Metastatic intradural primary spinal osteosarcoma: illustrative case

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BACKGROUND Osteosarcomas are a common primary bone neoplasm among adolescents but represent 0.2% of all malignancies with an incidence of two to four cases per million persons annually worldwide. Although known to have significant metastatic potential, its rare incidence, treatment resistance, and poor prognosis have rendered it a poorly understood and infrequently documented pathology.

OBSERVATIONS Herein the authors present the first documented case of lumbosacral intradural metastasis of a primary osteosarcoma in a young patient, possibly via intradural dissemination following pinhole durotomy in a prior thoracic surgery.

LESSONS Osteosarcomas remain a difficult pathology to treat, particularly upon metastatic dissemination. The utility of adjuvant radiotherapy after resection of an osteosarcoma is increasingly evident in the reduction of local recurrence. In the context of intraoperative pinhole durotomies in resections of high-grade lesions, due consideration should be given to whole-spine radiation, although this remains an evidence-free zone.

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KEYWORDS intradural osteosarcoma; metastatic osteosarcoma; intradural dissemination

Osteosarcomas represent a primary malignancy of bone with a strong propensity for the metaphyses of long bones, particularly in the younger population.1 These aggressive tumors rarely occur in the axial skeleton and even more rarely metastasize to extraosseous tissue.1–3 Only three cases of intradural spinal osteosarcomas have been described in the literature, with the following case the first described instance of intradural dissemination of an osteosarcoma, likely via the spinal subarachnoid space.2

Illustrative Case

History
Referred to our neurosurgical department by her medical oncologist, a 28-year-old female had a 2-week history of progressive right lower-limb weakness, several episodes of fecal incontinence, urinary hesitancy, and sensory disturbance in the L2 dermatome. A neurological examination revealed normal cranial nerve and upper-limb function. Her lower-limb examination revealed normal tone bilaterally, with normal patellar and ankle jerk reflexes. Her Babinski plantar reflexes were negative bilaterally. Her left lower limb had preserved 5/5 power throughout all myotomes. Right-sided hip flexion was 2/5 and hip extension 3/5, while knee flexion and extension were 3/5. Distal power of dorsiflexion, plantarflexion, and hallucis extension were 2/5. Light touch sensation was grossly preserved on the left and diminished in the L3–S1 dermatomes on the right. Her gait was reflective of dense monoparesis of the right lower limb.

A preliminary computed tomography study revealed a large, heterogeneous mass at the level of L2–3 with high attenuation in keeping with significant calcification. A magnetic resonance imaging (MRI) study of the patient’s whole spine revealed the mass was intradural, causing severe canal stenosis, cauda equina compression, and foraminal compression of the exiting right L2 nerve root (Fig. 1). There was progression of a previously identified T9 paravertebral mass and new vertebral body metastases to C4, L4, and S2.

Oncological History
An osteosarcoma of the left proximal humerus had been diagnosed several years earlier, and wide resection of the primary lesion had been performed. The formal histopathological diagnosis was mixed parosteal and dedifferentiated high-grade osteosarcoma. The patient was subsequently treated with adjuvant chemotherapy using methotrexate, doxorubicin, and cisplatin.

ABBREVIATIONS MRI = magnetic resonance imaging.

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Months later, the patient presented with mild hypoesthesia below the level of the umbilicus and was found to have a distant metastasis within the T10 vertebral body resulting in mild cord compression. A posterior T10 en bloc spondylectomy was performed, followed by a T8–12 pedicle screw fixation. During the procedure, the dura was found to be intimately adherent to the diseased T10 vertebral body, and a pinhole durotomy occurred. Postoperatively, the patient was neurologically intact, and her sensory deficits resolved. The histopathological diagnosis was consistent with the previous diagnosis, and she subsequently received high-dose palliative radiotherapy (50.4 Gy in 28 fractions) to the operative site.

After completion of her radiotherapy, the patient was restaged and found to have additional metastatic deposits within the left acetabulum and adjacent ischium. These were treated with further radiation (39 Gy in 13 fractions), and she was then started on cabozantinib. Further imaging revealed disease progression with metastases of the left scapula and iliac crest, prompting six cycles of ifosfamide and etoposide, which were completed approximately 2 months prior to her latest presentation.

Operation
The patient underwent an L2–3 laminectomy, with a midline durotomy and subsequent piecemeal debulking of the intradural tumor. Intraoperatively, the tumor was significantly heterogeneous: calcified areas resembling cortical bone, interspersed with soft components and pockets of subacute hematoma. Some nerve roots traversing the tumor were deemed nonsalvageable and were transected. Near-total resection was achieved with small fragments of tumor in-separable from nerve roots left in situ.

Postoperative Course
The patient’s postoperative recovery was unremarkable. The histopathological diagnosis was unchanged. After surgery, prior to discharge to a rehabilitation ward, the patient’s neurological examination confirmed normal revealed nil sphincter function. Her sensory losses were unchanged apart from complete anesthesia of the right thigh. Her left lower-limb function was intact with her right lower-limb examination revealing persistent weakness of hip flexion and extension (1/5), knee flexion and extension (3/5), and dorsiflexion (1/5) but resolution of plantarflexion (5/5). In the weeks after surgery, she was found to have improved proximal power of right hip flexion and extension (2/5). The patient required assistance to mobilize with a Zimmer frame. At the time of composition, 30 Gy of radiation over 10 fractions to the L2 lesion was planned, and the patient was started on last-line chemotherapy in the form of docetaxel and gemcitabine.

Patient Informed Consent
The necessary patient informed consent was obtained in this study.

Discussion
Osteosarcomas remain the most common malignant primary neoplasm of bone among children and young adults, although they are a rare entity constituting 0.2% of all malignancies. They are characterized by immature osteoid matrices laid by mesenchymal or osteogenic progenitor cells, with an estimated annual worldwide incidence of two to four cases per million persons. They have a bimodal distribution, with a peak predominant incidence among children ages 13 to 16, coinciding with growth spurts, and a small peak above the age of 60. A significant predilection to metastasize confers high rates of treatment failure and poor prognosis among osteosarcoma patients. A unique series of molecular alterations culminate in distinct biological behavior of metastatic colonies in comparison to the index tumor, allowing metastases to virtually any site and organ, with a proclivity for the lung, appendicular or axial bone, and lymph nodes. The most common sites of primary osteosarcoma in younger patients are the metaphyses of long bones, with 10% of cases within the proximal humerus. Primary spinal osteosarcomas are a rare entity, making up less than 3% of all osteosarcomas, whereas primary extraosseous osteosarcomas are obscure presentations.

In the literature, there have been five described cases of primary intradural extramedullary osteosarcomas, two of which were cervical lesions in dogs. Curiously, all three human cases involved the lumbosacral region (Table 1). The first two reported patients were both elderly women, sharing a previous history of iofendylate myelography. A provisional hypothesis postulated the consequent adhesive
arachnoiditis and subsequent metaplasia as the potential etiology for the development of intradural osteosarcoma after the use of the oil-based radiocontrast agent. Other possible correlations include malignant dedifferentiation of meningiomas after radiotherapy but have not been clearly implicated in intradural metastasis of osteosarcoma.2,10

Mehra et al.2 published a case report in 2019, reporting a primary intradural osteoblastic osteosarcoma with the radiological absence of alternative sites of disease. This was the first reported incidence of intradural osteosarcoma in the absence of historical exposure to iofendylate or prior radiation therapy.

**Observations**

To our knowledge, this is the fourth reported case of intradural osteosarcoma. However, it is the first reported case of metastatic primary intradural osteosarcoma, although the pinhole durotomy during the patient’s en bloc corpectomy might have caused intradural dissemination. We hypothesize that the fluid constitution of the spinal subarachnoid space may have facilitated its lodgement within the lumbosacral region, akin to “drop” metastases in other neuroaxial malignancies, such as ependymomas and choroid plexus papillomas.11 This is more likely than the alternative probability of hematogenous spread into an intradurally discrete mass free of surrounding osseous disease. Regardless, the intradural excursion of metastatic disease is consistent with the well-documented ability of osteosarcoma to disseminate to unusual sites, both local and distant.1

**Lessons**

Historically, the ascribed radioresistance of osteosarcomas had made radiation treatment unpopular in their management. Although previously reserved for incompletely resected tumors or palliative management of symptomatic metastases in nonoperative cases, more recent applications have yielded a favorable response to radiotherapy as a surgical adjunct.12,13 In our patient, there was no local recurrence at the treated T10 level on radiological surveillance. However, the patient did develop multifocal metastatic disease in the spine elsewhere, both intradural and osseous.

In other solid tumors with leptomeningeal involvement, neuroaxis radiation is often considered.14 In retrospect, in the context of intradural dissemination possibly attributable to an intraoperative durotomy, could prophylactic whole-spine radiation have prevented or delayed intradural spread? Although associated with significant toxicity, including myelosuppression and enteritis, perhaps whole-spine radiation postoperatively is worth considering in inadvertent durotomy during resection of high-grade lesions such as osteosarcomas.14 Given the obscurity of intradural osteosarcoma metastases, there is a paucity of literature on this area of contention, which remains an active field of ongoing research.

**References**


**TABLE 1. Summary of reported cases of intradural osteosarcoma**

<table>
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<tr>
<th>Feature</th>
<th>Pal et al., 20089</th>
<th>Schiller et al., 20137</th>
<th>Mehra et al., 20192</th>
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<td>Metastasis</td>
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<td>L2 to L3, previously T10</td>
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<td>Iophendylate myelogram (20 yrs prior)</td>
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Authors & Year
Disclosures
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
Conception and design: Thiruvengadam, Honeybul. Acquisition of data: Thiruvengadam, Lam. Analysis and interpretation of data: Thiruvengadam, Lam. Drafting the article: Thiruvengadam. Critically revising the article: all authors. Reviewed submitted version of manuscript: Thiruvengadam. Approved the final version of the manuscript on behalf of all authors: Thiruvengadam. Study supervision: Lam, Honeybul.

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