**Myeloid Sarcoma: Acute Myeloid Leukemia Beyond the Bone Marrow**

Carol Velez Martinez, MD1, Katie Kennedy, DO1, Kiran Kaur, MD1, Sara Cowles, DO1, Musharraf Navaid, MD1

1. Department of Internal Medicine, Brody School of Medicine at East Carolina University, Greenville, NC, USA

---

**KEYPOINTS**

- MS is a rare extramedullary manifestation of hematological disorders
- MS can present de novo or concomitant with bone marrow involvement
- Diagnosis ultimately relies on immunohistochemistry studies of tissue biopsy
- Treatment is based on individual risk stratification and performance status
- Systemic treatment is preferred over local therapy even in cases of isolated MS

---

**INTRODUCTION**

- MS is a mass consisting of immature myeloid cells
- Manifestation of leukemia, MPN, or MDS
- Male preference in both adult and pediatric cases
- Most common sites involved are the skin, bones, lymph nodes, CNS, or oral cavity
- Extramedullary invasion modulated by cytokines and adhesion molecules
- Common associated cytogenetic and molecular abnormalities include t(8;21), inv(16), complex karyotype, MLL, NPM1, FLT3-ITD, TET2, KIT
- Treatment modalities include chemotherapy, radiotherapy, hematopoietic stem cell transplant, and targeted therapies

---

**CASE SUMMARY**

- 72 female who presented for evaluation of worsening back pain and new abdominal pain affecting her activities of daily living
- Past medical history notable for hypertension, bilateral osteoarthritis of the knees, class 3 obesity, performance status ECOG 3
- Vitals were stable on presentation and bloodwork notable for normocytic anemia
- CT abdomen showed posterior mediastinal soft tissue density extending from the carina into upper abdomen, and encasing the lower thoracic to upper abdominal aorta
- Underwent EUS with biopsy of mass
- Developed peripheral blast concerning for AML and worsening back pain as well as associated lower extremities paresthesia, and urinary incontinence
- MRI revealed the posterior mediastinal mass extending from T6 through L1 and epidural involvement from T4 through L2 resulting in SCC
- Molecular studies showed adverse risk – poor prognosis mutations: FLT3, EZH2, DMNT3A, NPM1, trisomy 8
- Received AML high intensity induction treatment with cytarabine and idarubicin complicated by tumor lysis syndrome and hypoxic respiratory failure
- Subsequent imaging, including CT and MRI, showed significant mass reduction and resolution of SCC, though neuro-deficits did not improve significantly

---

**DISCUSSION**

- MS carries a poor prognosis given its association with adverse risk genetic abnormalities
- Hematological disease specific chemotherapy is the preferred treatment
- SCC from MS incurs in poor prognosis
- Case reports of SCC managed with local therapy with decompression surgery showed the worse overall prognosis
- Combined surgery and systemic chemotherapy showed improved survival than monotherapy
- HSCT and maintenance chemotherapy resulted in longer disease-free survival
- Surveillance imaging plays an important role in monitoring for relapse
- Relapse can present as intra or extramedullary involvement and is treated similarly
- This case helps expand our knowledge on the prognostic factors and therapeutic approach

---

**REFERENCES**

- Zhao H. et al. Clinical Characteristics, treatment, and prognosis of 278 cases of myeloid sarcoma. Scientific Reports 12, 6752-6762 (2022)

---

**ABBREVIATIONS**

- Myeloid sarcoma MS, acute myeloid leukemia (AML), myeloproliferative neoplasm (MPN), myelodysplastic syndrome (MDS), central nervous system (CNS), matrix metalloproteinase (MMP) 9, leukocyte B2 integrin, tissue inhibitor of metalloproteinase 2 (TIMP2), enhancer of Zeste 2 (EZH2), mixed-lineage-leukemia (MLL) ten-eleven translocation 2 (TET2), nucleophosmin (NPM1), cat scan (CT), radiotherapy (RT), endoscopic ultrasound biopsy (EUS), magnetic resonance imaging (MRI), spinal cord compression (SCC), Hematopoietic stem cell transplant (HSCT)