Upper cervical intramedullary schwannoma of the spinal cord presenting with myelopathy: illustrative case

Shyam Duvuru, MS, MCh,1 Vivek Sanker, MBBS,2 Naureen Syed, MD,3 Shubham Mishra, MBBS,4 Sayantika Ghosh, MBBS,5 and Tirth Dave6

1Department of Neurosurgery, Apollo Specialty Hospitals, Madurai, Tamil Nadu, India; 2Department of Neurosurgery, Trivandrum Medical College, Kerala, India; 3Department of Clinical Research, UT MD Anderson Cancer Center, Houston, Texas; 4Deanery of Clinical Sciences, College of Medicine and Veterinary Medicine, The University of Edinburgh, Edinburgh, United Kingdom; 5Department of Microbiology and Immunology, Georgetown University Medical Center, Washington, DC; and 6Department of Internal Medicine, Bukovinian State Medical University, Chernivtsi, Ukraine

BACKGROUND Intramedullary schwannomas account for 1.1% of all spinal schwannomas. Preoperative diagnosis is best accomplished by thoroughly evaluating clinical and radiological characteristics, accompanied by a high index of suspicion. The authors report a case of C2–3 intramedullary schwannoma in a young male who presented with neck pain and vertigo. The current literature is also reviewed.

OBSERVATIONS The authors reviewed the data of a young male with a 2-month history of neck pain and vertigo. Magnetic resonance imaging of the brain and cervical spine showed an intramedullary mass at C2–3 with a syrinx extending into the cervicomedullary junction. Laminectomy, myelotomy, and microsurgical excision of the mass under intraoperative neurological monitoring (IONM) were done. Postoperative pathology reported the specimen as a schwannoma.

LESSONS Gross-total resection of a schwannoma using IONM is the treatment of choice because of the lesion's benign nature, a better prognosis, and defined cleavage plane. Schwannomas should be included in the differential diagnosis of intramedullary spinal tumors. Because of its progressive nature, early surgery is recommended in symptomatic patients.

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KEYWORDS schwannoma; intramedullary spinal tumor; myelopathy

Primary tumors of the spinal cord are infrequent and comprise approximately 2% to 10% of all central nervous system (CNS) tumors.1 Typically, these tumors are of a benign nature and are categorized into extradural, intradural-extradural, and intradural spinal tumors based on their locations. Intradural spinal cord tumors are the least common among them all, accounting for 2% to 5% of all spinal tumors. They originate from the glial cells and grow inside the spinal cord, invading and destroying the gray and white matter.2 It has been postulated that the underlying cause of these tumors may be attributed to the presence of small perivascular bundles of peripheral nerves within the spinal cord.3 Few of them have been documented at the level of the conus medullaris, and the majority are in the cervical region.4 As previously reported, 30% of primary intraspinal neoplasms are schwannomas.5 Intramedullary schwannomas account for 0.3% of intraspinal tumors and 1.1% of intraspinal schwannomas.6 However, intramedullary schwannomas are rarely seen and account for 1.1% of all spinal schwannomas.7 The Schwann cell is thought to be the schwannoma’s cell of origin, although it is usually not found in the brain and spinal cord.8 Because of its scarcity among intraspinal tumors and variable clinical presentation, preoperative diagnosis is best accomplished by thoroughly evaluating clinical and radiological characteristics with a high index of suspicion. Spinal magnetic resonance imaging (MRI) is the gold standard for diagnosing intradural spinal tumors.
Still, there are instances in which other diagnostic methods, such as computed tomography (CT) scans, cerebrospinal fluid (CSF) analyses, and neurological examinations, can provide valuable insights. In cases in which patients experience symptoms, early surgical intervention is recommended to prevent further deterioration of neurological deficits. Intramedullary schwannomas are exceedingly rare, but the differential diagnoses of intramedullary tumors have very good outcomes when safe gross-total resection (GTR) is completed. Because this is a rare tumor, only case reports are available, and we discuss all the cases that have been published. Diagnosis is established with histopathology, and the origin is still enigmatic. Safe GTR with intraoperative neuromonitoring (IONM) guidance is the treatment of choice, and the role of radiotherapy is dubious. The present report describes the surgical removal of an intramedullary schwannoma from a 23-year-old male together with subsequent histological confirmation.

Illustrative Case
Clinical History
A 23-year-old male presented with a 2-month history of neck pain and vertigo. There was no other preexisting illness. Physical and neurological examinations were within normal limits with no neurological deficits. Routine blood investigations at admission were also within normal limits.

Imaging Examination
MRI of the cervical spine and brain was performed with contrast. Imaging revealed an intramedullary tumor in the cord at the C2–3 level with a syrinx from the lower medulla until C5. The patient was diagnosed with C2–3 intramedullary space-occupying lesion (SOL; Fig. 1).

Management
After a pre-anesthetic check, the patient underwent C2–4 laminectomy and midline myelotomy while under endotracheal tube intubation general anesthesia. Gross-total excision of the lesion under IONM guidance was performed. The patient was positioned prone with the head held in a Mayfield 3-pin head holder; a midline vertical skin incision was made from C1 to C5. After subperiosteal dissection and muscle retraction, C2–4 laminectomy was performed. Under a microscope, durotomy was performed and retracted followed by a midline myelotomy. The tumor was identified and removed using cavitron ultrasonic surgical aspirator (CUSA), and hemostasis was achieved. The dura mater was closed watertight, and a drain was placed. Throughout the procedure, there was no significant drop in motor evoked potentials (MEPs) and somatosensory evoked potentials (SSEPs). The wound was closed in layers, and a sterile dressing was applied (Fig. 2).

Utilizing IONM is essential during the resection of such lesions, as it ensures safe and complete removal of the tumor. In this case, we used SSEPs and MEPs for monitoring the patient’s neural function. Additionally, D wave monitoring could be considered in similar scenarios. The implementation of these neuromonitoring techniques allows real-time assessment of neurological integrity during the surgical procedure, minimizing the risk of postoperative neurological deficits and contributing to favorable patient outcomes.

Histopathology and Immunohistochemistry
The tumor appeared soft, well-defined, moderately vascular, and grayish on gross examination. Microscopically, the section showed lesions composed of a biphasic component, Antoni-A, a hypercellular area, and Antoni-B, a hypocellular area. Cellular areas of the specimen exhibited nuclear palisading around the fibrillary process (Verocay bodies). The cells present in these areas were narrow and elongated and displayed a wavy pattern with tapered ends. These cells were interspersed with collagen fibers. Large vessels were seen in the Antoni-B area (Fig. 3). All findings were suggestive of intramedullary cervical schwannoma. Figure 4 presents an
intraoperative image demonstrating a well-defined lesion at the C2–3 level, which was debulked using CUSA.

At the 2-year follow up, the patient had symptoms of decreased fine touch and vibration below his neck. There was no weakness or neck pain. His imaging showed a resolving syrinx and no residual or recurrent tumor.

**Patient Informed Consent**

The necessary patient informed consent was obtained in this study.

**Discussion**

In this case report, we describe the case of a 23-year-old male patient who was diagnosed with an intramedullary SOL at the C2–3 level. Subsequently, it was revealed that the lesion was an intramedullary schwannoma. Along with neurofibromas, these tumors account for most (30%) primary spinal tumors. Based on a study, the rarity of intramedullary schwannoma is evident, as only 70 cases have been reported up to 2018. One possible explanation for this rarity is the lack of Schwann cells within the CNS. Typically, intramedullary schwannomas manifest as solitary tumors. However, some cases have also been sporadically associated with neurofibromatosis. The male/female ratio of intramedullary schwannoma is approximately 3:1. The most common location of this lesion is the cervical spinal cord (63%), followed by the thoracic (26%) and lumbar (11%) spinal cord.

Common clinical manifestations of intramedullary schwannoma include sensitivity complaints, sphincter dysfunction, muscle fasciculations, and radicular pain initially, which later progresses to pyramidal and sensory tract dysfunctions. It is worth noting that many of the clinical features of our patient were classic, which are well documented in previous studies (Table 1). We know that the brain and spinal cord are devoid of Schwann cells; this makes the pathogenesis of intramedullary schwannoma controversial. However, there have been some proposed hypotheses that try to explain its origin.

These hypotheses include the presence of ectopic Schwann cells during embryological development, the proliferation of Schwann cells ensheathing aberrant intramedullary nerve fibers, the extension of Schwann cells along the intramedullary perivascular nervous plexus, Schwann cells resulting from the differentiation of primitive multipotential mesenchymal cells, and Schwann cells originating from the posterior nerve root at the root entry zone.

**Observations**

In our case, the patient presented with a history of neck pain and vertigo. MRI revealed a solitary lesion with a syrinx in the lower medulla to C5 level. An MRI scan is considered the most helpful for diagnosing intramedullary schwannomas. Generally, the tumor demonstrates hypo-intense signals compared to the spinal cord on T1-weighted scans and hyperintense alerts along with isointense or low signals on T2-weighted scans. Differentiating intramedullary schwannoma from astrocytoma or ependymoma can be challenging; however, the presence of an exophytic component related to the nerve root is suggestive of the former. The presence of a thickened enhancing nerve root connected to the tumor is the primary diagnostic indicator on MRI, although it is seldom observed. A centrally enhancing lesion accompanied by polar cysts suggests a likelihood of ependymoma. Although radiological investigations are often helpful in the preoperative diagnosis, one should always relate clinical and histopathology to prevent the lesion from being misdiagnosed as glioma.

**Lessons**

Intramedullary schwannomas are exceedingly rare. In the differential diagnosis of intramedullary tumors, schwannomas are rare but have very good outcomes when safe GTR is completed.
<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>Age (yrs)/Sex</th>
<th>Location</th>
<th>Symptoms</th>
<th>Procedure/Treatment</th>
<th>Outcome</th>
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<tbody>
<tr>
<td>Dandpat et al., 2021&lt;sup&gt;13&lt;/sup&gt;</td>
<td>17/M</td>
<td>Medulla–C5</td>
<td>Progressive spastic quadriaparesis &amp; paraesthesia for 6 mos</td>
<td>C1–3 laminoplasty</td>
<td>Died after postoperative day 10 due to ventilator-associated pneumonia</td>
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<tr>
<td>Shahab et al., 2022&lt;sup&gt;15&lt;/sup&gt;</td>
<td>52/M</td>
<td>T11</td>
<td>Instability in gait &amp; numbness in bilat lower limbs</td>
<td>T10 partial, T11 complete, &amp; T12 partial laminectomy</td>
<td>No new motor or sensory deficits in lower limb with overall improvement</td>
</tr>
<tr>
<td>Decharin et al., 2022&lt;sup&gt;16&lt;/sup&gt;</td>
<td>63/M</td>
<td>C4–6</td>
<td>Severe spastic tone, progressive weakness, &amp; numbness in both legs &amp; hands since 5 yrs</td>
<td>Laminectomy</td>
<td>6-mo postoperative, patient improved, including better hand grip power &amp; ambulation</td>
</tr>
<tr>
<td>Tahta et al., 2022&lt;sup&gt;17&lt;/sup&gt;</td>
<td>38/M</td>
<td>Cervico-medullary junction</td>
<td>Constant &amp; radiating rt arm &amp; shoulder pain for 1 mo</td>
<td>Craniotomy &amp; myelotomy</td>
<td>Postoperative MRI of brain &amp; cervical spine revealed no residual lesions &amp; symptoms improved gradually</td>
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<tr>
<td>Varshney et al., 2020&lt;sup&gt;18&lt;/sup&gt;</td>
<td>70/M</td>
<td>T11–L2</td>
<td>Progressive paraparesis &amp; sphincter dysfunction for 2.5 yrs</td>
<td>T12–L1 laminectomy</td>
<td>Neurological functions remained unchanged postoperative; 3 mos later, motor function was 4/5 in lt &amp; 3/5 in rt lower extremity &amp; bladder control regained</td>
</tr>
<tr>
<td>Dai et al., 2019&lt;sup&gt;19&lt;/sup&gt;</td>
<td>34/M</td>
<td>C3–4</td>
<td>Nonradiating neck pain w/ progressive weakness of lt limbs &amp; sensory loss of both lower extremities</td>
<td>C3–4 laminectomy</td>
<td>Postoperative MRI verified successful tumor removal &amp; resolution of cranial spinal cord edema</td>
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<tr>
<td>Navarro Fernández et al., 2018&lt;sup&gt;20&lt;/sup&gt;</td>
<td>19/M</td>
<td>C6–7</td>
<td>Pain, numbness, &amp; dysesthesias in rt arm &amp; hand w/ progressive spastic quadriparesis</td>
<td>C6–7 laminoplasty</td>
<td>At 4-wk FU, strength recovery (score 4– on MRC scale), increased sensitivity, &amp; modified McCormick score III; postoperative MRI indicated complete tumor removal</td>
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<tr>
<td>Jagannatha et al., 2016&lt;sup&gt;21&lt;/sup&gt;</td>
<td>11/M</td>
<td>T8–9</td>
<td>Weakness in both lower limbs</td>
<td>T10–12 laminotomy</td>
<td>Postoperative progress smooth &amp; significant improvement at 6-mo FU</td>
</tr>
<tr>
<td>Li et al., 2017&lt;sup&gt;22&lt;/sup&gt;</td>
<td>30/M</td>
<td>C3–5</td>
<td>Lt chest pain &amp; arm weakness</td>
<td>—</td>
<td>Died before procedure</td>
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<tr>
<td>Gupta et al., 2015&lt;sup&gt;23&lt;/sup&gt;</td>
<td>48/M</td>
<td>C3–4</td>
<td>Paraesthesia of all limbs, progressive spastic quadriparesis, &amp; urinary urgency for 5 mos</td>
<td>C3–4 laminectomy</td>
<td>Postoperative, reduced limb spasticity &amp; improved bilateral grip strength; at 1-yr FU, notable decrease in paraesthesia &amp; spasticity</td>
</tr>
<tr>
<td>Karatay et al., 2017&lt;sup&gt;24&lt;/sup&gt;</td>
<td>30/F</td>
<td>T12–L1</td>
<td>Gait disturbance, back pain, &amp; progressive numbness in both legs for 2 mos</td>
<td>T12 laminectomy</td>
<td>Noticeable improvement observed in neurological exam</td>
</tr>
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<td>Ejejbouri et al., 2013&lt;sup&gt;24&lt;/sup&gt;</td>
<td>10/M</td>
<td>T7–9</td>
<td>Bladder &amp; bowel incontinence w/ lower limb weakness</td>
<td>T7–9 laminectomy</td>
<td>At 15-mo FU, neurological recovery; sensation &amp; strength in all limbs returned to normal, but deep tendon reflexes in rt lower limb remained exaggerated</td>
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Because this is a rare tumor, only case reports are available, and we have discussed all cases that have been published. The diagnosis is established with histopathology, and the origin is still enigmatic. It is mostly seen in young males. MRI is the imaging modality of choice, and the cervical spine is the most common location. Safe GTR with IOM guidance is the treatment of choice, and the role of radiotherapy is dubious.

### References


### TABLE 1. Summary of some of the previous studies on intramedullary schwannoma

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<thead>
<tr>
<th>Authors &amp; Year</th>
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<th>Procedure/Treatment</th>
<th>Outcome</th>
</tr>
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<tr>
<td>Ohttonari et al., 2009</td>
<td>29/M</td>
<td>Conus medullaris</td>
<td>Sexual impotence, bladder dysfunction, &amp; paraesthesia in buttocks for 8 mos</td>
<td>T12–L1 laminectomy</td>
<td>Initially, deterioration in bladder function, but gradually recovered over time</td>
</tr>
<tr>
<td>Hayashi et al., 2009</td>
<td>78/F</td>
<td>T11–L1</td>
<td>Pain &amp; numbness in both lower limbs</td>
<td>T10–L2 laminectomy</td>
<td>Postoperative, transient worsening of lower-extremity weakness &amp; sensory disturbance, but pain improved; JOA score remained at 2 points</td>
</tr>
<tr>
<td>Nicácio et al., 2009</td>
<td>40/M</td>
<td>C4–6</td>
<td>Sphincter disturbances &amp; spastic quadriaparesis</td>
<td>C3–5 laminotomy</td>
<td>At the 24-mo mark, a notable regression of motor &amp; sphincter dysfunction observed</td>
</tr>
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</table>

FU = follow-up.
Disclosures
The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author Contributions
Conception and design: Duvuru, Sanker, Syed, Ghosh. Acquisition of data: Duvuru. Analysis and interpretation of data: Duvuru, Sanker, Mishra. Drafting the article: Dave, Sanker, Syed, Ghosh. Critically revising the article: Dave, Duvuru, Sanker, Mishra, Ghosh. Reviewed submitted version of manuscript: Dave, Duvuru, Sanker, Ghosh. Approved the final version of the manuscript on behalf of all authors: Dave. Statistical analysis: Duvuru. Administrative/technical/material support: Duvuru. Study supervision: Duvuru, Sanker.

Supplemental Information
Previous Presentations
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Correspondence
Tirth Dave: Bukovinian State Medical University, Chernivtsi, Ukraine. tirth.snehal.dave@gmail.com.