Intramedullary Spinal Cord Metastasis from Endometrioid Adenocarcinoma

Alireza Khoshnevisan MD MPH¹, Sina Abdollahzade MD MPH², Isa Jahan zad MD³, Anoshirwan Niknezhad MD², Soheil Naderi MD³, Masoud Khadivi MD³

Abstract
Intramedullary spinal cord metastases are rare. The majority of these metastases reportedly spread from lung cancer in the cervical region; however, they have been seen to arise from a variety of other primary sources. Here, we report what is, to the best of our knowledge, the first known case of an intramedullary spinal cord metastatic lesion in the conus region arising from primary endometrioid adenocarcinoma.

Keywords: Endometrioid carcinoma, intramedullary tumor, metastases, spinal cord


Introduction
Intramedullary spinal cord metastasis (ISCM) is a rare site for systemic cancer invasion. It accounts for about 1–3% of all intramedullary spinal cord neoplasms and 2% of central nervous system metastases. Although lung and breast cancers are the most common primary tumor sites in ISCM, lymphoma, renal cell carcinoma, colorectal carcinoma, and unknown primary tumors were also reported. The prognosis of patients with ISCM is very poor and many have neurological deficits and pain. Radiotherapy, chemotherapy and surgery can be effective in selected cases.

Case Report
A 61-year-old woman presented with a two-month history of low back pain radiating to both legs. Since 2 weeks before presentation, her pain aggravated and she progressively developed lower extremity weakness as well as sensory loss and urinary incontinence. General physical exam was unremarkable except for pale conjunctiva. On neurologic exam, motor scores were reduced in both proximal and distal sites of lower extremities and she could not walk on her own. Partial sensory loss was also noted along with saddle hypothermia. Deep tendon reflexes were diminished and plantar reflexes were downward bilaterally. The patient’s history was significant for endometrial carcinoma, endometrioid adenocarcinoma grade 1B. She had undergone total abdominal hysterectomy and bilateral salpingo-oophorectomy about 15 months before admission. No metastasis was found at that time.

A magnetic resonance imaging scan of whole neuroaxis was performed, revealing an enhancing intra-axial mass lesion at T12-L1 level with peritumoral edema (Figure 1). Whole body bone scan and computed tomography (CT) scan of the chest, abdomen, and pelvis were negative for other lesions. An investigation for other possible sources of infection or tumor was negative. As intramedullary target of endometrioid adenocarcinoma has not been encountered in the literature, she was scheduled for surgical resection of the lesion with an impression of primary conus pathology. The patient underwent T12-L1 laminectomy and partial tumor removal. The tumor appeared as an ill-defined grayish mass without clear margins from surrounding cord. Histopathologic examination of sample demonstrated glandular composition of neoplastic cells consistent with metastasis from endometrioid adenocarcinoma (Figure 2). Immunohistochemistry staining also provided more evidence to support this. IHC was positive for Pan-Cytokeratin (CK), CK7, Vimentin, Carcinoembryonic Antigen (CEA) and Progesterone receptors and negative for Thyroid Transcription Factor 1, CK20, Synaptophysin, Wilm’s tumor 1 protein (WT1) and Estrogen receptors (Figure 2). Postoperatively, the patient experienced moderate improvement in her pain while her limb force or continence did not change. She was assigned for radiotherapy following discharge. The radiotherapy protocol consisted of 20 Gray dose in 5 fractions. Three months post-op following the completion of radiotherapy, a control image was performed (Figure 3). Although there was slight reduction in tumor size, concomitant syrinx shrank dramatically (Figure 4). The patient was then assigned for chemotherapy which was prematurely ceased because of medical complications. She has survived beyond 18 months (May 2013) since the detection of metastasis without any change in neurological status.

Discussion
Intramedullary spinal cord metastases (ISCM) are considered to be quite rare; they account for 1 to 3% of all intramedullary spinal cord lesions. The majority of intramedullary metastases are from lung and breast cancer; however, other tumors have also been cited in the literature.
Figure 1. MR imaging of thoracolumbar spine depicting intramedullary spinal cord lesion (A) Sagittal T1-weighted MRI without contrast showing hyposignal area at T12-L1 level of spinal cord (B) Sagittal post Gadolinium T1-weighted image revealing peripheral avid enhancement of the lesion. (C) Sagittal T2-weighted MRI revealing central hyposignal area of the lesion with associated cord edema and syringomyelia (D) Axial post Gadolinium T1-weighted image confirms intramedullary central and posterior location of tumor.

Figure 2. Histopathology slide of tumor (A) Spinal cord tissue infiltrated with neoplastic tissue composed of glandular structures, lined by cylindrical atypical cells with hypochromatic nuclei and high N/C ratio. Necrosis is also evident. Hematoxylin and eosin stain, magnification ×100. (B) Enlarged view, better illustrating glandular structures magnification ×400 (C) IHC staining for Cytokeratin 7.
Metastasis to the intramedullary spinal cord rarely occurs with systemic cancer. Autopsy series have reported its prevalence in cancer patients to range from 0.9% to 2.1%. Lung cancer is the most common source of ISCM, comprising approximately 50% of reported cases. Breast carcinoma is the second observed source, accounting for 8% to 14% of ISCM. Other primary sources for ISCM include renal cell carcinoma, melanoma, colorectal carcinoma, and lymphoma.

To our best knowledge, there is no reported case in the English literature for endometrial adenocarcinoma metastasis in intramedullary spinal cord. Also, few cases of ISCM have been reported in the conus medullaris region. In a series of 138 cases of intramedullary spinal cord metastasis of any origin, none targeted conus medullaris. Hence, our case characterizes a unique picture of primary tumor involvement as well as the affected site of spinal cord and clinical presentation.

Various routes of tumor spread to spinal cord have been described: hematogeneous seeding, Batson’s venous plexus, leptomeningeal seeding via cerebrospinal fluid and direct extension through dura mater or nerve roots. Given the proximity of primary tumor, the most probable route for cancer dissemination in our patient is regional venous extension; however, hematogeneous seeding could not be ruled out.

Our patient presented with progressive paraparesis and sphincteric disturbance, a finding that often portends an ISCM in cancer patients. Most reported cases of ISCM also had focal neurologic deficits on initial encounter contrary to vertebral metastasis which mainly manifests with pain.

The prognosis of intramedullary metastasis has been repeatedly reported as extremely poor. Several studies found a median survival of less than 12 months. No consensus outlined management of these lesions and therapeutic decisions are made on case by case basis. Fractionated radiotherapy is advocated especially in radiosensitive primary tumors. Surgery is an option when rapid neurological deterioration is encountered or when histopathology of the lesion is unclear, as in our case. Patients with poor general health, severe neurologic deficits and multiple metastases are poor candidates for surgery. An improved survival has been confirmed in patients undergoing surgical tumor removal. However, the pros and cons of surgery must be carefully weighed in discussions with individual patients and their families.

Here, we presented the first case of intramedullary endometrial adenocarcinoma metastasis for which surgical removal of the lesion followed by radiotherapy performed with reasonable outcome.

References

Figure 3. Sagittal and axial post Gadolinium T1-weighted MRI demonstrating decreased tumor size after surgery and radiotherapy

Figure 4. Preoperative (left) and 3 months postoperative (right) Sagittal T2-weighted MRI verifying marked decline in patient’s syrinx size.


