A 19-year-old, otherwise healthy, white man presented with a 6-month history of progressive right leg spasticity, right arm weakness, and left-sided lower extremity numbness. Two days prior to being seen by his physician, he had developed loss of bowel and bladder control. Magnetic resonance imaging showed an enhancing, dumbbell-shaped epidural and intradural mass at C6–7 with cord compression and extension into the right brachial plexus (Figure 1). A C5–7 laminectomy demonstrated a firm, whitish, focally hemorrhagic tumor with an intradural component compressing and displacing the spinal cord to the left. The intracanalicular portion of the tumor was completely resected; however, a dissection into the brachial plexus could not be performed. The nerve roots at C6 and C7 were involved by the tumor and were sacrificed. Postoperatively, the patient received radiation and chemotherapy. Six months after surgery, he is neurologically intact and has no evidence of tumor by magnetic resonance imaging.

Histologically, a focally necrotic malignant tumor growing in broad sheets infiltrated the peripheral nerve roots (Figure 2). The cells were large, uniform, and polygonal with vesicular nuclei, prominent nucleoli, and abundant cytoplasm (Figure 3). Numerous, slightly fibrillary to hyaline paranuclear inclusions were present; these inclusions were positive for cytokeratin. Immunostains for S100, desmin, CD30, HMB-45, and CD45 were negative. Electron microscopy demonstrated numerous paranuclear whorled arrays of closely packed intermediate filaments (Figure 4).

**What is your diagnosis?**
Pathologic Diagnosis: Extrarenal Rhabdoid Tumor

Malignant rhabdoid tumor was originally described in 1978 as a result of a review of a large number of Wilms tumors. It was histologically characterized by sheets of large polygonal cells with prominent nucleoli and cytoplasmic inclusions of hyaline material, which were subsequently shown to be frequently positive for cytokeratin and to correlate ultrastructurally with a whorled paranuclear array of intermediate filaments. Since then, morphologically similar tumors have been described in other organs and soft tissue locations. A histologic similarity between rhabdoid tumor and epithelioid sarcoma has also been noted in recent years, one result of which has been to suggest that the separation of these two tumors is clinical and not pathologic. Acknowledging that the present report cannot resolve this issue, it will be designated for convenience as an extrarenal rhabdoid tumor.

Of all the various locations in which extrarenal malignant rhabdoid tumors have occurred, one of the least common has been paravertebral soft tissue with involvement of the spinal cord. The Table lists the features of the present case and of 5 cases documented in the literature. Only those cases in which compatible histopathology was supported by immunostaining, electron microscopy, or both were included. As in most cases of extrarenal rhabdoid tumor, the patients were young. Despite attempts to extirpate the tumors, total removal is seldom successful, and in such cases postoperative radiation and chemotherapy have not been effective in arresting a lethal outcome. The only patient with a complete excision was disease-free at 12 months. Although surgical removal was incomplete in the present case, most of the grossly visible tumor was removed, and the patient is well and neurologically intact 6 months after surgery. Despite the small number of cases reviewed in the Table, it may be that the success of additional therapeutic modalities with this tumor was strongly influenced by the completeness of excision.

Much discussion in the literature has been concerned with the nonspecific morphologic features of extrarenal rhabdoid tumor, since various carcinomas, melanomas, or primitive sarcomas may exhibit the same features. In the clinical context of a malignant tumor arising in a soft tissue location in a young patient (in this case aged 19 years), an epithelial malignancy or melanoma would be extraordinary. The differential diagnosis should center on large round-cell sarcomas, one of which is extrarenal rhabdoid tumor, or hematologic malignancies. Rhabdoid tumor is both a defined tumor entity and a well-established term that connotes a particular morphology associated with aggressive clinical behavior. Given the rarity of the lesion, the potential for efficacious therapy and reliable prognosis are uncertain.

References


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**Primary Paravertebral Malignant Rhabdoid Tumor: Review of Literature**

<table>
<thead>
<tr>
<th>Reference No.</th>
<th>Age/Sex</th>
<th>Symptom</th>
<th>Location</th>
<th>Treatment</th>
<th>Adjuvant Treatment*</th>
<th>Postoperative Follow-up</th>
</tr>
</thead>
<tbody>
<tr>
<td>4</td>
<td>59 y/M</td>
<td>Weight loss, anoxia</td>
<td>T6, 8 cm</td>
<td>Decompressive laminectomy</td>
<td>No</td>
<td>Died at 6 mo; progressive paraplegia; malignant pleural effusion</td>
</tr>
<tr>
<td>5</td>
<td>11 mo/F</td>
<td>Weight loss</td>
<td>L5-S1, not measured</td>
<td>Partial excision</td>
<td>C and R</td>
<td>Died at 16 mo; no autopsy</td>
</tr>
<tr>
<td>6</td>
<td>3 mo (sibling)/F</td>
<td>Mass</td>
<td>Right paraspinal not measured</td>
<td>Biopsy only</td>
<td>C and R</td>
<td>Died at 6 mo; no autopsy</td>
</tr>
<tr>
<td>7</td>
<td>15 y/M</td>
<td>Cord compression</td>
<td>T6-9, 7 cm</td>
<td>Complete excision</td>
<td>C</td>
<td>Disease-free at 12-mo follow-up</td>
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<td>Present case</td>
<td>2.5 y/F</td>
<td>Weight loss, weakness of right arm</td>
<td>C6-T1, not measured</td>
<td>Partial excision</td>
<td>C</td>
<td>Died at 8 wk; no autopsy</td>
</tr>
<tr>
<td></td>
<td>19/M</td>
<td>Brachial plexus palsy, cord compression</td>
<td>C6-7, 3 cm</td>
<td>Partial excision</td>
<td>C and R</td>
<td>Disease-free at 6 mo; neurologically intact</td>
</tr>
</tbody>
</table>

* C indicates chemotherapy; R, radiation.