room 5-224, East Joseph, St. Mary’s Hospital, Rochester, Minnesota 55901.