Cervical, Intraspinal Tumor: A Case Report and Review of the Literature

Abstract
Solitary fibrous tumors (SFTs) are rare, spindle cell neoplasms of the mesenchymal origin. Lesions localized to the spine are exceptionally uncommon, only described in the literature in case reports and small case series. While these lesions are typically benign, there are a few reports in which they recur or present as malignancies. The patient presented in the case herein was a 72-year-old male, who presented with a 1-year history of lower extremity weakness, pain, and numbness and was found to have a cervical, intradural extramedullary tumor. In addition to the case report, the authors perform a thorough review of all previously published cases of spinal SFT.

Keywords: Solitary fibrous tumor, spine, tumor

Introduction
Solitary fibrous tumors (SFTs) are rare spindle cell neoplasms of the mesenchymal origin. These lesions were first described in 1931[1] as a localized form of pleural mesothelioma. In the time since, SFTs have been reported in numerous extra-pleural locations, including the orbit, upper respiratory tract, nasopharyngeal sinuses, perioesum, soft tissues, skin, prostate, meninges, epiglottis, liver, and thyroid.[2-12]

The first seven cases of SFT of the central nervous system were described in 1996 by Carneiro et al., two of which were intraspinal.[13] A comprehensive review by Bisceglia et al. in 2011 found that of the 220 cases of SFT reported in the literature since 1996, roughly one-fifth were intraspinal lesions.[14] Since the initial report of intraspinal SFTs, approximately 85 additional intraspinal SFTs have been described in the literature.[13-72] Presented herein is an illustrative case of an SFT with an extensive literature review, focused predominately on cervical/thoracic, intradural, and extramedullary tumors.

Illustrative Case Solitary Fibrous Tumor
The patient, in this case, was a 72-year-old male who presented with a 1-year history of lower extremity weakness, pain, and numbness. Symptoms were initially localized to the right lower extremity and progressively worsened, evolving to include the left lower extremity. Three weeks before presentation, the patient began having difficulty walking and experiencing instability at the knee joint. Interestingly, the patient reported that the pain was limited to his right lower extremity, while the only symptom on the left side was weakness. The patient underwent magnetic resonance imaging (MRI) of the cervical and thoracic spine and was found to have an intradural enhancing lesion, with associated spinal cord compression [Figure 1a and b].

The mass, located along the ventral and right lateral surface of the thoracic cord at the C7 vertebral level, appeared to be causing prominent mass effect, severe canal narrowing, and hydrosyringomyelia.

The patient underwent C5–C7 laminectomies with intraoperative neuromonitoring. After opening the dura, a large, extremely hard, and fibrous extramedullary tumor was found closely adherent to the spinal cord [Figure 2a]. The tumor was debulked, and a biopsy was sent for frozen section. Subsequently, microdissection was performed to remove the tumor from the cord. All tumors were removed, including some areas with a poor resection plane [Figures 1c, d and 2b]. The operation was well tolerated by the patient, with no complications during the follow-up period.

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Histological Findings

Histologically, the specimen was composed of spindle-shaped cells arranged in intersecting fascicles within an abundant collagen network. Immunohistochemical stains demonstrated that the mass was strongly, diffusely positive for CD34 and BCL-2 and negative for S100 and EMA. In addition, stain for Ki-67 revealed a low proliferation rate. Collectively, these findings supported a diagnosis of SFT.

Discussion

Spinal SFTs can be classified, according to their compartment of development, as intramedullary, intradural extramedullary, or extradural.[41] To the authors’ knowledge, there have been only three cases of cervical, intradural extramedullary SFTs, including the present case [Table 1].[18,20,21,24,26,31,34,46,47,59,61,62,68,71,73‑75] As these lesions are rare, no large case series have been performed on spinal SFTs; however, comprehensive reviews have been conducted by Fargen et al.,[76] Bisceglia et al.,[14] and more recently Albert and Gokden.[16] According to these studies, the majority of spinal SFTs are intradural and occur in the cervical and thoracic segments. Males and females are roughly equally affected.[14,76] Fargen et al. reported that the patients included in their analysis almost universally presented with pain, sensory loss, motor weakness, urinary dysfunction, or a combination of these symptoms.[76] Generally, SFTs are considered to be benign or indolent; however, malignant cases have been reported in the literature.[77‑79] The current consensus treatment for spinal SFTs consists of surgical resection via laminotomy or laminectomy. The extent of surgical resection has been implicated as the most important prognostic factor.[80,81] Fargen et al. reported that 25% of cases exhibited recurrences, half of which underwent subtotal resection (STR). Further analysis showed that STR was associated with a 16-fold increased odds of recurrence (odds ratio 15.9, 95% confidence interval 5.5–46.1).[76] The role of radiotherapy and chemotherapy is yet to be defined against these tumors.[24] However, it is suggested that there is minimal benefit in benign cases and that these therapies are likely ineffective for malignant SFTs.[14]

Although gross total resection carries a good prognosis and lower risk of recurrence, these resections are not without risks.[24] As treating these lesions requires a challenging microsurgical resection, the possibility of postoperative morbidity remains. Damage to the spinal tracts, particularly in intramedullary SFTs, is a potential complication and must be mitigated through the use of neuromonitoring.[20,25] In cases where there is no clear plane of resection and close adherence to the spinal cord, the use of cutting instruments and lasers is found to be useful.[77]

The ability to distinguish SFTs from other spindle cell tumors is important for clinicians as these lesions have similar features. The differential diagnosis includes meningioma, schwannoma, and neurofibroma.[36] A definitive diagnosis can be made through a combination of histopathological and immunohistochemical analysis.[82] Histologically, SFT cells are found encircled by dense collagen networks in fascicular, storiform, herringbone, or patternless
Table 1: Literature review of reported cases of cervical/thoracic, intradural extramedullary solitary fibrous tumors

<table>
<thead>
<tr>
<th>Authors (years)</th>
<th>Age/sex</th>
<th>Location</th>
<th>Compartment</th>
<th>Treatment</th>
<th>Follow-up</th>
<th>Outcome/notes</th>
</tr>
</thead>
<tbody>
<tr>
<td>Malek et al. (1997)</td>
<td>33/male</td>
<td>T7-8</td>
<td>Intradural, extramedullary</td>
<td>GTR</td>
<td>4 months disease-free</td>
<td></td>
</tr>
<tr>
<td>Brunori et al. (1999)</td>
<td>46/female</td>
<td>T12-L1</td>
<td>Intradural, extramedullary</td>
<td>GTR</td>
<td>7 months no recurrence</td>
<td></td>
</tr>
<tr>
<td>Vorster et al. (2000)</td>
<td>51/male</td>
<td>T2-3</td>
<td>Intradural, extramedullary</td>
<td>GTR</td>
<td>12 months disease-free</td>
<td></td>
</tr>
<tr>
<td>Kurtkaya et al. (2001)</td>
<td>70/female</td>
<td>T3</td>
<td>Intradural, extramedullary</td>
<td>GTR</td>
<td>15 months no recurrence</td>
<td></td>
</tr>
<tr>
<td>Caroli et al. (2004)</td>
<td>54/male</td>
<td>C7-T1</td>
<td>Intradural, extramedullary</td>
<td>GTR</td>
<td>18 months no recurrence</td>
<td></td>
</tr>
<tr>
<td>Pizzolitto et al. (2004)</td>
<td>36/male</td>
<td>T7-8</td>
<td>Intradural, extramedullary</td>
<td>GTR</td>
<td>Recurrence 14 years later $\rightarrow$ S3-5 and coccygeal nerve roots; intradural, extramedullary $\rightarrow$ GTR</td>
<td></td>
</tr>
<tr>
<td>Pakasa et al. (2005)</td>
<td>27/male</td>
<td>T5-7</td>
<td>Intradural, extramedullary</td>
<td>STR</td>
<td>18 months disease-free</td>
<td></td>
</tr>
<tr>
<td>Arantes et al. (2009)</td>
<td>22/male</td>
<td>T1-2</td>
<td>Intradural, extramedullary</td>
<td>GTR</td>
<td>Tumor embedded in posterior nerve rootlets</td>
<td></td>
</tr>
<tr>
<td>Bisceglia et al. (2011)</td>
<td>47/male</td>
<td>T3-4</td>
<td>Intradural, extramedullary</td>
<td>GTR</td>
<td>11.5 years disease-free</td>
<td></td>
</tr>
<tr>
<td>Vassal et al. (2011)</td>
<td>52/female</td>
<td>T8-9</td>
<td>Extradural and intradural, extramedullary</td>
<td>GTR</td>
<td>62 months disease-free</td>
<td></td>
</tr>
<tr>
<td>Mariniello et al. (2012)</td>
<td>75/female</td>
<td>T6-7</td>
<td>Intradural, extramedullary</td>
<td>GTR</td>
<td>1 year disease-free</td>
<td></td>
</tr>
<tr>
<td>Brigui et al. (2013)</td>
<td>56/male</td>
<td>T6-7</td>
<td>Intradural, extramedullary</td>
<td>GTR</td>
<td>29 months disease-free</td>
<td></td>
</tr>
<tr>
<td>Hwang et al. (2014)</td>
<td>48/male</td>
<td>C7-T1</td>
<td>Intradural, extramedullary</td>
<td>STR</td>
<td>No recurrence at 6 months</td>
<td></td>
</tr>
<tr>
<td>Robert et al. (2014)</td>
<td>49/female</td>
<td>T9-10</td>
<td>Intradural, extramedullary</td>
<td>STR</td>
<td>No recurrence at 6 months</td>
<td></td>
</tr>
<tr>
<td>Yuan et al. (2014)</td>
<td>48/male</td>
<td>T9</td>
<td>Intradural, extramedullary</td>
<td>GTR</td>
<td>Dumbbell-shaped; communicating with thoracic cavity</td>
<td></td>
</tr>
<tr>
<td>Sade et al. (2015)</td>
<td>43/male</td>
<td>Thoracic</td>
<td>Intradural, extramedullary</td>
<td>Surgery</td>
<td>Local recurrence and pulmonary metastases at 5 months $\rightarrow$ palliative radiotherapy and chemotherapy</td>
<td></td>
</tr>
<tr>
<td>Biswas et al. (2017)</td>
<td>35/female</td>
<td>T10-11</td>
<td>Intradural, extramedullary, and extramedullary</td>
<td>STR (2 stages)</td>
<td>Malignant tumor</td>
<td></td>
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</tbody>
</table>

Present case

<table>
<thead>
<tr>
<th>Age/sex</th>
<th>Location</th>
<th>Compartment</th>
<th>Treatment</th>
<th>Follow-up</th>
<th>Outcome/notes</th>
</tr>
</thead>
<tbody>
<tr>
<td>72/male</td>
<td>C6-7</td>
<td>Intradural extramedullary</td>
<td>GTR</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

GTR – Gross total resection; STR – Subtotal resection; Surgery – Otherwise unspecified surgical resection

arrangements on hematoxylin and eosin staining.\[^{[13,83]}\]
Positive staining for CD34, vimentin, BCL-2,\[^{[13,46,84,85]}\] and CD99\[^{[86]}\] and negative staining for EMA and S-100\[^{[84]}\] are hallmark findings in SFTs.

Hemangiopericytomas (HPCs) display many of the same characteristics as SFTs, sometimes making differential diagnosis a challenge. A new paradigm has gained traction among pathologists in the past decade, which views HPC as a variant within the broader spectrum of SFT.\[^{[87]}\] Recent evidence supports this view, including a study by Schweizer et al., where a similar NAB2-STAT6 fusion protein was found in both SFT and HPC.\[^{[88]}\] However, this is not universally accepted. Given the better prognosis associated with SFTs, particularly in the central nervous system, most experts retain that distinguishing the two entities remains clinically significant.\[^{[14,89]}\]

On MRI, SFTs appear isointense on T1-weighted sequences and hypointense on T2-weighted sequences.\[^{[36,47]}\] Intraoperative appearance of SFTs can aid in distinguishing them from other, similar neoplasms. Intradural extramedullary SFTs lack involvement of the spinal roots (unlike neurinomas) and have a hard tumor consistency, little to no vascularization, and an absent or weak dural adherence (unlike meningiomas). In addition, unlike schwannomas and meningiomas, there is a firm attachment to the spinal cord and no clear arachnoidal interface. Intramedullary SFTs also have a hard consistency (unlike metastases and astrocytomas) and scarce vascularization (unlike hemangioblastomas).\[^{[47]}\]

**Conclusion**

We report a rare case of a cervical, intradural extramedullary SFT of the spinal cord. To date, with the inclusion of the case herein, there are only three similar cases reported in the literature. Thus, continual reports must be contributed to inform clinicians regarding how to identify, differentiate, classify, and treat these lesions.

**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and
other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest
There are no conflicts of interest.

References
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