



Leukemia Cutis: A Rare Clonal Presentation

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Introduction

- Chronic myelomonocytic leukemia (CMML) is a hematopoietic malignancy with monocytes.¹
- Bone marrow dysplasia can cause progression and transformation to acute myeloid leukemia (AML).²
- Diagnosis occurs via bone marrow biopsy and can present with fever, pancytopenia, and skin manifestations such as leukemia cutis.

Case Presentation

A 65-year-old male with atrial fibrillation initially presented with cytopenias, weight loss, splenomegaly, and intermittent abdominal pain. He was found to have significant increase in blasts and promonocytes accounting for ~10% of marrow cellularity. His final diagnosis favored CMML-2 and he underwent treatment with darbepoetin alfa and decitabine.

Case Presentation (cont.)

Repeat bone marrow biopsy revealed 24% of blasts, which was consistent with transformation to AML. Therefore, he subsequently underwent reinduction therapy with liposomal daunorubicin-cytarabine in this hospital. His day 22 bone marrow showed a complete response. Prior to re-induction, a papular violaceous rash developed on the upper trunk of his body and rapidly progressed. Skin biopsy confirmed leukemia cutis, a dermatologic involvement of leukemia.

Through treatment with daunorubicin-cytarabine, his skin manifestations did not show improvement. Concurrent bone marrow biopsy upon blood count recovery showed a morphologic complete response to treatment. He continued to have the rash consistent with leukemia cutis despite a bone marrow that responded to therapy. Due to the rash, he had to undergo re-induction with azacitidine and venetoclax. Ultimately, he did achieve response of AML to therapy in both his marrow and leukemia cutis.

PE: diffuse violaceous papular lesions of variable size covering portions of chest, abdomen, back and face.

Peripheral blood smear and bone marrow biopsy prior to treatment	Skin biopsy	Peripheral blood smear and bone marrow biopsy after treatment
<ul style="list-style-type: none"> ❖ thrombocytopenia, leukopenia with absolute neutropenia, normocytic normochromic anemia ❖ Diagnosis: AML 	<ul style="list-style-type: none"> ❖ diffuse perivascular infiltrate of neoplastic cells with large round to folded nuclei, prominent nucleoli, variable cytoplasm. Frequent mitotic figures. Tumor cells + for lysozyme ❖ Diagnosis: Leukemia cutis 	<ul style="list-style-type: none"> ❖ normal number of platelets & WBC; neutrophils exhibit occasional hypersegmentation with normocytic normochromic anemia ❖ Diagnosis: No evidence of residual leukemia

Conclusions

- Leukemia cutis presents as dermatologic infiltration of leukemic cells that commonly presents with papules or nodules.^{2,3}
- Most cases present with active leukemia and involvement of the marrow, however may present as an independent and isolated leukemic clone such as in this case.⁴
- This rare presentation shows persistence of leukemia cutis AML clone despite a bone marrow with complete response.
- It is important to be aware that variant clones of a single disease process may behave differently and may have discordant responses to therapies.

References

- Kwon J. Diagnosis and treatment of chronic myelomonocytic leukemia. *Blood Res.* 2021;56(S1):S5-S16. doi:10.5045/br.2021.2020321
- Jeong Hee Cho-Vega, MD, PhD, L. Jeffrey Medeiros, MD, Victor G. Prieto, MD, PhD, Francisco Vega, MD, PhD, Leukemia Cutis, *American Journal of Clinical Pathology*, Volume 129, Issue 1, January 2008, Pages 130-142.
- Parsi M, Go MS, Ahmed A. Leukemia Cutis. [Updated 2022 Apr 30]. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2022 Jan-.
- Estey, EH. Acute myeloid leukemia: 2019 update on risk-stratification and management. *Am J Hematol.* 2018; 93: 1267267- 1291