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The Clinical Picture
Leukemia cutis

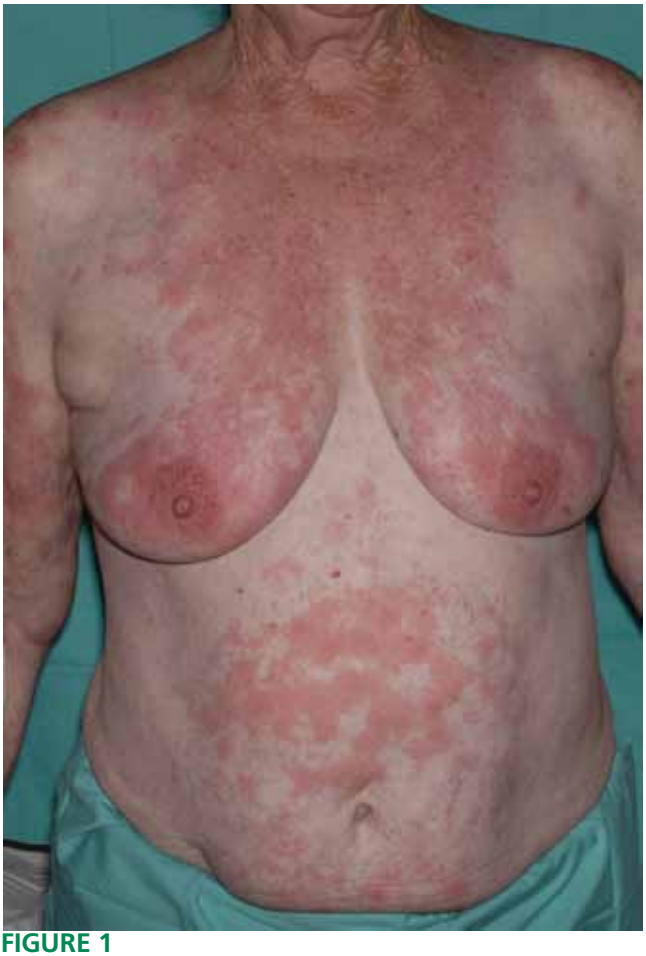

FIGURE 1

A 72-year-old woman presents with a 3-week history of diarrhea, pyrexia, and a florid but asymptomatic skin eruption consisting of infiltrated erythematous papules and nodules that coalesce into large plaques (**FIGURES 1 AND 2**). These initially afflicted her thorax before spreading to her back, arms, and legs. Pancytopenia is noted on her admission hemogram.

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FIGURE 2

She is a smoker but has been in good health and is on no regular medications.

Q: What is the most likely diagnosis?

- ☐ Leukemia cutis
- ☐ Drug reaction
- ☐ Sweet syndrome
- ☐ Erythema multiforme
- ☐ Urticaria

A: The correct answer is leukemia cutis, defined as a cutaneous infiltration by neoplastic leukocytes.[1] When the leukocytes are primarily granulocytic precursors, the terms myeloid sarcoma, granulocytic sarcoma, chloroma, and primary extramedullary leukemia have been used.[2] The term monoblastic sarcoma has been used when the cutaneous infiltrate is composed of neoplastic monocytic precursors.[2]

Leukemia cutis most commonly manifests as erythematous papules and nodules, single or multiple, of varying sizes, and afflicting one or more body sites; it typically is asymptomatic.[3] It occurs in 10% to 15% of patients with acute myeloid leukemia[4] and is itself a poor prognostic sign.[5] The cutaneous changes may pre-

date the hematologic manifestations and may even herald a relapse.[6]

In patients presenting with extramedullary leukemia and no bone marrow or blood involvement, the importance of preemptive chemotherapy for acute myelogenous leukemia has recently been emphasized.[7]

CASE CONTINUED

The patient undergoes further testing with bone marrow aspiration and trephination, which are diagnostic of acute myeloid leukemia with myelodysplastic changes: the studies reveal a clear excess of myeloblasts (accounting for 40% to 50% of nucleated cells) and clearly dysplastic erythropoiesis and myelopoiesis. Bone marrow cytogenetic analysis reveals a complex abnormal female karyotype with multiple numerical and structural abnormalities, in particular deletion of the long arm of chromosome 5, suggestive of a poor prognosis.

Skin biopsy reveals a normal epidermis but dermal perivascular involvement with a reactive T-lymphocyte infiltrate associated with immature myeloid elements, characterized by positive staining to myeloperoxidase, in keeping with leukemia cutis. It should be noted that histopathologic confirmation of leukemia cutis can be challenging, as the condition can adopt a variety of patterns, and clinicopathologic correlation is often warranted.

The patient is treated with a cycle of cytarabine-based chemotherapy, and her skin eruption transiently improves. However, her clinical condition subsequently deteriorates; she has a relapse of leukemia, with the rash returning more florid and angry-looking than previously. She is subsequently managed palliatively and passes away 3 weeks later.

THE OTHER DIAGNOSTIC CHOICES

Sweet syndrome or acute febrile neutrophilic dermatosis is often seen in association with hematologic malignancies, but the lesions are typically tender, and histopathology reveals an intense dermal neutrophilic infiltrate.[6]

Erythema multiforme is associated with malignancy, but its characteristic concentric "target" lesions are typically acral and symmetrical in their distribution; their histopathology is inflammatory.[8]

The patient had not been on any regular medications and her rash could not have been medication-induced.

Urticaria presents with pruritic evanescent wheals, which rarely last more than 12 hours.[9] Our patient had a fixed and entirely asymptomatic rash, which in addition did not have the histopathologic features of urticaria—namely, dermal edema involved with an infiltrate made of lymphocytes and eosinophils.[9]

Bone marrow studies confirm acute myeloid leukemia with myelodysplasia

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