Case Report

An 83-year-old female presented with a chief complaint of multiple skin lesions on her bilateral lower extremities, in particular on the pretibial regions. The lesions first appeared three weeks prior to presentation to the office. She stated that she awoke with the lesions present and did not note any associated events or activity the night before. They had not changed in appearance or number of lesions since onset. The lesions did not itch, had no drainage, and were not painful. She denied any other symptoms, including fevers, chills, fatigue, nausea, vision changes, oral lesions, or any other rash/cutaneous changes elsewhere. She also denied a history of lower extremity edema or trauma.

Pertinent recent history included a hospitalization three months prior for atrial fibrillation and an acute colitis of unknown etiology. She did not have a history of lower extremity edema or trauma. The only cutaneous manifestations prior to, during, or after her hospitalization was an anemia of uncertain etiology without white blood cell count change since discharge was an anemia of unknown etiology. She did not have any other dermatologic involvement.

The differential at that time included excoriations, traumatic ecchymosis, vasculopathy, leukocytoclastic vasculitis, Schamberg’s disease, and early stasis dermatitis changes. Two 4-mm punch biopsies, one from each lower extremity (right proximal pretibial and left proximal pretibial locations), were obtained and sent for histology. On histologic exam, both specimens showed a superficial dermal infiltrate of atypical inflammatory cells that were enlarged and hyperchromatic with surrounding red blood cell extravasation (Figures 2, 3). A periodic acid-Schiff stain for fungi was negative for both specimens. Immunohistochemistry staining was then applied to the biopsy samples. In both biopsies, the atypical-appearing cells were highlighted by CD68 (Figures 4, 5). CD3 staining was also completed and showed only focally positive in one biopsy specimen and negative in the other. Last, a CD20 stain was non-significant with the exception of a hospitalization for acute colitis three months prior to presentation (12 weeks prior to rash development).

Upon examination, the patient had non-pruritic, violaceous, poorly demarcated macules of varying sizes on both pretibial areas (Figure 1). On the patient’s left lower extremity, one of the lesions had focal ulceration with a distinct border; otherwise, no secondary characteristics were appreciated. A full body examination was performed and revealed no other dermatologic involvement.

The patient had a past medical/surgical history of anxiety, arthritis, atrial fibrillation, hypertension, hypercholesterolemia, breast cancer with mastectomy (right), cholecystectomy, bilateral hip replacements, and squamous cell carcinoma of the upper lip. Social history included no alcohol use and no history of smoking. Current medications included low-dose aspirin, bisoprolol fumarate, digoxin, lisinopril, and pravastatin. She denied any known allergies.

Disclosures: None

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Abstract

We present the case of an 83-year-old female who presented with red macules of varying sizes, including one area of focal ulceration located on the pretibial region of her lower extremities. The lesions had been present for three weeks. She denied any systemic symptoms or current complaints at the time of presentation. Her past medical and surgical histories were non-significant with the exception of a hospitalization for acute colitis three months prior to presentation (12 weeks prior to rash development).

Punch biopsies were obtained, and histology showed an atypical lymphocytic infiltrate consistent with leukemia cutis. The patient was referred to Hematology/Oncology. Initial peripheral blood flow cytometry did not reveal atypical leukocytes, resulting in the diagnosis of aleukemic leukemia cutis. Six weeks later, she underwent a core bone-marrow biopsy and a repeat flow cytometry that revealed atypical leukocytes, leading to a final diagnosis of acute monocytic leukemia.
of the skin, which has been estimated to occur commonly associated with direct blast infiltration types of leukemia, monocytic leukemias are more and CD45. Myelomonocytic types are also lysozyme- and CD68-positive. Monocytic types IHC staining, myeloid subtypes are typically children with leukemia than adults.

Leukemia cutis is most commonly diagnosed in patients with known/active systemic leukemia; however, in rare cases, skin involvement can occur before involvement of bone marrow or peripheral blood, termed "aleukemic leukemia cutis" or "primary extramedullary leukemia." Aleukemic leukemia cutis is extremely rare, occurring in only 7% of all leukemia cutis cases. While rare, there have been reports in the literature regarding this condition since it was first termed and reported in 1948 by Epstein in the Archives of Dermatology and Syphiology. The majority of the articles are either case reports or case series reports. No formal, large-scale epidemiology studies have been completed to date, but a review in 2016 by Pena-Romero et al. involving 27 patients found that 60% of those affected were male, the mean age of diagnosis was 42 years, and the predominant leukemia type was acute myeloid leukemia.

Overall, the diagnosis of aleukemic leukemia cutis has an associated poor prognosis with an aggressive clinical course after original diagnosis similar to classic leukemia cutis. The average time between confirmed diagnosis of aleukemic leukemia cutis and involvement of blood or bone marrow is usually several months and typically less than one year. Another recent review found the mean duration between diagnosis and death to be about 10 months. However, longer latency periods have been rarely reported, including one case in which the development occurred seven years after original diagnosis. Based on the immunologic subtype, patients with aleukemic leukemia cutis are managed the same way as patients with classic leukemia cutis and patients with diagnosed blood or bone marrow involvement. There are reported cases of successful treatment remission using a variety of regimens in younger adult patients.

Conclusion

Aleukemic leukemia cutis is a rare cutaneous disease that is an early manifestation of systemic lymphoproliferative disease. The distinctive feature is atypical myeloproliferative cells in the skin prior to their detection in the peripheral blood, a distinguishing characteristic from classic leukemia cutis. The cutaneous manifestations can occur anywhere in the body but tend to favor areas of preexisting inflammation. Progression from histologic diagnosis on skin biopsy to detection in blood typically evolves over several months, although latency periods can rarely last years. Compared to classic leukemia cutis, there is no significant difference in prognosis or progression. Overall, the diagnosis of aleukemic leukemia cutis, much like classic leukemia cutis, carries a very poor prognosis with rapidly progressive disease. Typically, these patients have a high mortality rate, with progression in less than one year. We present this case for clinical interest and to highlight the importance of early recognition of this disease to decrease overall mortality.

References