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Case presentation

Paraneoplastic erythema annulare centrifugum eruption (PEACE)

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Abstract

Erythema annulare centrifugum (EAC) is a reactive erythema with distinct, annular, erythematous plaques with trailing scale. This condition has been associated with various etiologies, which include an associated malignant condition. EAC with cancers or paraneoplastic erythema annulare centrifugum eruptions (PEACE), is more likely to be associated with lymphoproliferative malignancies such as lymphomas and leukemias. Histopathologic features include a superficial and deep, lymphohistiocytic perivascular infiltrate. We present a patient with a history of diffuse large B cell lymphoma in remission for two years, who presented with a one-year history of EAC.

Case synopsis

History: A 39-year-old woman with a history of diffuse large B-cell lymphoma in remission for two years after six cycles of chemotherapy presented to the Bellevue Hospital Center Dermatology Clinic for the evaluation of multiple, erythematous, pruritic plaques on her chest, abdomen, and back that had appeared over the past year. Topical antifungal preparations, triamcinolone acetonide 0.1% cream, and clobetasol 0.05% ointment were used twice daily for several weeks with minimal improvement. The patient reported that whereas the topical glucocorticoids helped clear the old lesions, new lesions would appear. The patient had regular follow-up with her oncologist. The patient denied fevers, nausea, diarrhea, and weight loss.

Physical examination: Multiple, 2-to-8-cm, erythematous, discrete, annular plaques with central trailing scale were present on her chest, abdomen, and back.

Laboratory data: A comprehensive metabolic panel, complete blood count, and lactate dehydrogenase were normal. The most recent positron emission tomography-computed tomography showed no evidence of a recurrent malignant condition. KOH scrapings taken at three separate clinical visits were negative.

Histopathology: There is a superficial, perivascular, lymphocytic infiltrate with occasional eosinophils with overlying epidermal acanthosis, spongiosis, and few mounds of parakeratosis. A periodic acid-Schiff stain with diastase is negative for fungal elements.



Figures 1,2. Annular erythematous plaques with trailing scale

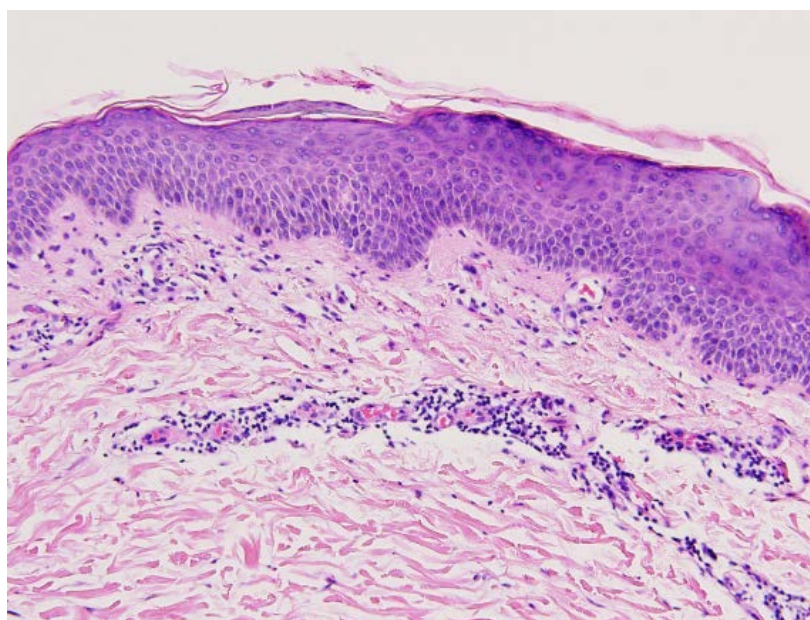


Figure 3. Perivascular lymphocytic infiltrate, acanthosis, spongiosis, focal parakeratosis

Discussion

Diagnosis: Paraneoplastic erythema annulare centrifugum eruption (PEACE)

Comment: First described in 1916, erythema annulare centrifugum (EAC) is a type of figurate erythema that is characterized by erythematous, annular papules or plaques that spread by peripheral extension [1]. EAC has been associated with numerous underlying etiologies, which include infections, such as fungal, bacterial, viral, and parasitic [2, 3]; food allergy [4]; drug reactions [5-7]; and endocrine disorders [8]. In addition, lymphoproliferative and solid tumors have been linked to the development of EAC [9-11].

In a recent review of EAC that was associated with malignant conditions or paraneoplastic erythema annulare centrifugum eruptions (PEACE), 63% of the 40 cases studied had associated lymphoproliferative diseases and 38% were linked with solid tumors [3]. In approximately one-half of the patients (46%), the appearance of EAC preceded the diagnosis of the associated cancer. In one third of the patients, the skin lesions appeared within one month of the discovery of a cancer. In 21% of patients, skin lesions followed the diagnosis of a tumor.

When EAC appears in patients with a history of cancer in remission, skin lesions may represent an early indication of a malignant recurrence. Investigators have proposed that an underlying malignant condition can elicit direct or indirect antigens or cytokines that stimulate cutaneous symptoms. If skin findings are observed and no associated tumor is initially appreciated, the tumor burden may be too low for laboratory or clinical recognition [3]. Thus, patients with a history of a malignant condition that develop EAC must be closely monitored. Studies have observed the appearance of EAC in patients with a recurrence of lymphoproliferative malignant conditions but not in those with solid tumors [3].

The histopathologic categorization of EAC is a topic of ongoing debate. In 1978, Ackerman categorized EAC into superficial and deep variants in *Histologic Diagnosis of Inflammatory Skin Diseases* [12]. However, other authors maintain that the superficial and deep type of EAC are same pathologic reactive process [13]. Investigators studied 82 biopsies from 73 patients and concluded that EAC with a superficial infiltrate has several distinct characteristics when compared to lesions that involve a deeper dermal infiltrate [14]. Clinically, the superficial type involves scale trails the expanding borders along with associated pruritus. The deeper form of EAC has indurated borders and usually lacks scale or pruritus. Histopathologically, the superficial subtype exhibited parakeratosis, spongiosis, acanthosis, and papillary dermal edema. In contrast, the deeper pattern was more likely to have a sleeve-like infiltrate, melanophages, and individual necrotic keratinocytes. Neither pattern was particularly associated with systemic disease.

The differential diagnosis for EAC includes sarcoidosis, discoid lupus erythematosus, mycosis fungoides, psoriasis, tinea corporis, and other figurate erythemas, such as erythema chronicum migrans, erythema gyratum repens, erythema marginatum, and erythema multiforme.

Treatment of EAC centers on identifying any associated pathologies that may trigger the disorder. Deep and superficial forms of EAC may be treated with oral or topical glucocorticoids. Antihistamines have been used to alleviate pruritus. In the management of PEACE, one-half of all lesions clear with the diagnosis and treatment of the associated malignant conditions [15].

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