Aggressive Myeloid Sarcoma Causing Recurrent Spinal Cord Compression

Jacob R. Joseph[1], D. Andrew Wilkinson[1], Nathanael G. Bailey[2], Andrew P. Lieberman[2], Christina I. Tsien[3], Daniel A. Orringer[1]

Key words
- Myeloid sarcoma
- Spinal cord compression

Abbreviations and Acronyms
AML: Acute myelogenous leukemia
MRI: Magnetic resonance imaging

From the Departments of [1]Neurosurgery, [2]Pathology, and [3]Radiation Oncology, University of Michigan, Ann Arbor, Michigan, USA

To whom correspondence should be addressed: Daniel A. Orringer, M.D. [E-mail: dorringe@med.umich.edu]

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OBJECTIVE: **Myeloid sarcoma is a rare extramedullary solid tumor comprised of immature myeloid precursor cells, most commonly associated with acute myelogenous leukemia (AML).**
We present the case of a patient with a history of Shwachman-Diamond syndrome and AML who presented with myeloid sarcoma causing acute spinal cord compression.

CASE DESCRIPTION: **The patient was a 20-year-old man who presented with acute onset weakness and numbness in his lower extremities. Magnetic resonance imaging revealed a thoracic dorsal epidural mass. Despite the history of AML, we elected to forego image-guided biopsy and up-front radiation due to the rapidly progressive nature of his myelopathy. Immediate surgical decompression was performed, but the patient had recurrence of tumor leading to further compression 13 days postoperatively. Subsequently, emergent radiation was performed, leading to resolution of cord compression and local disease control.**

CONCLUSIONS: **To our knowledge, there are no randomized controlled trials examining the appropriate timing for postoperative radiation. Because most typical neuro-oncologic cases have no need for immediate postoperative radiation, our practice has been to wait 14 days to initiate postoperative radiation to ensure wound healing. One unique feature of our case was the rapid recurrence of symptoms due to tumor progression. Given this observation, we believe that radiation therapy should be considered as soon as possible after confirmatory pathology diagnosis for patients presenting with neurological compromise due to myeloid sarcoma of the spine.**

INTRODUCTION

Myeloid sarcoma, previously known as granulocytic sarcoma or chloroma (9), is a rare extramedullary solid tumor comprised of immature myeloid precursor cells. Tumor locations are widely variable, but have been known to involve the spinal canal. They are most commonly associated with a previous diagnosis of acute myelogenous leukemia (AML), and have an incidence of 2.9%–3.1% in these patients (11, 14). They can also precede the diagnosis of AML (16). Myeloid sarcoma has also been associated with other myelodysplastic and myeloproliferative disorders, but may occur in isolation (2). Presence of myeloid sarcoma is generally regarded as a poor prognostic indicator from a hematologic standpoint (18), although nonleukemic myeloid sarcoma may have increased survival in comparison with AML (20). We present a case of recurrent, aggressive myeloid sarcoma presenting as acute spinal cord compression in a patient with a history of Shwachman-Diamond syndrome and AML.

CASE DESCRIPTION

History and Physical Examination

A 20-year-old man with a history of Shwachman-Diamond syndrome and refractory AML presented with acute onset weakness and numbness in his lower extremities. Previous treatment history included chemotherapy and bone marrow transplant. Approximately 1 week before presentation, he developed midthoracic pain that migrated to his left scapular region and was initially resolved with nonsteroidal anti-inflammatory agents. On the day before presentation, the patient noticed onset of right lower extremity paresthesias, followed by lower extremities weakness. By the day of presentation, he was unable to walk and was brought to our facility.

On physical examination, the patient had significant weakness (Medical Research Council grade 2/5) in the bilateral lower extremities, and a T7 sensory level to fine touch and pinprick with preserved proprioception. He had upgoing toes bilaterally and hyperreflexia at the bilateral patellar tendons. Rectal tone and perineal sensation were normal.

Treatment Course

An infusion of 30 mg/kg methylprednisolone sodium succinate was initiated, and mean arterial pressures were kept at more than 90 mm Hg. Emergent magnetic resonance imaging (MRI) of the total spine revealed a dorsal epidural mass that was marginally hyperintense on T_1 signal, and measured 1.0 cm × 2.2 cm × 8.3 cm, extending from T4 to T9, resulting in severe spinal cord compression (Figure 1). Due to the rapidly progressive nature of his myelopathy, the amount of

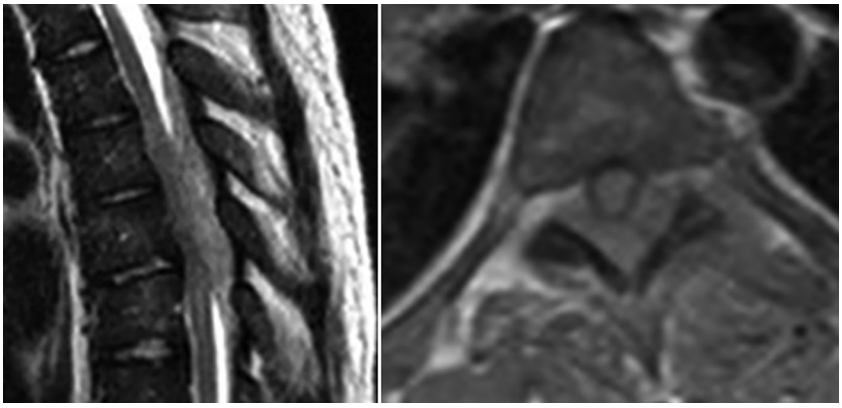

Figure 1. Preoperative T_2-weighted sagittal (*left*) and axial (*right*) magnetic resonance imagings of posterior epidural mass causing spinal cord compression.

time to obtain an image-guided biopsy and confirm pathology was deemed unacceptable, therefore we elected to forego image-guided biopsy with likely up-front radiation, despite the history of AML. The patient was taken immediately to the operating room, where a T5-T8 decompressive laminectomy was performed. There was a dense fibrous mass in the epidural space causing significant compression. Subtotal resection of the dorsal mass was performed for decompression and to restore pulsatility. Frozen section of the mass revealed a hematopoietic malignancy. Sheets of neoplastic cells morphologically compatible with blasts were present in the epidural resection, infiltrating dense fibrous tissue and adipose tissue. Neoplastic cells exhibited weak expression of CD68 (KP1) and partial expression of CD43 and CD117, without detectable expression of lymphoid antigens or keratins. Findings were diagnostic of a myeloid sarcoma (Figure 2).

Postoperatively, the patient's lower extremity strength and bladder control improved. However, on postoperative day 13, he developed increased urinary retention and lower extremity weakness. Repeat MRI showed regrowth of the epidural mass, particularly in the lateral recesses (Figure 3). Because of the morbidity associated with a transpedicular decompression and long-segment fusion, we opted to proceed with emergent radiation therapy, with a total of 30 Gy administered daily during 10 days. With radiation, lower extremity motor function gradually improved.

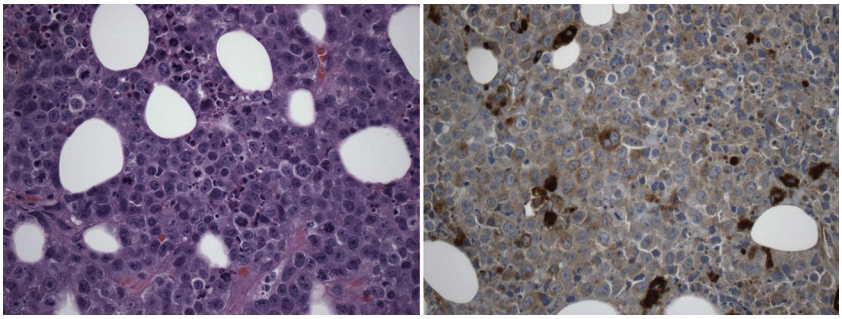

Figure 2. Hematoxylin and eosin stained section (*left*, ×400 magnification) displays sheets of neoplastic cells characterized by large size, with large round to slightly indented nuclei, fine immature chromatin, and prominent nucleoli. There are numerous areas of karyorrhexis and frequent mitoses. Immunohistochemistry (*right*, ×400 magnification) confirms the diagnosis of myeloid sarcoma, demonstrating weak positivity for CD68 (KP1, pictured) and partial expression of CD43 and CD117 (not shown).

Three months postoperatively, the patient was able to ambulate with the use of a walker. Follow-up MRI 2 months after radiation therapy showed significant decrease in cord compression and epidural tumor size (Figure 4). Shortly thereafter, the patient was found to be in liver failure due to suspected leukemic infiltration. He was eventually transitioned to hospice care, and expired 5 months postoperatively.

DISCUSSION

Myeloid sarcoma is relatively uncommon, occurring in 3% of patients with AML (11, 14). Those involving the spinal column or spinal cord are rare, and the optimal treatment for cases presenting with neurological complications has not been established. A recent case series of the literature identified 55 cases with myeloid sarcoma and spinal complications, with most receiving a combination of surgery, chemotherapy, or radiotherapy (12). However, the timing of these therapies in the acute setting of cord compression, as well long-term outcomes after treatment, have generally not been reported in individual case reports.

We describe a patient with acute onset myeloid sarcoma presenting with lower extremity motor and sensory disturbance. We presumed that neurological symptoms were due to mass effect on the spinal cord and chose to treat the patient with high-dose steroids and emergent surgical decompression within 24 hours of the onset of symptoms. It was deemed at this initial presentation that the amount of time needed for confirmation of pathology from a biopsy would be unacceptably long. Upon relapse of motor and sensory deficits on postoperative day 13, we elected to irradiate the patient and observed good clinical and radiologic improvement.

As with most types of tumors of the central nervous system, optimal treatment combines surgery and radiotherapy. Our case raises the issue of appropriate timing of surgical decompression and radiotherapy in myeloid sarcoma. In the initial emergent presentation, rapid surgical decompression provides immediate resolution of spinal cord compression and establishes a tissue diagnosis. It does bear mentioning that an alternative option would be to perform a biopsy followed by emergent radiation.