Peripheral nerve tumors are soft-tissue tumors that can occur in any nerve throughout the body. The majority of peripheral nerve tumors arise from elements of the nerve sheath with the two most common being neurofibromas and schwannomas. More than 90% of all peripheral nerve tumors are benign. When there is peripheral nerve involvement in metastatic carcinoma, it is often via contiguous spread from the primary mass; hematogenous seeding to a peripheral nerve is seldom seen. In this report the authors describe the even rarer case of metastatic renal cell carcinoma mimicking a schwannoma in a dorsal root ganglion. Cases from the literature show the rarity of this finding and its late clinical appearance. Given that survival in patients with metastatic carcinoma continues to increase, dorsal root ganglion metastasis may become more common over time.

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KEY WORDS renal cell carcinoma; metastasis; dorsal root ganglion; schwannoma; dumbbell lesion; oncology
eral nerve or spinal cord symptoms were present; motor strength was 5/5 throughout, reflexes were normal and symmetrical, toes were downgoing bilaterally, sensation was intact, and gait was normal.

**Operation**

Given that the lesion was growing, the decision was made to proceed with surgical biopsy for diagnosis. Intraoperatively, the proximal nerve was visualized exiting the foramen, and there did not appear to be a tissue plane between the tumor mass and the nerve. Distal to the tumor mass the nerve appeared to branch into multiple fascicles; the resection margin was distal to these fascicles. The tumor specimen was sent for pathological analysis. Intraoperative blood loss was 800 ml, with most of the blood loss coming from epidural veins; the tumor did not bleed significantly.

**Pathological Findings**

Microscopy of the tumor revealed a mass with an attached spinal nerve, encapsulated by part of the normal DRG structures (Fig. 2A). While normal DRG cells were found at the periphery (Fig. 2C, arrow), the remainder of the ganglion had been replaced by large fields of tumor cells with optically empty cytoplasm and round to oval, fairly regular, nuclei. Many thin-walled capillaries were also visualized within the tumor. The tumor extended to the proximal surgical margin.

Immunohistochemical analysis showed that both the tumor and the adjacent nerve were positive for the mesenchymal marker vimentin (not shown), while only the adjacent nerve was positive for the Schwann cell marker S100 (Fig. 2B). The tumor cells were positive for PAX-8 (Fig. 2D), the RCC marker (Fig. 2E), and CD10 (not shown). Because most cases of RCC are associated with a deletion of the *von Hippel Lindau* gene on the short arm of chromosome 3, fluorescence in situ hybridization (FISH) was performed which confirmed the loss of the 3p25 region in all tumor cells. Taken together, these findings are consistent with an RCC metastasis.

At the 3-month follow-up, the patient was free of motor, sensory, or autonomic spinal cord or peripheral nerve symptoms. Neither radiotherapy nor chemotherapy was given at this point. Further resection was offered to the patient; however, because she had other metastatic lesions that were progressing and because she was asymptomatic with respect to the thoracic lesion, the patient refused further treatment. Five months postsurgery the patient was started on sunitinib malate to control growth of metastatic disease elsewhere. Currently, 1 year after surgery, there is no recurrence of the DRG lesion, and the patient has no peripheral nerve or spinal cord symptoms.

**Discussion**

Most tumors involving the spinal nerve roots and DRG are primary neoplasms that arise from components of the neural sheath, schwannomas being the most common.
Schwannomas arise from neoplastic Schwann cells that proliferate and displace the normal components of the nerve to the periphery. Normally, Schwann cells are responsible for myelinating axons in peripheral nerves and are analogous to oligodendrocytes in the central nervous system. Neurofibromas are another common type of primary peripheral nerve tumor, which arise from the proliferation of several cell types normally present in the neural sheath: Schwann cells, perineuronal cells, and fibroblasts.

The central nervous system is frequently affected by metastatic tumor. The peripheral nervous system is also often affected by tumor elsewhere, by perineural and neural growth directly in or around a tumor—factors that are often used in grading tumors and are considered a poor prognostic sign. Hematogenous spread to peripheral nerves is rare, and despite the frequency of metastatic cancer in general, metastatic cancer to the DRG is even rarer. To date, only 7 reports of a total of 9 cases have been reported in the literature (Table 1). The first part of this series comprises 3 autopsy reports documenting 1654 autopsies, in which a total of 5 cases of DRG metastasis were found.1,3,13 For those studies with details, the lesions were documented in 58 routine autopsy cases from different institutions, revealing a metastatic lesion with positive symptoms, or the symptoms such as radicular pain are attributed to other aspects of the disease, and that therefore the true frequency of DRG metastases is higher.

Management in our case was guided initially by radiological findings that led to a presumptive diagnosis of schwannoma. Indeed, many of the radiological features of the tumor identified in our patient were characteristic for an extramedullary intradural schwannoma. Specifically, the extra- and intraforaminal location and dumbbell shape of the tumor were consistent with a slow-growing, benign mass that extended through the intervertebral foramen with some associated remodeling.7 Moreover, the tumor in our case was consistent with a small schwannoma in that it showed homogeneous enhancement after gadolium.8 However, the central hypointensity on sagittal T1-weighted images differed from the previously described schwannoma “target sign” and may have represented a flow void caused by the highly vascular tumor.2 Conversely, the tumor was also consistent with RCC, which tends to be hypo- to isointense on T1-weighted images, iso- to hyperintense on T2-weighted images, and avidly enhancing.9 In hindsight, the radiological features in our case corresponded well with the histological appearance of the tumor: a vascular neoplasm, centrally infiltrating the DRG, and displacing the normal parenchyma to the periphery.

For most patients with spinal nerve tumors, surgery is indicated for symptom relief. Patients often present with symptoms of a radiculopathy, including weakness, paresthesia, and pain.2 Even though she was asymptomatic, our patient was treated surgically to obtain a diagnosis because the tumor was increasing in size and was starting to compress the spinal cord. The procedure was uneventful. Subsequent pathological analysis of permanent sections, however, revealed a metastatic lesion with positive margins.

Given the ever-increasing survival in cancer patients and the fact that DRGs are sites that, from a clinical point of view, are involved late in metastasis, it may well be that the frequency of DRG metastasis will increase in the future. This report therefore highlights the need to always include metastasis in the differential diagnosis for a new lesion in a patient with a known primary cancer, even when the lesion has the typical presentation of another common disease, or if survival after initial disease is long.

### TABLE 1. Review of cases in the literature

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>Study</th>
<th>Primary Organ</th>
<th>Primary Tumor</th>
<th>Interval (yrs)*</th>
<th>Symptoms</th>
</tr>
</thead>
<tbody>
<tr>
<td>Chason et al., 1963</td>
<td>1096 autopsy cases</td>
<td>Unknown</td>
<td>Unknown</td>
<td>Unknown</td>
<td>Unknown</td>
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<tr>
<td>Van Ketel, 1979</td>
<td>58 routine autopsy cases</td>
<td>Uterus</td>
<td>Cervical carcinoma</td>
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<td>Unknown</td>
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<tr>
<td>Johnson, 1977</td>
<td>500 routine autopsies</td>
<td>Lung</td>
<td>Small cell</td>
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<tr>
<td></td>
<td></td>
<td>Colon</td>
<td>Adenocarcinoma</td>
<td>1</td>
<td>No</td>
</tr>
<tr>
<td>Wigfield et al., 2003</td>
<td>Case report</td>
<td>Colon</td>
<td>Adenocarcinoma</td>
<td>12</td>
<td>Yes</td>
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<td>Uchida et al., 2008</td>
<td>Case report</td>
<td>Uterus</td>
<td>Adenocarcinoma</td>
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<tr>
<td>Schultz et al., 2009</td>
<td>Case report</td>
<td>Breast</td>
<td>Ductal carcinoma</td>
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<td>Shakeel et al., 2009</td>
<td>Case report</td>
<td>Kidney</td>
<td>RCC</td>
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<td>Yes</td>
</tr>
<tr>
<td>Current case</td>
<td>Case report</td>
<td>Kidney</td>
<td>RCC</td>
<td>6.5</td>
<td>No</td>
</tr>
</tbody>
</table>

* In patients without symptoms, interval refers to the time between primary tumor diagnosis and autopsy or surgery. In patients with symptoms, interval refers to the time between primary diagnosis and symptom onset.
References


Author Contributions
Conception and design: Jansen, Wasserman. Acquisition of data: Wasserman, Tsai, Glikstein, Mai. Analysis and interpretation of data: Jansen, Wasserman, Tsai, Glikstein. Drafting the article: Jansen, Wasserman, Tsai, Glikstein. Reviewed submitted version of manuscript: Wasserman, Tsai, Glikstein, Mai. Study supervision: Jansen.

Correspondence
Gerard H. Jansen, Eastern Ontario Regional Laboratory, Civic Campus—Lab Med Building Rm. 121, 1053 Carling Ave., Ottawa, ON K1Y 4E9, Canada. email: gjansen@toh.on.ca.