Intra-axial tumors of the cervicomedullary junction

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The authors present their experience with the operative management of 20 intra-axial tumors of the cervicomedullary junction. There were two distinct modes of clinical presentation: lower cranial nerve dysfunction and spinal cord dysfunction. Both groups of patients had indolent courses; in 75% of the patients the symptoms had been present for 6 months to 2 years. Radical excision was carried out in all patients. There was no surgical mortality. Postoperative neurological recovery was directly related to the preoperative status; one patient had a significant new deficit. The authors conclude that intrinsic gliomas of the cervicomedullary junction are amenable to radical excision and that an aggressive surgical approach offers the potential for both neurological recovery and long-term survival. The neuroradiological evaluation and operative technique are discussed.

KEY WORDS • glioma • cervicomedullary junction • brain-stem neoplasm • spinal cord neoplasm • diagnostic imaging • cranial nerve

Radical surgical excision of intra-axial tumors in the region of the cervicomedullary junction has not been employed with any regularity because it is generally assumed that extensive dissection in this vital region will result in significant morbidity and mortality. For this reason, radiation therapy (with or without biopsy) has been considered the treatment of choice and, while this may be compatible with transient remission of symptoms, progressive disability and death have been considered inevitable.

In previous publications, the authors have described 185 operations for intrinsic tumors of the spinal cord and brain stem. Among these cases were eight neoplasms that were located at the cervicomedullary junction. These tumors extended from the medulla to the cervical spinal cord, and the preoperative clinical states were improved or stabilized following radical surgical excision. Five of these neoplasms were low-grade astrocytomas and three were gangliogliomas. This small experience suggested that cervicomedullary neoplasms were often “benign” and amenable to surgery. The present report summarizes the first author’s further experience with radical surgical excision of 20 intra-axial tumors occurring in the region of the cervicomedullary junction.

Summary of Cases

This series included 15 children and five adults. Primary clinical manifestations were roughly divisible into two categories: lower cranial nerve dysfunction and spinal cord dysfunction. Both groups of patients commonly had indolent courses, and in 15 patients symptoms were present from 6 months to 2 years prior to diagnosis (Table 1).

Cranial Nerve Dysfunction

Eight patients noted lower cranial nerve dysfunction manifested by difficulty in swallowing, nasal speech, and (much later) hemiparesis or quadriplegia. Three of these patients described paresthesias and dysesthesias occurring in the arms and legs; in two of these patients a tentative diagnosis of multiple sclerosis was made on the basis of exacerbations and remissions of the subjective complaints. Three patients complained of chronic nausea associated with recurring vertigo.

Spinal Cord Dysfunction

Twelve patients had the insidious onset of motor dysfunction, and obvious lower cranial nerve dysfunction followed usually months but occasionally years later. In two of these patients severe pain in the neck antedated objective neurological dysfunction by 12 to 36 months, and in six of the remaining patients severe spinal pain was described just before or after the onset of symptoms. In two very young patients with large tumors of the cervicomedullary junction, neck pain was the only complaint and no neurological dysfunction was disclosed by either the medical history or examination.
### TABLE 1

**Summary of clinical course in 20 patients**

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age (yrs), Sex</th>
<th>Duration of Symptoms (mos)</th>
<th>Preop Neurological Status</th>
<th>Diagnosis</th>
<th>Irradiation</th>
<th>Follow-Up Period (mos)</th>
<th>Neurological Status</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>10, M</td>
<td>36</td>
<td>ataxia, hemiparesis, nasal speech, dysphagia</td>
<td>II</td>
<td>+</td>
<td>48: alive, asymptomatic</td>
<td></td>
</tr>
<tr>
<td>2</td>
<td>11, M</td>
<td>12</td>
<td>ataxia, hemiplegia, dysarthria, dysphagia</td>
<td>III-IV</td>
<td>+</td>
<td>9: dead</td>
<td></td>
</tr>
<tr>
<td>3</td>
<td>17, M</td>
<td>14</td>
<td>quadripareis, ataxia</td>
<td>II</td>
<td>+</td>
<td>24: alive, improved</td>
<td></td>
</tr>
<tr>
<td>4</td>
<td>2, M</td>
<td>4</td>
<td>neck pain</td>
<td>I</td>
<td>0</td>
<td>24: alive, asymptomatic</td>
<td></td>
</tr>
<tr>
<td>5</td>
<td>11, F</td>
<td>14</td>
<td>nausea, vertigo, nasal speech</td>
<td>II</td>
<td>0</td>
<td>24: alive, asymptomatic</td>
<td></td>
</tr>
<tr>
<td>6</td>
<td>6, F</td>
<td>12</td>
<td>vertigo, nausea</td>
<td>I</td>
<td>+</td>
<td>24: alive, asymptomatic</td>
<td></td>
</tr>
<tr>
<td>7</td>
<td>15, M</td>
<td>72</td>
<td>vertigo, ataxia, dysphagia</td>
<td>II</td>
<td>+</td>
<td>24: alive, moderate</td>
<td></td>
</tr>
<tr>
<td>8</td>
<td>31, M</td>
<td>24</td>
<td>quadripareis, ataxia, dysarthria, dysphagia</td>
<td>II</td>
<td>+</td>
<td>24: alive, improved</td>
<td></td>
</tr>
<tr>
<td>9</td>
<td>5, F</td>
<td>3</td>
<td>facial diplegia, quadripareis, dysphagia</td>
<td>GG</td>
<td>+</td>
<td>3: dead</td>
<td></td>
</tr>
<tr>
<td>10</td>
<td>7, M</td>
<td>18</td>
<td>hemiparesis, facial pain, dysarthria</td>
<td>GG</td>
<td>+</td>
<td>60: alive, asymptomatic</td>
<td></td>
</tr>
<tr>
<td>11</td>
<td>6, F</td>
<td>6</td>
<td>monoparesis rt/UE</td>
<td>III</td>
<td>0</td>
<td>24: alive†</td>
<td></td>
</tr>
<tr>
<td>12</td>
<td>2, F</td>
<td>3</td>
<td>neck pain, ataxia</td>
<td>II</td>
<td>0</td>
<td>24: alive, asymptomatic</td>
<td></td>
</tr>
<tr>
<td>13</td>
<td>8, M</td>
<td>12</td>
<td>hemiatrophy of tongue, hemiparesis</td>
<td>GG</td>
<td>+</td>
<td>36: dead</td>
<td></td>
</tr>
<tr>
<td>14</td>
<td>16, F</td>
<td>6</td>
<td>quadripareis, ataxia</td>
<td>III</td>
<td>+</td>
<td>6: dead</td>
<td></td>
</tr>
<tr>
<td>15</td>
<td>31, M</td>
<td>6</td>
<td>neck pain, UE weakness</td>
<td>I</td>
<td>0</td>
<td>6: alive, improved</td>
<td></td>
</tr>
<tr>
<td>16</td>
<td>62, M</td>
<td>12</td>
<td>neck pain, mild quadripareis</td>
<td>II</td>
<td>+</td>
<td>24: alive, improved</td>
<td></td>
</tr>
<tr>
<td>17</td>
<td>12, F</td>
<td>9</td>
<td>advanced quadripareis, lower cranial nerve dysfunction</td>
<td>III</td>
<td>+</td>
<td>9: alive, quadriplegic since surgery</td>
<td></td>
</tr>
<tr>
<td>18</td>
<td>12, M</td>
<td>36</td>
<td>neck pain, torticollis</td>
<td>I</td>
<td>0</td>
<td>6: alive, asymptomatic</td>
<td></td>
</tr>
<tr>
<td>19</td>
<td>24, F</td>
<td>18</td>
<td>neck pain, spastic quadripareis, lower cranial nerve dysfunction</td>
<td>E</td>
<td>0</td>
<td>6: alive, improved</td>
<td></td>
</tr>
<tr>
<td>20</td>
<td>36, F</td>
<td>36</td>
<td>quadripareis</td>
<td>E</td>
<td>+</td>
<td>36: alive, postop sleep apnea for 3 mos improved</td>
<td></td>
</tr>
</tbody>
</table>

* Abbreviations: I to IV = grades of astrocytoma; GG = ganglioglioma; UE = upper extremity; E = ependymoma.
† The tumor recurred after 6 months, requiring repeat radical excision and irradiation.

### Neurodiagnostic Investigation

Magnetic resonance imaging (MRI) was indispensable to the preoperative investigation of tumors in this location. Whereas computerized tomography was often unreliable in the region of the foramen magnum, MRI provided excellent visualization of the medulla and cervicomedullary junction and very precise anatomical delineation of the tumor's rostral-caudal extension.

In eight patients the medullary component of the lesion was a non-neoplastic cyst that extended from the rostral pole of a cervical astrocytoma. In 11 patients the medullary component of the neoplasm was noncystic or contained a small intratumor cyst (Fig. 1). Three tumors extended throughout the cervical cord, but in no case did a tumor extend rostral to the pontomedullary junction. Two patients harbored intratumor cysts.
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FIG. 2. Left: Preoperative magnetic resonance image showing a solid tumor (solid arrow) and cyst (open arrow). Center: Intraoperative ultrasound scan, longitudinal view, prior to dural incision demonstrating a tumor (solid arrow) and a cyst (open arrow). Right: Ultrasound image, transverse view, showing a solid tumor (arrows).

that had apparently developed after primary radiation therapy.

Surgical Technique

Monitoring of brain-stem evoked potentials was a valuable surgical adjunct: the electrical activity became relatively disordered as the interface between tumor and normal neural tissue was approached. It is essential to use a monitoring system that updates information every few seconds, as it is of very little practical value to know that the potentials were disrupted during dissection at some time in the previous 30 to 120 seconds when it is too late for corrective measures (temporarily halting the tumor excision).

All operations were carried out with the patient in the prone position. The approach was through a cervical laminectomy with or without a small suboccipital craniectomy. In most cases it was not necessary to remove the occipital bone if the medullary component of the neoplasm was cystic; if a cystic lesion did not extend above the foramen magnum, it could be drained as the rostral portion of the neoplasm was removed (Fig. 2 left).

Before the dura was opened, intraoperative transdural ultrasonography was used and was found indispensable in defining the rostral-caudal limits of the neoplasm and the presence or absence of associated cysts' (Fig. 2 center and right). After the dura was opened, the myelotomy was carried out over the entire length of the solid component of the neoplasm (Fig. 3). Tumor excision was initiated in the region where the neoplasm was most voluminous, as documented by both the ultrasound study and visual inspection. The Cavitron ultrasonic surgical aspirator and the surgical laser were utilized to gradually debulk the neoplasm. It was usually possible to obtain a gross total excision of neoplasms and to identify a glia-tumor interface circumferentially around the tumor. Tumors in the medulla were also exposed through a myelotomy in the area where the neoplasm was closest to the surface of the area postrema or the floor of the fourth ventricle. Tumors in the brain stem and in the spinal cord were removed from inside out until white matter was ob-

Fig. 3. Operative exposure before (left) and after (right) tumor excision.
served or brain-stem evoked potentials became disor-
dered as compared to the baseline. The latter event
suggested that the tumor-brain stem interface was
nearby, and the dissection was suspended.

Ultrasonography was utilized to monitor the tumor
excision as surgery proceeded. Without this imaging
technique, intratumor or rostral cysts in the medulla
would have been missed and the surgery suspended
prematurely in three cases. In addition, ultrasonography
was very helpful in identifying the anterior surface of
the cervical spinal cord vis-à-vis the residual cavity
from which the tumor had been excised. This either
confirmed the surgical impression that the tumor had
been removed or encouraged the surgeon to proceed
further (Fig. 4 left).

Operative Results

There was no surgical mortality in this group of
patients. One patient had a sleep apnea syndrome for 6
weeks and required support from a ventilator during
that period of time. She gradually recovered and had
regained nearly normal respiratory function 3 years
after surgery. Three patients had impaired position
sense in the upper extremities postoperatively, and six
had transient weakness and spasticity in the lower ex-
tremities. One 10-year-old girl who was moderately
disabled preoperatively became quadriplegic postop-
eratively.

The postoperative neurological recovery was directly
related to the preoperative neurological status. Patients
with a mild disability made a normal or nearly normal
recovery, while patients who were severely disabled
preoperatively were much more likely to sustain a neu-
rological injury as a result of the surgical procedure.
There was no correlation between the presence of cysts
and the degree of recovery. On postoperative MRI the
cervicomedullary junction had returned to normal or
nearly normal in eight patients (Fig. 4 right) and re-
mained expanded in 11 others.

Pathology

There were five grade I astrocytomas, six grade II
astrocytomas, three gangliogliomas, two ependymomas,
and four anaplastic tumors. All patients with malignant
astrocytomas died from tumor progression within 6 to
9 months after surgery. One patient with a ganglio-
glioma died from tumor progression 3 months postop-
eratively. All of the remaining patients in this series are
alive 3 months to 6 years postoperatively.

Discussion

Epstein and McCleary have described the results of
radical surgical excision of 35 intrinsic non-exophytic
brain-stem gliomas. While the surgery was relatively
well tolerated, it was apparent retrospectively that this
therapy did not have a favorable impact on malignant
tumors. Although transiently improved, those patients
all succumbed to tumor progression within 6 to 9
months of surgery. Radical removal of benign neo-
plasms resulted in significant neurological improve-
ment and long-term clinical remission. While only two
of 22 neoplasms rostral to the medulla were benign
gliomas, eight consecutive cervicomedullary neoplasms
were grade I or II astrocytomas. On the basis of these
observations, we decided to analyze our surgical and
clinical experience with cervicomedullary neoplasms to
determine if they were a separate subgroup of brain-
stem spinal cord neoplasms that may be amenable to
radical surgical excision.

Our results in this series of 20 patients suggest that
intrinsic tumors arising at the cervicomedullary junc-
tion represent a melting-pot of pathology, being an
admixture of neoplasms that are commonly seen in the
spinal cord as well as those that occur in the brain stem.
Whereas most tumors rostral to the medulla are malig-
nant, it seems that tumors in the lower brain stem are
more similar to neoplasms that occur in the spinal
cord where at least 90% of tumors are low-grade gli-
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omas. 2-4,6 Perhaps in some circumstances the medul-

lary component of the neoplasm represents the rostral

extension of what was primarily a cervical spinal cord
tumor.

In this regard, it is of interest that there seemed to be

some correlation between the primary clinical symp-
toms and the primary location of the neoplasm. Pa-

tients in whom lower cranial nerve dysfunction was an

early complaint almost invariably had a noncystic med-

ullary component of the neoplasm. In other patients

with early spinal cord symptoms and/or neck pain the

solid part of the neoplasm was in the high cervical cord

and was associated with an apparently non-neoplastic
cyst which “capped” the neoplasm and extended into

the medulla. These latter tumors were identical to the

cystic astrocytomas of the spinal cord which are com-

monly associated with huge non-neoplastic rostral and

caudal cysts.1-5,7,8

Although it is clear that there is only an occasional

role for radical surgical excision of brain-stem tumors

rostral to the medulla, our experience with this series of

20 patients strongly suggests that tumors at the cervi-

comedullary junction are a subgroup of neoplasms

for which surgery may be considered a viable therapeu-
tic option. In other words, while surgery has no positive

role in altering the biology of the most common ma-

lignant brain-stem neoplasms, it has potentially great

therapeutic value for low-grade astrocytomas at the

cervicomedullary junction. On the basis of this small

number of patients and the relatively short follow-up

period, it would be premature to predict the duration of

surgical remission or likelihood of permanent cure, but

it is clear that neurological function as well as survival

have been significantly extended by radical surgical ex-

cision of these neoplasms.

There are two major apparent predictors of potential

success or failure of surgery. These are the duration of

symptoms prior to definitive diagnosis and the micro-

scopic pathology of the neoplasm. Patients who have

had symptoms for years prior to surgery almost invari-

ably had indolent tumors and were in the most

favorable group in terms of significant neurological

improvement and long-term survival following surgery.

Obviously, radical but subtotal excision of a tumor

that is growing very slowly is more likely to be associ-

ated with a long-term remission than is radical removal

of a rapidly growing tumor. Interestingly, one patient

with an apparently typical ganglioglioma died from tu-

mor progression a few months following surgery. Al-

though the microscopic appearance of this neoplasm

would have suggested a long-term remission, the child

had a very short preoperative neurological course (3

months) and was severely disabled at the time of sur-

gery. In this case, the growth potential of the tumor was

more reflected by the rapid evolution of signs and symp-
toms than by the microscopic appearance.

The pathological grade of the neoplasm was likewise
directly related to the postoperative neurological course.

Patients with a grade III or IV neoplasm did not do

well even if there was a very prolonged preoperative

course. In other words, in these circumstances it seems

likely that the tumor had undergone malignant degener-

ation and the microscopic pathology predicted the

unsatisfactory surgical result in terms of recurrence and

longevity.

Our results in this series of patients suggest that it is

technically feasible to radically excise intrinsic tumors

of the cervicomedullary junction with little permanent

morbidity. This is possible because the expanding mass

of a slow-growing glioma probably displaces neural

structures in a circumferential manner but does not
diffusely infiltrate them to the same extent as do malig-
nant gliomas. Therefore, as long as the dissection is

carried out within the bulk of the neoplasm, there is

little risk to adjacent functioning neural tissues.

Conclusions

We have described intrinsic neoplasms at the cervi-

comedullary junction as a “subgroup” of neoplasms

which are similar to both low brain-stem and upper
cervical spinal cord tumors. They are commonly slow-
growing gliomas and, following radical excision, both

neurological recovery and extended longevity may be

anticipated. It is premature to speculate on the duration

of remission or the likelihood of cure, and at this

juncture we may only state that surgery palliates the

neurological course.

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