RESIDENT ROUNDS: PART III

Granulomatous Slack Skin in an Adolescent Girl

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CASE REPORT

After immigrating to the United States from El Salvador, a 16 year-old Hispanic female presented for evaluation of a long-standing eruption. She developed hypopigmented patches over her trunk and arms at the age of 5 that subsequently became more extensive, dark, and pruritic. She was treated for presumed sarcoidosis with topical steroids without response. Our physical exam revealed extensive, indurated, dusky red plaques on the extremities, breasts, buttocks, lower back, and flanks with overlying scale and atrophy (Figure 1). Bulky, redundant skin in intertriginous areas was noted (Figure 2).

A skin biopsy demonstrated a patchy and diffuse dermal and subcutaneous infiltrate with histiocytes and multinucleated giant cells. Small to medium sized lymphocytes with irregular nuclear contours were noted with focal minimal epidermotropism. There was marked reduction of elastic fibers within the infiltrate. T-cell receptor gene clonal rearrangement showed clonality. Laboratory work-up, including flow cytometry and HTLV-1 testing, was unremarkable with the exception of hypercalcemia of 14.4. A PET scan did not show systemic involvement. Pediatric Endocrinology was consulted for the hypercalcemia, which is being managed with oral hydration. Given the extensive cutaneous involvement, she is scheduled to see Radiation Oncology for consideration for treatment with total skin electron beam.

DISCUSSION

Mycosis fungoides (MF) is the most common cutaneous T-cell lymphoma. Data from cancer registries of the Surveillance, Epidemiology, and End Results (SEER) Program found the incidence of MF to be 0.64 per 100,000 person years, with a median age at diagnosis of 55 to 60 years.[1-3] The incidence of MF/Sezary syndrome (SS) is rare before the age of 20, with an incidence rate of 0.05 per 100, 000 persons per year; however, it has been reported in children as young as 10 months old.[4, 5]

Up to 6.3% of MF cases may contain a prominent granulomatous infiltrate, the vast majority of which are due to granulomatous MF.[6] Skin manifestations of granulomatous MF are similar to classic MF with patches and plaques.[6] In contrast, distinct bulky skin folds limited to intertriginous regions characterize granulomatous slack skin.[7] Histologically, granulomatous slack skin cannot be discriminated from granulomatous MF with both defined by either prominent sarcoid-like granuloma formation or histiocytic infiltrate accounting for more than 25% of the infiltrate.[7, 8] Thus, differentiating between the two is based purely on physical exam findings.[7]

> "Histologically, granulomatous slack skin cannot be discriminated from granulomatous MF"

Granulomatous MF has been observed to have a more aggressive course and inadequate response to skin-direct treatment.[6] It has also been shown to be a poor prognostic marker with five-year mortality rate of 40%.[7, 9] In contrast, despite poor response to therapy, granulomatous slack skin appears to have a persistent but stable clinical course.[7] However, this is based on limited number of cases and is not well studied.[10]

Hypercalcemia is an exceedingly rare manifestation of MF, but has been reported with both granulomatous MF and

FIGURE 1. Numerous indurated plaques over the extremities.

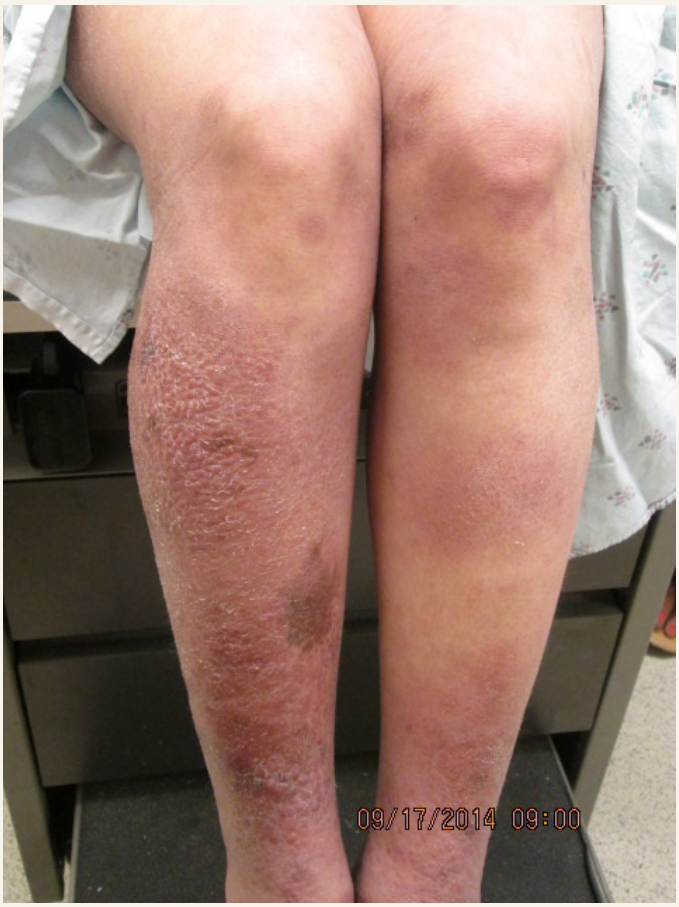
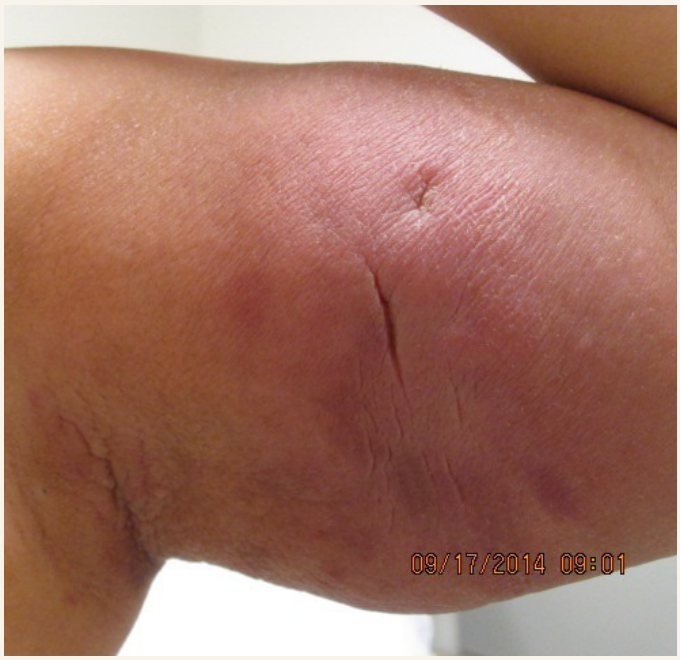


FIGURE 2. Bulky,redundant skin in the axilla.



granulomatous slack skin.[11,12] As this case demonstrates, the presence of a granulomatous infiltrate in MF warrants an evaluation for possible hypercalcemia.

DISCLOSURES

The authors have no conflicts of interest.

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