Radiation-induced intradural malignant peripheral nerve sheath tumor of the cauda equina with diffuse leptomeningeal metastasis

Case report

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Malignant peripheral nerve sheath tumors (MPNSTs) are rare, affecting only a small portion of the general population. In many cases, MPNSTs occur in association with neurofibromatosis Type 1 and at times arise secondary to previous radiation therapy (RT). These tumors can be found essentially anywhere a peripheral nerve is present, but they rarely originate primarily from the spinal nerve or cauda equina and cause leptomeningeal spread. This report describes the treatment course of a 43-year-old man with a history of testicular seminoma treated with RT a decade before, who was found to have a large sacral MPNST. The patient underwent complete sacrectomy for gross-total resection. Despite this effort, he was eventually found to have metastatic lesions throughout the spine and brain, ultimately resulting in acute hydrocephalus and death. Biopsy results of these metastatic lesions proved to be characteristic of his original MPNST. The literature is also reviewed and the diagnostic modalities, management strategies, and prognosis of MPNST are discussed.

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KEY WORDS • cauda equina • malignant peripheral nerve sheath tumor • radiation therapy • oncology

The term “malignant peripheral nerve sheath tumor” (MPNST) was coined by the World Health Organization to describe tumors originating from the connective tissue surrounding peripheral nerves, and it replaced previous nomenclature, including malignant neurilemmoma, malignant schwannoma, and neurofibrosarcoma. These tumors are rare, with an incidence of 0.001% in the general population.15,17 Approximately 20%–50% occur in association with neurofibromatosis Type 1,5,10,17 and up to 11% arise secondary to previous radiation therapy (RT).10,27 The incidence of MPNST has a slight female predominance, and the tumor is seen in patients of all ages, with most presenting in their 3rd decade of life.10 The most common locations for MPNSTs are in the extremities and trunk; they occur less commonly in the head and neck.27 Primary intradural MPNST involving the cauda equina is extremely rare, with only 4 case reports in the literature.1–3,5,9,21,25,28,29 Leptomeningeal spread from such lesions is extremely uncommon.29 We report a unique case of a patient with RT-induced intradural MPNST of the spine complicated by diffuse CNS and leptomeningeal metastases. We also review the literature to discuss the diagnostic modalities, management strategies, and prognosis of MPNST.

Case Report

Presentation and History. A 43-year-old man with a history of testicular seminoma presented with severe back pain and paresthesia radiating to his left buttock and posterolateral thigh for 5 months. He had undergone orchiectomy and adjuvant RT to his right retroperitoneal and inguinal nodal chain 10 years before. There was no personal or family history of neurofibromatosis. On examination, he was neurologically intact. Oral analgesia and physiotherapy were initiated but failed to resolve his symptoms. Sagittal and axial T1-weighted contrast-enhanced MRI of the spine revealed a contrast-enhancing, 5.8 × 5.3–cm sacral mass that encased the S-2 and S-3 vertebral bodies and extended into the sacral spinal canal, involving bilateral sacral nerve roots (Fig. 1).

Abbreviations used in this paper: MPNST = malignant peripheral nerve sheath tumor; RT = radiation therapy.

* Dr. Lau and Mr. Moon contributed equally to this work.
Treatment Course and First Surgery. Biopsy guided by CT revealed a high-grade spindle cell sarcoma, consistent with MPNST. The tumor was positive for S100 protein and negative for caldesmon, desmin, and muscle-specific actin. The patient underwent 4 cycles of neoadjuvant chemotherapy, alternating between ifosfamide/doxorubicin and ifosfamide/etoposide, to decrease tumor burden and thus increase the potential for complete resection. Despite this, the tumor continued to grow to $6.2 \times 6.3$ cm, and the patient developed difficulty with urination. At this time, a decision was made to proceed with complete resection. The patient was then referred to an outside institution for surgical management of the MPNST.

The patient underwent complete sacrectomy via a 2-staged approach. By report, an anterior approach was taken to free up the anterior bony portions of the sacrum via bilateral sacroiliac osteotomy, bilateral iliac osteotomy, and osteotomy of the L-5 vertebral body. The second stage consisted of a posterior approach for en bloc removal of the sacral tumor and spinal reconstruction. Osteotomies of the posterior portions of the sacrum, ilium, and L-5 vertebral body were performed, freeing the sacrum and tumor from all bony connections. The sacrum (containing the tumor) was removed en bloc. Therefore, the physical nature and vascularity of the tumor were not directly encountered. The L-5 nerve roots were attached to the specimen by soft-tissue connections during removal but were easily dissected free. Negative margins were confirmed. Spinal reconstruction consisted of L2–5 pedicle screw placement, double iliac screw fixation, and dual rod placement, in conjunction with placement of bilateral fibular strut allograft from the supraacetabular bone to the anterior portion of the inferior endplate of L-5. Gross-total resection via sacrectomy was achieved (Fig. 2A and C). However, the surgery was complicated by multiple issues, including postoperative wound infection, which required surgical debridement, a yearlong course of antibiotics, and pelvic screw fracture (Fig. 2B).

Postoperative Course. The patient had bilateral lumbar radiculopathies/plexopathies after surgery, resulting in bilateral paraparesis with partial motor preservation of knee extensors, thigh adductors, and hip flexors. Sensation to light touch and pinprick was significantly diminished in L5–S4 distribution. At the time of discharge, the patient had Grade 4+ of 5 bilateral knee extension strength, Grade 4− of 5 hip flexor strength, and absent hip extension and ankle motion. In addition, the patient had a neurogenic bowel with flaccid anal sphincter tone and neurogenic bladder, requiring, respectively, colostomy at the time of sacrectomy and self-catheterization.

The patient was evaluated for RT after surgery, but the radiation oncologist felt the patient would not benefit from additional RT at the time, given clear surgical margins and previous RT exposure to the area for seminoma. Instead, the patient underwent an additional 4 cycles of adjuvant chemotherapy with the same preoperative regimen.

Metastasis. The patient subsequently returned to our institution for further management of his MPNST. About 24 months after completing adjuvant chemotherapy and 30 months after sacrectomy, the patient was found to have renal metastasis of his MPNST. He was treated with cryo-
therapy alone, which resulted in remission of his disease, and after his treatment course he continued to do well, attaining Karnofsky Performance Scale score of 70.

Second Surgery and Histopathology Analysis. Twenty-four months after remission from his renal metastasis, the patient presented with acute pain and weakness in his right upper extremity. However, he also endorsed worsening bilateral lower-extremity weakness that started 5 months before. He also had decreased sensation in bilateral lower extremities. In his right upper extremity he had Grade 2 of 5 deltoid strength, Grade 3 of 5 bicep strength, and Grade 4+ of 5 hand grip strength. Otherwise, he had Grade 5 of 5 strength in the right triceps and left upper extremity. There was no movement in bilateral lower extremities and no sensation below approximately the T-12 dermatome.

Sagittal T1-weighted contrast-enhanced MRI of brain and spine demonstrated leptomeningeal enhancement and contrast-enhancing masses throughout brainstorm and upper cervical cord surfaces (Fig. 3 left). The largest mass was in the midcervical region, with marked cord compression (Fig. 3 right). The patient underwent urgent cervical decompression via C3–6 laminectomy and biopsy of the large cervical mass. The mass adhered widely over the pial surface of the spinal cord. Biopsy staining with H & E revealed features characteristic of MPNST, including high cellular density, fascicles of tissue weaving in different directions (Fig. 4A), and pleomorphic long, wavy nuclei with tapered ends (Fig. 4B), confirming metastatic MPNST. There were at least 5 mitoses per high-power field (Fig. 4C). Type IV collagen was present around individual and small clusters of cells (Fig. 4D). The biopsy was negative for S100 protein and glial fibrillary acidic protein, and the tumor did not overexpress p53. Histological features of specimens from first and second surgeries were similar.

Outcome. The patient remained at his preoperative neurological baseline after biopsy. However, 6 days later, he developed new-onset headaches and seizures. Noncontrast head CT revealed hydrocephalus, likely secondary to biopsy of the large cervical mass. The mass adhered widely over the pial surface of the spinal cord. Biopsy staining with H & E revealed features characteristic of MPNST, including high cellular density, fascicles of tissue weaving in different directions (Fig. 4A), and pleomorphic long, wavy nuclei with tapered ends (Fig. 4B), confirming metastatic MPNST. There were at least 5 mitoses per high-power field (Fig. 4C). Type IV collagen was present around individual and small clusters of cells (Fig. 4D). The biopsy was negative for S100 protein and glial fibrillary acidic protein, and the tumor did not overexpress p53. Histological features of specimens from first and second surgeries were similar.

Discussion

Most MPNSTs arise de novo from normal peripheral nerves, perineuromas, and neurofibromas.2,9,15 Thus, MPNSTs may arise from any site, but they are most commonly located in the extremities, followed by the trunk, and less commonly the head and neck region.21 Primary intradural MPNST involving the spine is extremely rare, with only 20 adult cases reported in the literature: 7 cervical,2,9,15,20 6 thoracic,3,9,15,20 3 lumbar,2,25 and 4 cauda equina.2,21,28 (Table 1). Here we present the fifth case of an intradural MPNST of the cauda equina; what makes this case unique compared with the other case reports of intradural MPNST is that our patient developed diffuse leptomeningeal and CNS metastases causing mass effect on the brainstorm and spinal cord. In addition, long-term follow-up is provided.

It is well recognized that a history of RT increases the risk of MPNST.2,7,8,30 Our patient underwent adjuvant RT for testicular seminoma 10 years before presenting with a sacral mass. Interestingly, there are 4 reported cases of patients developing RT-induced MPNST after treatment for testicular seminomas. The oncological histories of these patients are very similar to that of our patient, each with an MPNST diagnosis made 7–10 years after adjuvant RT.5,18,33 Three of the 4 patients underwent RT to the ipsilateral paraaortic and/or inguinal regions and subsequently developed MPNST involving the sacrum: L3–S1,5 S-2 nerve root and plexus,18 and S-2 nerve root.33 Studies have shown that RT for testicular cancer confers increased risk of developing secondary malignancies, with one study showing a 2.6-fold increase.30 As a result, National Comprehensive Cancer Network guidelines updated in 2009 recommend surveillance as the preferred option for management of Stage I seminoma patients,22 and European Association of Urology guidelines revised in 2011 recommend against RT for localized testicular seminoma.4 The alternative to RT is single-agent carboplatin for Stage I seminoma,20 which may not be associated with secondary cancers.37 Although RT-induced MPNSTs are extremely rare, suspicion is warranted, as there is a real risk of developing secondary cancers a decade after RT.

The diagnosis of spinal intradural MPNST depends on high clinical suspicion on both imaging and histopathology analyses,6,11 especially since patients present with nonspecific signs and symptoms of pain, paresthesia, weakness, and/or incontinence, depending on the anatomical site. Imaging alone may lead to an incorrect diagnosis.19 In general, spinal MPNSTs are described as large tumors with irregularly lobulated borders that are indistinguishable from the surrounding soft tissue and bone, suggestive of malignant properties.19 On MR images, MPNSTs are inhomogeneously contrast enhancing and have scattered areas of T2-weighted hyperintensities,
likely representing areas of necrosis. There is also evidence of abundant blood supply and edema.

Histologically, MPNST can resemble other tumors (fibrosarcoma, leiomyosarcoma, and cellular schwannomas). Typically, MPNST cells weave in different directions, and long, wavy nuclei have tapered ends. Fascicle formation, mitoses, necrosis, and extreme nuclear anaplasia are also common. In addition, a wide variety of “divergent” histological patterns may be present: epithelial structures, rhabdomyoblastic differentiation, cartilage, and even bone. On immunohistochemistry, MPNSTs are negative for glial fibrillary acidic protein and variably immunoreactive for S100 protein (40% lack S100 protein) and Type IV collagen. Cellular schwannomas, in contrast, are diffusely positive for S100 protein and Type IV collagen and have fewer mitoses than do MPNSTs. Leiomyosarcomas have desmin. Fibrosarcomas lack S-100 protein. Alterations in the p53- and Rb-dependent pathways are common in MPNSTs. Necrosis and positive S100 protein immunohistochemistry can be focal features and could be missed on some specimens.

Patients with MPNSTs are best treated by complete resection with wide margins, an independent factor associated with improved survival. However, resectability and extent of resection depend greatly on tumor location. The ability to obtain complete tumor resection ranges from 20% for spinal tumors to 95% for extremity tumors. Spinal tumors are often less amenable to wide resection, given their intimate proximity to the spinal cord, major vessels, and vital nerves. In this patient, gross-total resection via complete sacrectomy offered him the best survival benefit but resulted in loss of ambulatory ability and...
TABLE 1: Cases of primary intradural MPNST reported in the literature*

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>Age (yrs), Sex</th>
<th>NF History</th>
<th>Previous RT</th>
<th>Presentation</th>
<th>Location</th>
<th>Surgery (biopsy or partial or complete resection)</th>
<th>RT</th>
<th>Chemotherapy Treatment</th>
<th>Recurrence</th>
<th>Metastasis</th>
<th>Survival</th>
</tr>
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<tbody>
<tr>
<td>Acharya et al., 2001</td>
<td>32, M</td>
<td>not mentioned</td>
<td>partial</td>
<td>back pain, leg weakness, bowel &amp; bladder dysfunction</td>
<td>cauda equina</td>
<td>yes</td>
<td>no</td>
<td>no</td>
<td>no</td>
<td>alive at 18 mos</td>
<td></td>
</tr>
<tr>
<td>Adamson et al., 2004</td>
<td>37, M</td>
<td>6 yrs previously for nodular sclerosing Hodgkin lymphoma</td>
<td>partial</td>
<td>cervical</td>
<td>partial</td>
<td>yes</td>
<td>no</td>
<td>no</td>
<td>no</td>
<td>dead after 1 yr &amp; a few mos</td>
<td></td>
</tr>
<tr>
<td></td>
<td>30, F</td>
<td>5 yrs previously for nodular sclerosing Hodgkin lymphoma, &amp; again for recurrent lymphoma</td>
<td>partial</td>
<td>cervical</td>
<td>partial</td>
<td>no</td>
<td>no</td>
<td>no</td>
<td>no</td>
<td>dead after 1 yr</td>
<td></td>
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<tr>
<td>Albayrak et al., 2006</td>
<td>25, M</td>
<td>NF1</td>
<td>not mentioned</td>
<td>paraparesis &amp; urinary incontinence</td>
<td>upper thoracic</td>
<td>complete</td>
<td>no</td>
<td>no</td>
<td>yes, in 7 wks</td>
<td>thoracic spine, lung</td>
<td>alive at 7 wks</td>
</tr>
<tr>
<td>Amin et al., 2004</td>
<td>38, M</td>
<td>10 yrs previously for testicular seminoma; paraaortic strip (total 30 Gy/15 fractions over 3 wks)</td>
<td>partial</td>
<td>back pain, leg weakness, bowel &amp; bladder dysfunction</td>
<td>cauda equina</td>
<td>biopsy</td>
<td>no</td>
<td>yes, palliative</td>
<td>not mentioned</td>
<td>not mentioned</td>
<td>not mentioned</td>
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<tr>
<td>Celli et al., 1995</td>
<td>52, F</td>
<td>not mentioned</td>
<td>complete</td>
<td>pain for 8 mos, weakness</td>
<td>thoracic</td>
<td>no</td>
<td>no</td>
<td>no</td>
<td>no</td>
<td>alive &amp; disease free at 6 yrs</td>
<td></td>
</tr>
<tr>
<td></td>
<td>68, F</td>
<td>not mentioned</td>
<td>complete</td>
<td>pain for 9 mos, weakness</td>
<td>lumbar</td>
<td>no</td>
<td>no</td>
<td>no</td>
<td>no</td>
<td>alive &amp; disease free at 2 yrs</td>
<td></td>
</tr>
<tr>
<td></td>
<td>43, M</td>
<td>not mentioned</td>
<td>complete</td>
<td>pain for 3 mos</td>
<td>lumbar</td>
<td>no</td>
<td>no</td>
<td>no</td>
<td>no</td>
<td>alive &amp; disease free at 6 yrs</td>
<td></td>
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<tr>
<td></td>
<td>36, F</td>
<td>not mentioned</td>
<td>complete</td>
<td>pain for 5 mos</td>
<td>thoracic</td>
<td>yes</td>
<td>no</td>
<td>yes</td>
<td>no</td>
<td>alive &amp; disease free at 4 yrs</td>
<td></td>
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<tr>
<td></td>
<td>22, F</td>
<td>NF1</td>
<td>not mentioned</td>
<td>pain for 2 yrs, weakness &amp; incontinence</td>
<td>cervical</td>
<td>complete</td>
<td>yes</td>
<td>no</td>
<td>yes</td>
<td>lung</td>
<td>dead at 6 mos</td>
</tr>
<tr>
<td></td>
<td>30, M</td>
<td>not mentioned</td>
<td>complete</td>
<td>pain for 6 yrs, weakness</td>
<td>thoracic</td>
<td>yes</td>
<td>no</td>
<td>yes</td>
<td>lung</td>
<td>dead at 14 mos</td>
<td></td>
</tr>
</tbody>
</table>

(continued)
### TABLE 1: Cases of primary intradural MPNST reported in the literature* (continued)

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>Age (yrs), Sex</th>
<th>NF History</th>
<th>Previous RT</th>
<th>Presentation</th>
<th>Location</th>
<th>Surgery (biopsy or partial or complete resection)</th>
<th>RT</th>
<th>Chemotherapy Treatment</th>
<th>Recurrence</th>
<th>Metastasis</th>
<th>Survival</th>
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</thead>
<tbody>
<tr>
<td>Mitsuhara et al., 2013</td>
<td>47, F</td>
<td>NF2</td>
<td>15 yrs previously for Stage Ib uterine cervical cancer (total 22 Gy adjuvant intraop paraaortic RT)</td>
<td>back pain, paraparesis of rt leg, urinary/bowel incontinence, acute consciousness disturbance</td>
<td>cauda equina</td>
<td>partial yes, adjuvant, total 36 Gy entire spine &amp; brain, additional 14.4 Gy to lumbo-sacral lesion/28 fractions</td>
<td>no</td>
<td>not mentioned</td>
<td>no</td>
<td>not mentioned</td>
<td></td>
</tr>
<tr>
<td>Seppälä et al., 1993</td>
<td>35, M</td>
<td>NF2</td>
<td>not mentioned</td>
<td>not mentioned</td>
<td>lumbar</td>
<td>complete no mentioned</td>
<td>not mentioned</td>
<td>yes</td>
<td>systemic</td>
<td>dead at 18 mos</td>
<td></td>
</tr>
<tr>
<td></td>
<td>23, F</td>
<td>not mentioned</td>
<td>not mentioned</td>
<td>not mentioned</td>
<td>upper thoracic</td>
<td>complete no mentioned</td>
<td>no</td>
<td>yes</td>
<td>systemic</td>
<td>dead at 8 mos</td>
<td></td>
</tr>
<tr>
<td></td>
<td>37, F</td>
<td>not mentioned</td>
<td>not mentioned</td>
<td>not mentioned</td>
<td>lower cervical</td>
<td>complete no mentioned</td>
<td>yes</td>
<td>after 2 yrs</td>
<td>systemic</td>
<td>dead at 6 yrs</td>
<td></td>
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<tr>
<td>Thomeer et al., 1981</td>
<td>42, M</td>
<td>not mentioned</td>
<td>back pain for 9 yrs worsening for 4 mos, impotence for 4 wks, leg weakness for 3 wks</td>
<td>cauda equina</td>
<td>complete yes, adjuvant, T11–S4 2.5 Gy 4x/wk for 6 wks (total 60 Gy)</td>
<td>yes, at time of recurrence; Adriamycin 120 mg every 3 wks (total 1080 mg)</td>
<td>yes, after 3 yrs, in T9–11</td>
<td>no</td>
<td>alive at 3 yrs w/ recurrence</td>
<td></td>
<td></td>
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<tr>
<td>Valdueza et al., 1991</td>
<td>43, F</td>
<td>not mentioned</td>
<td>low-back pain for 1 mo, leg weakness for 2 wks</td>
<td>lower thoracic</td>
<td>partial yes, total 24 Gy after 1st op, total 32 Gy after 2nd op at time of recurrence</td>
<td>no</td>
<td>yes, after 8 yrs</td>
<td>no</td>
<td>alive at 10 yrs w/ recurrence</td>
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<tr>
<td></td>
<td>47, M</td>
<td>NF1</td>
<td>not mentioned</td>
<td>neck pain radiating to rt shoulder for 9 mos, rt arm weakness for 1 mo</td>
<td>cervical</td>
<td>complete yes, total 10 Gy</td>
<td>no</td>
<td>yes, after 3 mos</td>
<td>lumbar spine, brain</td>
<td>dead at 18 mos</td>
<td></td>
</tr>
<tr>
<td></td>
<td>18, M</td>
<td>not mentioned</td>
<td>It shoulder &amp; arm pain for 4 mos</td>
<td>cervical</td>
<td>complete no</td>
<td>no</td>
<td>no</td>
<td>no</td>
<td>alive &amp; disease free at 8 mos</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>70, F</td>
<td>not mentioned</td>
<td>neck pain radiating to rt shoulder for 6 mos</td>
<td>cervical</td>
<td>complete no</td>
<td>no</td>
<td>no</td>
<td>no</td>
<td>alive &amp; disease free at 7 mos</td>
<td></td>
<td></td>
</tr>
<tr>
<td>present study</td>
<td>43, M</td>
<td>10 yrs previously, adjuvant therapy for testicular seminoma</td>
<td>severe back pain &amp; paresthesia radiating to lt buttock &amp; thigh for 5 mos</td>
<td>cauda equina</td>
<td>complete no</td>
<td>yes, neoadjuvant &amp; adjuvant therapy alternating b/wn ifosfamide/doxorubicin &amp; ifosfamide/etoposide</td>
<td>yes, about 60 mos</td>
<td>renal, cervical cord, &amp; brain-stem</td>
<td>died at 5 yrs from CNS metastasis</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

* NF = neurofibromatosis (NF1 = Type 1; NF2 = Type 2).
Cauda equina MPNST

bladder/bowel control. While aggressive resection is warranted, consideration of functionality and quality of life needs to be carefully balanced.

Postoperative adjuvant RT, chemotherapy, or both have yet to be systematically studied for the treatment of spinal MPNST, but there are indications that adjuvant RT may be beneficial.29 A large case series by Baehring et al.30 of 54 patients with MPNSTs in various locations showed a mortality hazard ratio of 0.22 for adjuvant RT on multivariate analysis. Adjuvant RT is particularly beneficial for local tumor control when radical resection is not possible, or for recurrent tumors.1 Chemotherapy has not proven to be effective in the treatment of MPNST2 but can be used in cases without alternatives for disseminated metastatic disease.13 The expanding scientific literature suggests that MPNST can be treated more effectively with the addition of agents that target and inhibit specific pathways and/or cellular processes: PI3K/mTOR pathway with XL765,12 autophagy with chloroquine,12 and platelet-derived growth factor receptor with sunitinib.35

The reported 5-year survival rate for patients diagnosed with MPNST is poor, ranging from 16% to 52%,10,18,34 Among reported cases of spinal intradural MPNST, the calculated 1-year, 3-year, and 5-year survival rates are 81%, 50%, and 25%, respectively. In a study of 21 patients with spinal MPNSTs, the local recurrence rate was extremely high: 71%2; this is approximately twice the rate associated with extraspinal MPNSTs (20%–40%),12-24,33 and is likely secondary to lower resectability. Of the 20 reported cases of patients with intradural MPNSTs of the spine, eventual metastasis was seen in 8 (40%), but these rates depend on the length of follow-up in each case report/series.

Intradural MPNSTs represent a unique clinical entity, as these tumors can be associated with greater morbidity and worse prognosis secondary to direct access for metastatic spread and seeding of the CNS. Our patient succumbed to diffuse leptomeningeal spread and seeding of the brainstem and cervical cord. This is an extremely rare event in patients with an intradural MPNST. Only 1 other patient in the literature demonstrated CNS spread of an intradural MPNST,29 which occurred a little more than 1 year after complete removal of the original tumor, with death shortly thereafter. There is only 1 other reported case of leptomeningeal spread of an MPNST,26 which is rapidly fatal. Additional surveillance for these potential sequelae should be carefully considered in patients with intradural MPNST.

Primary intradural MPNST of the spine is a rare tumor that may arise secondary to prior RT. It is difficult to diagnose and has a poor prognosis. Given that adjuvant RT is relatively common, RT-induced intradural MPNST of the spine may be more widespread than reported. Better awareness of this entity and its potential for metastasis to the CNS may enable physicians to provide early intervention in patients who have signs and symptoms associated with this condition.

Disclosure

Dr. Park reports being a consultant for Globus Medical and Medtronic and receiving royalties from Globus Medical.

Author contributions to the study and manuscript preparation include the following. Conception and design: Orringer, Hervey-Jumper. Acquisition of data: Lau, Moon, Hervey-Jumper, McKeever. Analysis and interpretation of data: Lau, Moon, Park, Hervey-Jumper, McKeever. Drafting the article: Lau, Moon. Critically revising the article: Orringer, Park, Hervey-Jumper, McKeever. Reviewed submitted version of manuscript: all authors. Study supervision: Orringer.

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