EXTRAMEDULLARY ACUTE MYELOID LEUKEMIA: TESTICULAR SARCOMA, LEUKEMIA CUTIS WITH LEPTOMENIGEAL INVOLVEMENT

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INTRODUCTION

Myeloid sarcoma also called granulocytic sarcoma, myeloblastoma, or chloroma is an extramedullary tumor of immature granulocytic cells. Extramedullary soft tissue manifestations are relatively rare in hematologic malignancies. One of the rarest manifestations is myeloid sarcoma (MS). MS develops as part of acute myeloid leukemia, myeloproliferative neoplasm, or myelodysplastic syndrome or at relapse, especially following allogeneic hematopoietic stem cell transplant. Demographically, it has a slight male predominance with a male to female ratio of 1.2:1. It may occur at any age and any site in the body leading to very varied clinical presentations. We report a rare case of extra medullary myeloid sarcoma appearing in testes and skin with leptomeningeal involvement.

Patient Description

44-year-old Caucasian male with past medical history of morbid obesity, hypertension, hyperlipidemia and diabetes mellitus type 2.

Clinical, Histopathology & Radiology images

CONSOLIDATION THERAPY

He was started on high dose cytarabine (HiDAC) for consolidation. Repeat PET scan showed 7 areas of hypermetabolic foci involving nodulated adjacent to bilateral lower anterior abdominal wall and subcutaneous stranding of bilateral buttock. Biopsy of these lesion were negative.

He completed 2 cycles of HiDAC but had repeated hospital admissions and therapy was switched to acrycinate and venetoclax.

He was seen by bone marrow transplant team and plan was for haplo (if possible) or MMUD Match. 9/10 available from registry, 2 half siblings were being typed.

Patient presented with severe throbbing lower back pain associated with bilateral lower extremity heaviness and weakness. He had also developed cellulitis at the site of ommaya reservoir.

MRI brain showed new patchy T2/FLAIR hypointense signal in the adjacent right hemisphere, monomerized cortex and subcortical, smooth ependymal enhancement in the lateral ventricles. Increased number and size of marrow replacing lesion throughout the visualized skeleton concerning for metastatic disease.

Patient underwent right iliac bone biopsy that showed >90% marrow involvement of previously diagnosed myeloid sarcoma (sheets of infiltrative cells with identical phenotype). Cytology is small round blue cells with somewhat eccentric cyttoplasm and a degree of maturation characterized by variable nuclear complexity. Positive for CD56 (>90% of marrow cellularity), CD4 and lymph. Possible small subset for CD68. Negative for: synaptophysin, chromogranin, S100, CD117, CD34, CD20, CD3, CD8, MPO, CD138.

The hospital course was complicated with acute kidney injury, electrolytes imbalance concerning for spontaneous tumor lysis syndrome. He became hemodynamically unstable and transferred to critical care for vasopressor support.

Once patient was stabilized, discussions were held for re-induction of chemotherapy with CLAG (G-CSF 300 mcg sc, cladribine 5 mg/m2 over 2 hours, and cytarabine 2 g/m2 over 4 hours beginning 2 hours after cladribine, all days times 5 days) vs palliative care consult.

Patient made the decision of pursuing comfort care collectively with his family and forego any aggressive treatment. He was transferred to palliative floor and passed away peacefully.

DISCUSSION

This case highlights a rare unique presentation of AML as myeloid sarcoma of testis with normal bone marrow. Misdiagnosis is not uncommon and can delay the appropriate treatment. Extra medullary involvement from leukemia is very aggressive and needs high suspicious and prompt treatment. Systemic chemotherapy using AML-like regimen should be commenced early, even in non leukemic disease. Allogeneic hematopoietic stem cell transplantation has demonstrated promising results, particularly in patients who achieved complete remission with AML-induction protocols, and recent advances in genetic profiling may enable the development of novel targeted therapies. Proactive multicenter controlled trials are required to further refine management decisions and investigate the role of novel targeted therapies.

REFERENCES