Surgical approach to C1-C2 nerve sheath tumors

Prasad Krishnan, Sanjay Behari, Deepu Banerji, Naveen Mehrotra, Devendra K. Chhabra, Vijendra K. Jain
Department of Neurosurgery, Sanjay Gandhi Postgraduate Institute of Medical Sciences, Lucknow - 226014, India.

Background: C1 and C2 nerve sheath tumors (NST) are unique in presentation, relationship to neighbouring structures and surgical approaches when compared to their counterparts in other regions of the spine. Aim: The strategies involved in the surgery for C1-C2 NST are discussed

Setting and Design: Retrospective study. Methods: 21 patients with C1 (n=6) and C2 (n=15) NST were operated based on their position with respect to the cord i.e. anterior (4), anterolateral (10), posterolateral (5), and posterior (2). The tumors had extra- and intradural components in 20 patients; while in one, the tumor was purely intradural. The operative approaches included the extreme lateral transcondylar approach (3); laminectomy with partial facetectomy (5); laminectomy (11); and, suboccipital craniectomy and laminectomy (2).

Results: Total excision was performed in 13 patients; while in 7, a partial extraspinal component, and in 1, a small intradural component were left, in situ. Thirteen patients showed improvement by one or more grades in the Harsh myelopathy score; 2 patients with normal power had significant decrease in spasticity; while 5 maintained their grade. One poor-grade patient succumbed to septicemia.

Conclusions: C1-C2 NST may have exuberant growth due to the capacious spinal canal and the absence of a “true” intervertebral foramen at this level. Surgical approaches are determined by its relationship to the cord. A “T incision” on the dura, the partial drilling of the facets, sectioning of the denticulate ligament, rotating the operating table 15 to 30 degrees, and at times sectioning the posterior nerve roots are all useful adjuncts for facilitating access.

Key Words: Spinal nerve sheath tumors, craniovertebral junction, laminectomy, extreme lateral transcondylar approach

Clinical spectrum
In this retrospective study, 21 patients (16 men and 5 women, aged 17-56 years, mean age 34.8 ±12.7 years) operated for C1-C2 NST between 1 January 1994 and 31 December 2002, were studied. The C1-2 NST accounted for 12.1% (21 out of 173) and 33% (21 out of 63) of all spinal and cervical NST, respectively. Their duration of symptoms ranged from 3 to 60 months (mean duration=21.8 ±19.5 months). The symptoms at the onset of illness were spasticity (n=10, 47.6%), tingling and numbness below neck (n=4, 19.0%), weakness (n=3, 14.3%) and neck pain (n=4, 19.0%).

Grading
The clinical disability of the patient was evaluated using the Harsh Myelopathy Grading 10 (Table 2) as: Grade 1: normal power with only hyperreflexia and Babinski’s sign (n=2, 9.5%); Grade 2: able to walk but not able to run (n=5, 23.8%); Grade 3: requiring support to ambulate (n=11, 52.4%) and Grade 4: completely bedridden (n=3, 14.2%).

Radiological findings
The patients were evaluated using lateral radiographs of the
Table 1: Clinical presentations of the 21 patients with C1-C2 nerve sheath tumors

<table>
<thead>
<tr>
<th>Presenting Features</th>
<th>Number of patients</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>Spasticity</td>
<td>21</td>
<td>100</td>
</tr>
<tr>
<td>Weakness</td>
<td>18</td>
<td>85.7</td>
</tr>
<tr>
<td>Spinothalamic tract involvement</td>
<td>11</td>
<td>52.4</td>
</tr>
<tr>
<td>Posterior column involvement</td>
<td>15</td>
<td>71.4</td>
</tr>
<tr>
<td>Horner’s syndrome</td>
<td>5</td>
<td>23.8</td>
</tr>
<tr>
<td>Sphincter dysfunction</td>
<td>6</td>
<td>28.6</td>
</tr>
<tr>
<td>Dyspnea at rest</td>
<td>3</td>
<td>14.3</td>
</tr>
<tr>
<td>Neck pain</td>
<td>10</td>
<td>47.6</td>
</tr>
<tr>
<td>Neck tilt</td>
<td>5</td>
<td>23.8</td>
</tr>
<tr>
<td>Restriction of neck movements</td>
<td>7</td>
<td>33.3</td>
</tr>
<tr>
<td>Suboccipital muscle wasting</td>
<td>4</td>
<td>19</td>
</tr>
<tr>
<td>Facial hypoaesthesia</td>
<td>4</td>
<td>19</td>
</tr>
<tr>
<td>Nystagmus</td>
<td>4</td>
<td>19</td>
</tr>
<tr>
<td>Papilledema</td>
<td>1</td>
<td>4.8</td>
</tr>
<tr>
<td>IXth-Xth nerve palsy</td>
<td>1</td>
<td>4.8</td>
</tr>
<tr>
<td>Neck swelling</td>
<td>1</td>
<td>4.8</td>
</tr>
</tbody>
</table>

Table 2: Pre-and postoperative Harsh Myelopathy Grade [13] of the patients

<table>
<thead>
<tr>
<th>Harsh Grade</th>
<th>Preoperative</th>
<th>Postoperative</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>2</td>
<td>3</td>
</tr>
<tr>
<td>2</td>
<td>5</td>
<td>16</td>
</tr>
<tr>
<td>3</td>
<td>11</td>
<td>1</td>
</tr>
<tr>
<td>4</td>
<td>3*</td>
<td>-</td>
</tr>
</tbody>
</table>

*one patient expired after surgery.

Results

Total excision was achieved in all patients except in 6 of the 10 (60%) anterolaterally placed and 2 of the 5 (40%) posterolaterally placed lesions. In 7 of these patients, a part of the tumor extending far laterally beyond the confines of the spinal canal was not removed; and, in 1 patient with an anterolaterally placed lesion, a small part of the lesion located anterior to the cord that did not have a good plane of cleavage, was left, in situ.

At clinical assessment performed at the time of discharge, 13 of the 21 patients showed improvement by one or more grades while 7 maintained their status. The pre- and postoperative disability grades are summarized in Table 2.

Surgical procedures (Table 3)

Of the 4 patients with anteriorly located tumors, 3 underwent total excision using the extreme lateral transcondylar approach,11,12,13 In 1 patient with a similar anteriorly placed tumor in whom the tumor was extending above the foramen magnum (FM) and laterally along the nerve root, the tumor was accessible using a suboccipital craniectomy and C1-C2 laminectomy.

The commonest location was the anterolateral one (n=10). In 5 of these patients, a laminectomy with partial facetectomy1 provided an oblique access to the tumor, while in 5 others, only a laminectomy sufficed. In all the 10 patients, a T-incision was given on the dura in order to connect the intra- and extradural components. Access to the part of the lesion located anterior to the cord within the intradural space was greatly facilitated by excision of the denticulate ligament.

The posterior and posterolaterally located lesions were approached by a laminectomy alone. In one patient, however, in whom the lesion was extending above the foramen magnum, a suboccipital craniectomy was also performed.

The NST were arising from C1 and C2 roots in 6 (28.6%) and 15 (71.4%) patients, respectively. One or more rootlets were sacrificed in all cases. In 9 cases, the entire nerve root from which the tumor was arising was sacrificed. A patch graft was used to close the dura in 11 cases (in 1 patient who underwent an extreme lateral transcondylar approach; in 5 patients who underwent laminectomy and partial facetectomy; in 4 patients who had a laminectomy with a “T” incision on the dura; and, in 1 patient who underwent a suboccipital craniectomy and laminectomy). It was possible to primarily close the dura without a patch in rest of the cases.

![Figure 1: Classification of tumors with respect to their location relative to the cord, roots and the denticulate ligament.](Image)
Figure 2: (a) T1-weighted sagittal MR image showing a nerve sheath tumor located anterior to the cord opposite the odontoid causing cervicomedullary compression; (b) T2-weighted MR image showing complete tumor removal using an extreme lateral transcondylar approach. Signal intensity changes are still persisting in the cord opposite the original site of lesion; and, (c) Axial CT image showing the resection of part of the C1 facet and total tumor removal.

Figure 3: (a) Preoperative T2-weighted coronal MR image showing a dumb-bell nerve sheath tumor with a large extraspinal component; (b) Intraoperative photograph of the C2 neurinoma exiting through the intervertebral foramina; and, (c) Intraoperative photograph showing total excision of the intra- and extradural component of the tumor.

Figure 4: (a) T1-weighted axial MR image showing an anterolaterally placed tumor extending above foramen magnum pushing the cord posterolaterally; (b) T1-weighted axial MR image showing minimal extraspinal extension.
gery that did not improve despite total excision of the tumor. He succumbed to bronchopneumonia and septicemia. The two patients (where a patch dural graft was placed) who developed CSF leak from the operative incision were managed by acetazolamide administration and lumbar drainage.

**Discussion**

**Incidence**

C1-2 NST represent approximately 5% of spinal and 18% of cervical NST reported while in the present series, they accounted for 12.1% and 33% of the spinal and cervical NST, respectively. Of these, 15 (71.4%) originated from the C2 root and 6 (28.6%) from the C1 root, respectively. This is in agreement with the studies published by Yasuoka, et al,7 with 18 tumors on C2 and 1 on C1 root; by Guidetti et al,15 with 6 on C2 and 3 on C1; and, George et al,2 with 44 on C2 and 6 on C1, respectively. The latter have also reported multiple lesions due to a high incidence of neurofibromatosis in their series.2 In our study, the two patients with neurofibromatosis had solitary lesions of the cervical spine. One of these patients had associated bilateral acoustic schwannomas, while the other had a NST within the lumbar spine.

**Anatomical considerations**

In the present series, 20 patients had dumb-bell lesions with both extra- and intradural extensions, while only one had a purely intradural location. Similarly, George et al have found 83% of C1-2 nerve sheath lesions having an extradural extension. In contrast, the total incidence of extradural extension of NST at other levels within the spine is only 16.2

The relatively large size of these lesions at the C1-2 level is due to the spacious spinal canal at this level. The mean sagittal diameters at the levels of the atlas and axis are 23 and 20mm, respectively, while in the subaxial spine, the average diameter is only 15mm.9 The cervical bulge of the spinal cord also begins below the axis.7 At the occipitocervical and atlantoaxial levels, the facetal pillars lie anterior to the nerve roots exiting through the intervertebral foramina. This also permits an exuberant growth of the lesion outwards since there is no posterior bony obstruction to its growth at the C1-2 levels.9 There is often a delay in the development of clinical symptoms until these tumors attain a fairly large size.4,9

**Surgical approaches (Table 4)**

The MR findings (Figure 1) were utilized to determine the surgical approach. The posteriorly located lesions were accessed using a laminectomy. In the cases of multi-segment involvement, laminoplasty and hemilaminectomy avoid the post-laminectomy kyphosis.7,12 The posterior approach also accessed the anterolateral or posteroventral lesions that, having shifted the cord towards the contralateral side, extended laterally towards the intervertebral foramen along the nerve roots. The intra-tumoral decompression of the lateral part of the tumor provided an oblique trajectory to access the part of the lesion situated in close proximity to the cord. In our series, sectioning of the denticulate ligament,17 rotating the operating table 15 to 30° and at times, sectioning the posterior nerve roots, as suggested by Bueci et al,3 in all anterolateral lesions and the one anteriorly placed lesion treated by the posterior approach, aided in gently mobilizing the tumor from the contact-surface of the cord. Placement of a “T” shaped dural incision, with the horizontal limb of the “T” connecting the intra- and extra-dural components of the tumor also facilitated access.

In the case of dumb-bell tumors, initially an intratumoral decompression was performed. Then, the intradural portion of the lesion was removed maintaining arachnoidal planes in the tumor-cord interface to avoid traction on the cord during the handling of the extradural portion. The use of operative microscope and neurophysiological monitoring facilitate tumor excision with minimal cord manipulation.

In the present series, the anatomical constraints posed by the posterior approach in accessing anterolaterally located lesions were overcome by extending the surgical corridor laterally by adopting a posterolateral trajectory via a laminectomy and partial facetectomy, as proposed by Bartolomei and Crockard.4 This approach is also advantageous in gaining an early control of the vertebral artery.4 Does laminectomy with partial facetectomy lead to cervical instability?16, 18,19 In the two series by George and Lot,2,14 of cervical NST, as well as the study by Bartolomei and Crockard15 of bilateral C2 neurofibromas, none of the patients have required fusion for cervical instability after laminectomy and partial facetectomy. At the level of C1-2, the anterior ligaments confer an additional stability to the cervical spine with less structural contribution from the posterior spinal column.5 The partial preservation of bilateral articular facets between C1 and C2 also confers stability. According to Welling et al,9 C1-C2 lateral joints have a capsule that is lax on the medial side. Most of the movement occurs along a synovial fold present medially. Removal of this part, therefore, does not lead to instability. In our patients with anteromedially placed NST, only a unilateral partial facetectomy with preservation of more than two thirds of the joint surfaces and capsule was performed and the contralateral joint was left completely intact. Thus, no spinal fusion was required.

For anterolaterally placed lesions, Verbeist20 and George et al,2 use an approach that partially removes the anterior portion of the vertebral body and transverse foramen but preserves the facet. The vertebral artery is mobilized from the foramen transversarium and the extraspinal tumor can also be safely excised. However, the extensive drilling and the risk of vertebral artery mobilization for resecting essentially the extraspinal component of the benign lesion (that was unlikely to cause spinal cord compression even in the distant future)
In patients with lesions situated mainly anterior to the cord, the extreme lateral transcondylar approach was used. Though an early control of the vertebral artery, a direct visualization of the cervicomedullary junction and the cord-tumor interface from a lateral aspect, and a field of view above and below the foramen magnum were obtained, the extensive bony and soft tissue dissection were the major deterrents to its frequent use in all dumb-bell, anteriorly placed lesions. The extrapharyngeal approach to the upper cervical spine may be an alternative approach. The transoral approach for midline, anterior, intradural NST has the risk of infection due to traversing through a potentially infected oral cavity; and, is unable to access tumors extending laterally from the midline.

**Implications of the C1/ C2 nerve root resection**

In removing the spinal NST, preservation of the anatomical integrity of the involved nerve root, although desirable, cannot always be achieved. This is specially in cases of large tumors; where both the sensory and motor roots are diffusely involved by the pathological tissue; and, when the tumor extends beyond the proximal pole of the dorsal root ganglion, where the roots are devoid of an arachnoidal sheath that usu-
ally separates them from the tumor tissue. Occasionally, the posterior sensory nerve root may be sacrificed in order to gain access to the anteriorly located tumor.

In our series, one or more rootlets were sacrificed in every case, while in 9 cases, the entire nerve root, from which the tumor arose, was resected. As the C1 and C2 nerve roots are nonappendicular and therefore relatively non-eloquent, this was not associated with any significant neurological deficit except an occasional mild hypoaesthesia in the suboccipital region. While working with eloquent nerves in the subaxial spine, Lot and George minimized postoperative deficits by electrically stimulating the nerve during surgery. When a response was recorded, the functional root was preserved even at the cost of leaving some residual lesion. This procedure was not required in our study since the resection of C1 or C2 root is usually not associated with any significant sensory or motor radicular symptoms.

**Implications of incomplete removal of the extra-spinal tumor**

In 7 patients, a part of the extraspinal tumor extending far laterally beyond the intervertebral foramen was not removed. Usually this part of the tumor has neither clinical recurrence nor a tumor regrowth into the spinal canal even after extremely long intervals ranging from 6 to 18 years. Preserving the nerve roots at the cost of a subtotal tumor removal is also not associated with increased recurrence rates. Seppala et al. have reported that of the 187 patients treated for spinal NST, in the 20 in whom less than total tumor excision was performed to avoid root injury, 18 did not have a symptomatic recurrence even after a median period of 19 years after surgery. In the patients with neurofibromatosis, the recurrence rates in the cases of partial tumor excision are higher. However, only two of our patients had neurofibromatosis and in both of them, a total tumor excision could be achieved.

In conclusion, C1 and C2 nerve sheath tumors are special when compared to their counterparts in other regions of the spine in terms of their tendency toward extradural and extraspinal spread, relationships to neighbouring structures and the surgical approaches required to deal with them. They were classified based on their position with respect to the cord determined by preoperative magnetic resonance imaging. A “T incision” on the dura, the partial drilling of the facets, sectioning of the denticulate ligament, rotating the operating table 15 to 30 degrees, and at times sectioning the posterior nerve roots are all useful adjuncts for facilitating access to these lesions.

**References**


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