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CASE REPORT – OPEN ACCESS

Rare Presentation of a Common Hematological Malignancy

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ABSTRACT

Background: Metastatic spinal cord compression (MSCC) is a well-recognized complication of underlying malignancies; however, MSCC is rarely seen in acute leukemia.

Case Report: A 24-year-old female presented with low backache, right lower limb radicular pain, right foot drop, and loss of bladder sensation, suggestive of cauda conus syndrome. Magnetic resonance images revealed an enhanced pre-/paravertebral lesion extending from the L5-S3 level with destruction of the vertebral body and intraspinal infiltration. A peripheral blood smear revealed atypical cells, which bone marrow analysis confirmed to be vacuolated myeloblasts. A diagnosis was made of acute myeloid leukemia (AML) type M4 with spinal myeloid sarcoma leading to cord compression. A cytarabine and doxorubicin-based chemotherapy regimen was initiated. Her symptoms improved after 2 cycles, however, she developed febrile neutropenia after 4 cycles of treatment and died.

Conclusion: MSCC, although rare, can be a presenting complaint of AML resulting from cord compression due to spinal myeloid sarcoma.

Keywords: Acute leukemia, acute myeloid leukemia, metastatic spinal cord compression, myeloid sarcoma

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INTRODUCTION

Metastatic spinal cord compression (MSCC) is a common complication of solid malignancies, but with the exception of multiple myeloma, is rarely encountered in hematological neoplasias. This report describes the case of a 24-year-old female with undiagnosed acute myeloid leukemia (AML) who presented with cauda conus syndrome secondary to a spinal myeloid sarcoma.

CASE REPORT

A 24-year-old female presented with sudden-onset low backache ongoing for 5 days. It was severe in intensity and was aggravated on change of posture, bending forward, and coughing/sneezing, affecting her daily activity. The patient also complained of an electric shock-like sensation occasionally radiating down the right lower limb. She had also noticed that she was unable to perceive hot or cold sensations on the lateral aspect of her right foot while bathing and she was finding it difficult to clear her right foot from the ground while walking. She also complained of bladder and bowel incontinence for 3 days. Other than a history of menorrhagia for 2 months, her history was not significant. A physical examination revealed tachycardia, pallor, and sternal tenderness. There were no visible spinal deformities. A right foot drop and a loss of touch/temperature sensation over the lateral aspect of the right leg and foot were observed. Right ankle and plantar reflexes could not be elicited, and it was determined that saddle anesthesia and a loss of the anal reflex were present.

Magnetic resonance imaging (MRI) of the lumbosacral spine revealed an enhanced pre- and paravertebral lesion extending from the L5-S3 level with destruction of vertebral body and intraspinal infiltration (Fig. 1a, b). Other than a finding of hyperuricemia (uric acid: 8.4 mg/dL), the biochemical panel was unremarkable. A complete blood count revealed anemia (hemoglobin: 6.2 gm/dL), leukocytosis (25,000/µL), and thrombocytopenia (52,000/µL). A peripheral blood smear showed atypical cells suggestive of blasts. Subsequently, a bone marrow analysis revealed abnormal promyelocytes and vacuolated blast cells (Fig. 1c), confirmed to be myeloblasts with immunocytochemistry. The patient was diagnosed with AML type 4. Given the clinical setting of AML and radiological features of paravertebral mass infiltrating the spinal cord, a diagnosis of myeloid sarcoma-related cauda conus syndrome was made. The patient refused a biopsy of the paravertebral mass.

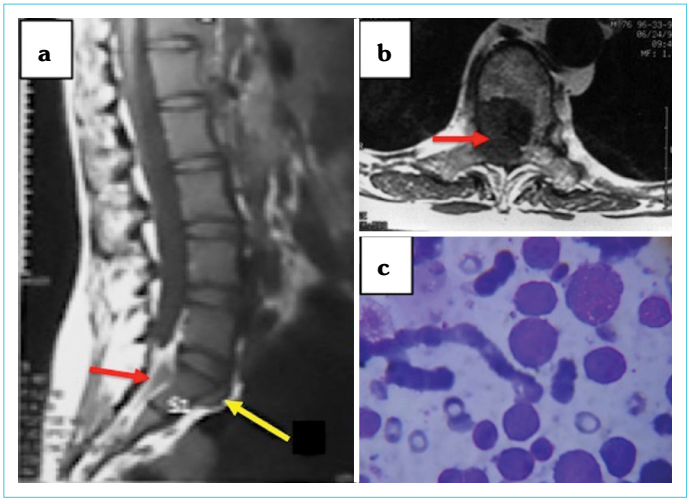

Figure 1. Magnetic resonance image (MRI) of the lumbosacral spine (T1-weighted, sagittal section) showing a pre-vertebral soft tissue mass in the L5-S1 region (yellow arrow) extending into the lower spinal cord (red arrow); (b) MRI of the lumbosacral spine (T1-weighted, axial section) showing destruction of the vertebral body (L5). (c) Photomicrograph of bone marrow aspirate showing well-defined vacuolated myeloblasts (100x, Wright-Giemsa Stain)

A cytarabine and doxorubicin chemotherapy regimen was initiated, along with parenteral dexamethasone to relieve the cord compression. Other supportive measures included packed red blood cell transfusion, allopurinol (to prevent tumor lysis syndrome), and antiviral, antifungal, and *Pneumocystis carinii* chemoprophylaxis. In addition, external beam radiotherapy was performed over the lower lumbar and sacral spine (30 Gy over 10 fractions). Corticosteroid use was continued for 2 weeks. Following 2 cycles of chemotherapy, her backache, foot drop, and bladder/bowel incontinence improved. However, after the fourth cycle, the patient developed febrile neutropenia with multidrug-resistant *Escherichia coli* and died.

DISCUSSION

MSCC is a well-recognized complication of cancer and usually presents as an oncological emergency. MSCC usually results from collapse or compression of a vertebral body destroyed by metastatic disease, but can also be caused by direct tumor extension into the spinal cord. An estimated 15% of all patients with advanced cancer develop MSCC. It is more commonly encountered in patients with solid malignancies, such as breast, prostate, or lung cancers (1). Among hematological malignancies, multiple myeloma is most commonly associated with MSCC; it is seldom seen in patients with leukemia (2).

Although occasional reports of spinal cord compression due to myeloid sarcoma have been described in the literature, the entity is extremely rare (3–6). Also known as granulocytic sarcoma, it is a tumor formed by myeloid precursors at an extra-medullary site. The prevalence of myeloid sarcoma in the spine is estimated to be <1% among all patients with acute and chronic myeloid leukemia (7). In a study that involved 32 patients with AML and spinal myeloid sarcoma, 9 patients had a spinal lesion as the initial manifestation of leukemia, as in the present case. The lumbosacral and thoracic regions of the spine were most commonly involved. Twenty-seven patients had multiple or contiguous multilevel spinal involvement. The type of spinal granulocytic sarcoma was classified according to location: epidural in the central spinal canal, epidural along the nerve course, thickening of the nerve root itself or prevertebral (7). The present case had prevertebral involvement with intraspinal extension, probably including the nerve roots. In addition, there was evidence of destruction of the vertebral body by the myeloid sarcoma.

The diagnosis of myeloid sarcoma is often challenging, especially in a patient presenting for the first time. The differential diagnosis of spinal myeloid sarcoma includes lymphoma, metastasis, extramedullary hematopoiesis, and neurogenic tumor with extramedullary imaging features. An imaging finding of multiple or a contiguous multilevel extramedullary mass of the spine with diffuse abnormal bone marrow signal intensity helps in the initial diagnosis of spinal myeloid sarcoma in association with leukemia. A solitary, dumbbell-shaped mass in the intervertebral foramen with diffuse bone marrow infiltration visualized on MRI may prompt consideration of spinal myeloid sarcoma. Unless evidence of diffuse bone marrow infiltration is seen on an MRI, the signal intensity of myeloid sarcoma can be used to differentiate a myeloid sarcoma from a neurogenic tumor. In this circumstance of mimicking a neurogenic tumor, the intermediate signal intensity (less high-signal intensity) of myeloid sarcoma on T2-weighted images is a helpful finding. Intermediate signal intensity on T2-weighted images and isointensity on T1-weighted images is seen in myeloid sarcomas (7). Post-gadolinium contrast images show homogenous enhancement.

Tissue diagnosis with immunocytochemistry is confirmatory. The diagnosis is, however, straightforward in those already diagnosed with AML. Systemic therapy directed against the underlying AML is the treatment of choice; as many as 28% and 38% of the patients respond to initial chemotherapy with complete or partial reduction in tumor volume, respectively (7). Although lumbosacral spine imaging was not repeated in this case, one can assume that there had been at least a partial reduction in the tumor volume, leading to improved symptoms. Adjuvant corticosteroid therapy during the acute stages can help reduce cord edema.

CONCLUSION

Spinal myeloid sarcoma can be a rare cause of metastatic spinal cord compression. It should be included as a first differential diagnosis in AML patients presenting with symptoms of cord compression.

Informed Consent: Written, informed consent was obtained from the patient's family for the publication of this case report and the accompanying images.

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