Neuroblastoma-like schwannoma of lumbar spinal nerve root

Case report

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Neuroblastoma-like schwannoma is a rare nerve sheath tumor with histological features resembling a neuroblastoma. A comprehensive literature review identified only 10 previous case reports of this condition. The authors present the first reported case of a neuroblastoma-like schwannoma at a spinal nerve root. The patient, a 61-year-old woman, presented with severe pain in the right lower extremity that failed to resolve after conservative management. Magnetic resonance imaging revealed an intradural enhancing lesion extending out of the right neural foramen at L1–2. A right L1–2 hemilaminectomy and facetectomy with gross-total resection of the tumor was performed without complications. Neuroblastoma-like schwannoma was diagnosed based on histopathological examination of the biopsied tumor specimen. A postoperative course of serial examination and imaging was chosen based on a suspected benign postoperative course as in the case of a completely resected schwannoma. The authors present the novel case of neuroblastoma-like schwannoma at a lumbar spinal nerve root and describe the distinguishing pathological features of this rare lesion. (DOI: 10.3171/2010.3.SPINE09251)

Key Words • neuroblastoma • schwannoma • tumor • nerve root

Schwannomas, or neurilemomas, are benign tumors of the peripheral nerve sheath with rare occurrences of malignant degeneration.3 Schwannomas typically appear as well-defined, encapsulated tumors composed predominantly of spindle-shaped neoplastic Schwann cells. These lesions often demonstrate a histological biphasic pattern of Antoni A (compact cellular areas) and Antoni B (loosely arranged hypocellular regions).3,4,8 Rarer variants of schwannoma include “ancient” schwannoma, melanotic schwannoma, plexiform schwannoma, epithelioid schwannoma, and the exceptionally rare neuroblastoma-like schwannoma.8,15

Neoplasms that may cause diagnostic confusion with neuroblastoma-like schwannoma include neuroblastoma and Ewing sarcoma/PNET. Neuroblastoma typically appears in young children, with approximately 60% of cases occurring within the first 2 years of life.1 This lesion may present anywhere along the distribution of the sympathetic chain.1 Microscopically, neuroblastomas are densely cellular with characteristic Homer-Wright rosettes consisting of clusters of neuroblasts surrounding regions of eosinophilic neuropil. Ewing sarcoma/PNET is a family of small round cell tumors that often demonstrate Homer-Wright rosettes. These tumors commonly occur in the soft tissue or bone of patients younger than 30 years old.13

To date, only 10 reported cases of schwannoma have pathological features consistent with neuroblastoma.3–6,8,14 We report the first case of a spinal neuroblastoma-like schwannoma, located at the right L-1 spinal nerve root.

Case Report

History and Examination. This 61-year-old woman presented with the chief complaint of chronic lower back pain unremitting for several years. After a recent episode of pain, her symptoms progressed to severe spasm and cramping in her lower back and right lower extremity. At presentation she had a constant, aching, radiating pain in her right lower extremity, which was exacerbated by standing, walking, or bending. She cited generalized...
weakness in her right lower extremity and abnormal sensation localized on her right anterior thigh. Attempts at conservative management were unsuccessful in resolving her symptoms. The patient’s medical history was significant only for resection of a benign right ovarian tumor. She denied any family history of neurocutaneous syndromes and lacked the typical stigmata of neurofibromatosis Type 1.

On examination, the patient had intact motor function with no apparent atrophy, fasciculations, or altered reflexes. Her only deficit was decreased sensation to light touch and pin-prick in the right L-2 distribution. Magnetic resonance imaging of the lumbosacral spine revealed a 12 × 10 mm intradural lesion within the right neural foramen at L1–2, severely compressing or involving the nerve root at that level (Fig. 1). The lesion was hypointense on T1-weighted MR imaging and enhanced avidly with contrast administration. Imaging results favored the presumptive diagnosis of nerve sheath tumor (schwannoma or neurofibroma) versus a juxtafacet cyst, based on the signal characteristics and intradural location. The patient consented to an operative course of nerve root decompression and tumor resection. Risk of late angulation or collapse due to unilateral facet sacrifice during surgery was included in the preoperative discussion.

Operation. The patient underwent a right L1–2 hemilaminectomy with an aggressive (near total) facetectomy and foraminotomy. After the L-1 lamina and a portion of the superior L-2 lamina were unroofed using a combination of rongeurs and drill, the L-1 pedicle and nerve root exiting through its foramen were identified. The dural root sleeve was markedly swollen and firm, confirming the intradural location of the mass as noted on preoperative MR imaging. The inferior articulating process of L-1 was resected using a drill and rongeurs. The neural foramen was then further unroofed with Kerrison rongeurs. Removal of the bulk of the L-2 superior facet provided additional exposure and decompression.

The dura was opened in the direction of the root sleeve, and a distinct plane between the firm, tan tumor and normal nerve fascicles facilitated circumferential microsurgical resection. After a clear plane was established proximally, the tumor was found to be clearly attached to a small distal rootlet. Intraoperative electromyography with monitoring of all major lower-extremity muscle groups did not suggest a motor origin for the lesion. The attached rootlet, which was presumed to be sensory in origin, was transected. The remainder of the tumor was delivered easily and sent in 2 pieces for pathological analysis. A portion of the tumor bivalved in the operating room for closer inspection grossly resembled the typical appearance of a schwannoma. With close examination of the root from its origin to its distal aspect, a portion of the nerve immediately proximal to the site of resection appeared somewhat firm and edematous. This finding appeared more consistent with reactive changes than with residual tumor. Gross-total resection was considered to have been achieved. The dura was closed with suture followed by fibrin sealant, and a standard wound closure was performed.

Histopathological Findings. The biopsy specimen showed a moderately cellular neoplasm composed predominantly of giant rosettes, characterized by a radial arrangement of neoplastic cells around large central eosinophilic collagenous regions (Figs. 2A and B). Occasional smaller rosettes were also interspersed throughout the specimen. The tumor cells exhibited round-to-oval
hyperchromatic nuclei, scant cytoplasm and minimal atypia. Dystrophic calcifications were noted, and no mitotic figures or necrosis was identified. The neoplastic cells were strongly immunoreactive for S100 protein (Fig. 2C). The cells were negative for expression of synaptophysin (Fig. 2D), neuron-specific enolase, keratin (AE1/AE3), and CD99 (MIC2). The MIB-1 labeling index was less than 1%. The histological features of large rosettes in conjunction with the immunohistochemical results were consistent with a diagnosis of neuroblastoma-like schwannoma.

Postoperative Course. The patient was hospitalized overnight, and examination revealed full motor strength with resolution of her radicular pain symptoms. We were encouraged to find that gross dermatomal examination of light touch in the affected distribution did not reveal significant new deficits compared with her preoperative examination. Magnetic resonance imaging with contrast administration at 2-month follow-up showed postoperative changes without any abnormal enhancement, consistent with complete resection of the lesion (Fig. 3).

Discussion

Intradural extramedullary tumors constitute as many as 40% of spinal tumors and arise from nerve root elements or the meninges. These typically benign, slow-growing lesions cause radiculopathy by nerve root compression. Our patient's intradural lesion is the only reported neuroblastoma-like schwannoma of spinal origin. A literature review yielded 10 cases of neuroblastoma-like schwannoma, underscoring the novelty of a spinal nerve root lesion within this rare class of tumors (Table 1).

The original 3 cases published by Goldblum et al. first brought attention to a variant of schwannoma with neuroblastoma-like histological appearance. Since its identification as a pathologically distinct lesion, 7 addi-
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Three reports described subcutaneous nodules at various locations (such as the knee, vulva, thigh, neck, palm, and flank) while Kukreja et al. described the first case of neuroblastoma-like schwannoma of the orbit. Seven of 10 cases were reported in women and 2 in men; sex was not specified in 1 case (Table 1). We present an additional case of neuroblastoma-like schwannoma in a woman, further contributing to a distribution favoring a higher female-to-male ratio. However, with so few reported cases of this schwannoma variant, the correlation between prevalence and sex cannot be determined.

Optimal treatment for this rare tumor variant is undocumented. Gross-total resection of the lesion using microsurgical technique to preserve normal neural tissue is suggested. Given the benign nature of the lesion, postoperative management includes long-term serial MR imaging to detect tumor regrowth. The 10 reported cases in addition to the present case showed no evidence of recurrence, with an average of 15 months follow-up. However, due to the exceedingly rare nature of these lesions, a definitive protocol for surgical and postoperative management has not been established. Care must be taken to avoid confusing neuroblasto-

TABLE 1: Summary of reported cases of neuroblastoma-like schwannoma

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>Age (yrs), Sex</th>
<th>Tumor Location</th>
<th>Tumor Size (cm)</th>
<th>Treatment</th>
<th>Follow-Up</th>
</tr>
</thead>
<tbody>
<tr>
<td>Goldblum et al., 1994</td>
<td>44, F</td>
<td>neck (superficial soft tissue, partly adherent to underlying skeletal muscle)</td>
<td>4.5</td>
<td>excision</td>
<td>no evidence of recurrence or metastasis 27 mos postop</td>
</tr>
<tr>
<td></td>
<td>32, M</td>
<td>palm</td>
<td>1.5</td>
<td>excision</td>
<td>no evidence of recurrence or metastasis 12 mos postop</td>
</tr>
<tr>
<td></td>
<td>46, F</td>
<td>right flank (eroding a portion of 11th rib)</td>
<td>1.3</td>
<td>excision</td>
<td>no evidence of recurrence or metastasis 2 mos postop</td>
</tr>
<tr>
<td>Bhatnagar et al., 1998</td>
<td>53, F</td>
<td>vulva</td>
<td>2.0</td>
<td>excision</td>
<td>no evidence of recurrence or metastasis 18 mos postop</td>
</tr>
<tr>
<td></td>
<td>35, F</td>
<td>knee (subcutaneous)</td>
<td>1.8</td>
<td>excision</td>
<td>no evidence of recurrence or metastasis 24 mos postop</td>
</tr>
<tr>
<td>de Saint et al., 2003</td>
<td>62, NA</td>
<td>neck (subcutaneous)</td>
<td>0.6</td>
<td>excisional biopsy</td>
<td>NA</td>
</tr>
<tr>
<td>Lewis et al., 2005</td>
<td>40, F</td>
<td>ankle (subcutaneous)</td>
<td>0.8</td>
<td>excision</td>
<td>no evidence of recurrence or metastasis 32 mos postop</td>
</tr>
<tr>
<td></td>
<td>24, F</td>
<td>knee (subcutaneous)</td>
<td>3.9</td>
<td>excision</td>
<td>no evidence of recurrence or metastasis 1 month postop</td>
</tr>
<tr>
<td>Vélez et al., 2006</td>
<td>29, F</td>
<td>buttock (subcutaneous)</td>
<td>NA</td>
<td>excision</td>
<td>NA</td>
</tr>
<tr>
<td>Kukreja et al., 2007</td>
<td>16, M</td>
<td>Lt orbit (displacing optic nerve temporally and medial rectus muscle laterally)</td>
<td>2.1</td>
<td>excision</td>
<td>proptosis reduced, left eye acuity (20/400 preop) improved to 20/200 4 wks postop and remained stable; no evidence of recurrence or metastasis 12 mos postop</td>
</tr>
<tr>
<td>present case</td>
<td>61, F</td>
<td>rt L-1 nerve root</td>
<td>1.2</td>
<td>excision</td>
<td>neurologically intact at discharge, no evidence of recurrence or metastasis 8 mos postop</td>
</tr>
</tbody>
</table>

* NA = not available.
ma-like schwannoma with lesions of similar pathology. A distinction should be made from tumors with Homer-Wright rosettes, notably neuroblastoma and Ewing sarcoma/PNET. In neuroblastoma, Homer-Wright rosettes are often identified and are typically smaller than those observed in neuroblastoma-like schwannoma. In addition, neuroblastomas commonly display mitotic figures, hemorrhage, and tumor necrosis, pathological features usually absent in neuroblastoma-like schwannoma. Furthermore, neuroblastomas typically demonstrate immunoreactivity for synaptophysin, chromogranin, and neuron-specific enolase, antibodies that are negative in schwannomas. In contrast, schwannomas characteristically are strongly S100 positive, although S100 expression may be diminished in Antoni B regions and some neuroblastomas may exhibit S100 positivity within regions of supporting stroma. Ewing sarcoma/PNET are small, round, blue cell tumors that also exhibit Homer-Wright rosettes and should be included in the differential diagnosis of a neuroblastoma-like schwannoma. These tumors characteristically demonstrate immunoreactivity for CD99 and may occasionally express AE1/AE3 unlike neuroblastoma-like schwannomas. The neoplastic cells in our patient’s tumor were strongly S100 positive. These cells were negative for the expression of neuronal markers synaptophysin and neuronal-specific enolase. They were also negative for CD99 (MIC2), a marker commonly expressed in Ewing sarcoma/PNET. In addition, the large rosettes identified in our patient’s tumor were typical of those described in other reports of this unusual tumor. Finally, the low MIB-1 labeling index, lack of mitotic activity, and absence of necrosis were all features consistent with a benign neoplasm. The histological findings, in conjunction with the immunohistochemical results, supported a diagnosis of a schwannoma with neuroblastoma-like differentiation. The benign histological features enumerated above further support conservative postoperative management.

In summary, we report a novel case of neuroblastoma-like schwannoma at the spinal root of L-1. This lesion was an uncommon, yet histologically distinct variant of schwannoma. Careful pathological examination is needed to differentiate and diagnose such tumors accurately.

Disclosure

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 to the study and manuscript preparation include the following: Conception and design: all authors. Acquisition of data: all authors. Analysis and interpretation of data: all authors. Drafting the article: all authors. Critically revising the article: all authors. Reviewed final version of the manuscript and approved it for submission: Theodore. Statistical analysis: all authors. Administrative/technical/material support: all authors. Study supervision: all authors.

References


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