Diagnostic Challenges of Aleukemic Myeloid Sarcoma

Shelby Boock, BS[1], Nathanial Hansen, DO[2]

[1]Philadelphia College of Osteopathic Medicine; [2]Department of Dermatology, Beaumont Hospital, Farmington Hills

BACKGROUND

- Myeloid sarcoma (MS) is an extramedullary manifestation of acute myeloid leukemia (AML), myeloproliferative neoplasms, or myelodysplastic syndromes, characterized by immature myeloid precursor cell proliferation. MS may present solely as a cutaneous entity, lacking peripheral blood or bone marrow involvement.
- The median age of people diagnosed is 37 years old, with females being twice as likely to have the disease. Presenting symptoms include red, brown, or violaceous masses, local bleeding, numbness, or pain. The most effective form of treatment is chemotherapy, with a complete remission rate of 57.9%. The relapse and progression to AML rates remain high at 73.7% and 47.4%, respectively. The median overall survival rate is 30 months.

CLINICAL COURSE

The patient is an 83-year-old female with a past medical history of hypertension and dyslipidemia, who presented with 6 weeks of enlarging, “red bumps” on her right anterior shin. Her review of symptoms was pertinent for a 10-pound weight loss in a year. There were five nontender, violaceous nodules located anteriorly in the center of the right shin. A punch biopsy, Figure 1, revealed a diagnosis of, “Malignant Hematolymphoid Neoplasm.” The patient was referred to hematology/oncology where repeat flow cytometry testing resulted in positive CD43, Ki-67, c-myc, CD117, myeloperoxidase, and negative for B and T-cell markers. A bone marrow biopsy and fluorescence in situ hybridization (FISH) panel were both negative for AML. The chosen course of treatment was decitabine and venetoclax. The patient’s treatment was complicated by chemotherapeutic side effects, leading to a discovery of a mass in the common bile duct. The biopsy was consistent with the patient’s already diagnosed myeloid sarcoma. A cholecystectomy was scheduled, the chemotherapy was postponed.

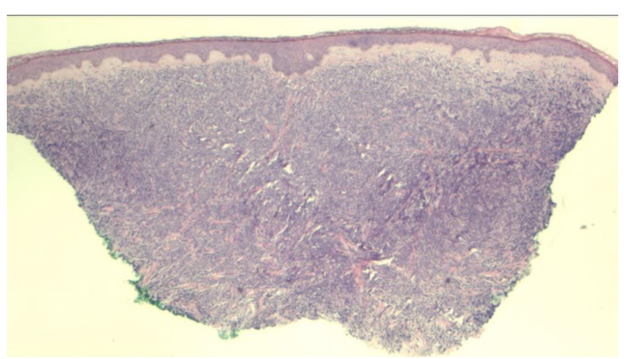

Figure 1. A punch biopsy of the largest lesion on the patient’s right leg revealed diffuse, dermal sheet growth pattern of lymphocytic infiltrates that extended to the deep dermis sparing the epidermis. Infiltrate was predominantly composed of large cells.

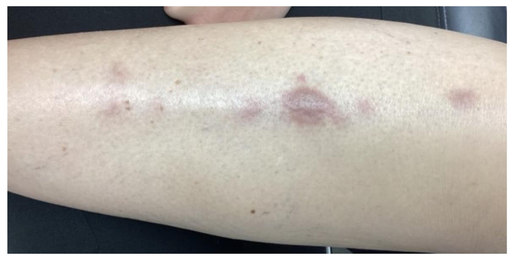

Figure 2. Lesions first seen on 10/17/2023

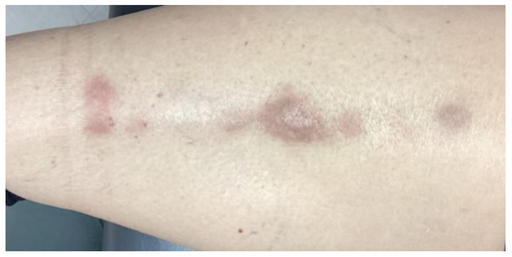

Figure 3. Lesions on 12/8/2023

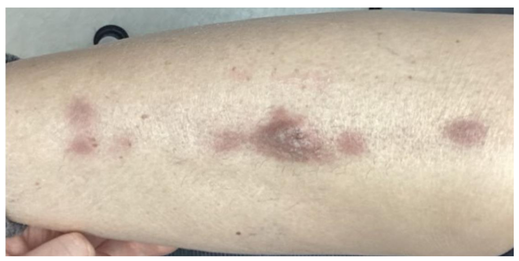

Figure 4. Lesions on 12/22/2023

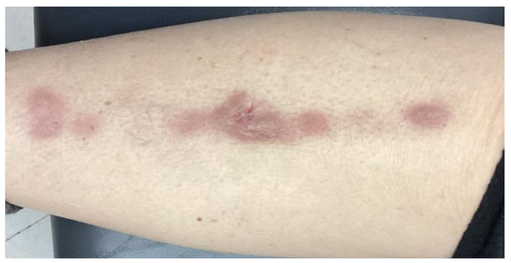

Figure 5. Lesions on 1/5/2024

DISCUSSION

To confirm a diagnosis of aleukemic MS, pathological confirmation via biopsy is necessary, peripheral blood or bone marrow testing must show an absence of leukemic cells, and no previous history of AML, myeloproliferative neoplasms, or myelodysplastic syndromes. Currently, there is no standardized chemotherapy regimen used for aleukemic MS. Various combinations of cytarabine, idarubicin, mitoxantrone, homoharringtonine, cyclophosphamide, vincristine, prednisone, adriamycin, and daunomycin have been attempted across several studies.

Exploring aleukemic MS delves into the interplay between hematological malignancies and dermatological manifestations. The ambiguous attributes of this cutaneous disease coupled with a lack of past medical history can create a diagnostic puzzle. Delay in appropriate treatment can lead to devastating outcomes in these patients as the progression is rapid. Three months after the patient’s initial presentation with the lesion depicted in Figure 2, MS was also found in the common bile duct. It is challenging to differentiate which MS-affected area originated first. To our knowledge, this is the only case with simultaneous cutaneous and bile duct involvement in MS without bone marrow or peripheral blood involvement.

REFERENCES

Zhao, H., Dong, Z., Wan, D. *et al.* Clinical characteristics, treatment, and prognosis of 118 cases of myeloid sarcoma. *Sci Rep* **12**, 6752 (2022). https://doi.org/10.1038/s41598-022-10831-7

Alexiev BA, Wang W, Ning Y, Chumsri S, Gojo I, Rodgers WH, Stass SA, Zhao XF. Myeloid sarcomas: a histologic, immunohistochemical, and cytogenetic study. Diagn Pathol. 2007 Oct 31;2:42. doi: 10.1186/1746-1596-2-42. PMID: 17974004; PMCID: PMC2186303.

Audouin J, Comperat E, Le Tourneau A, Camilleri-Broët S, Adida C, Molina T, Diebold J. Myeloid sarcoma: clinical and morphologic criteria useful for diagnosis. Int J Surg Pathol. 2003 Oct;11(4):271-82. doi: 10.1177/106689690301100404. PMID: 14615822.