

JAMA Dermatology Clinicopathological Challenge

Concentric Annular Truncal Eruption

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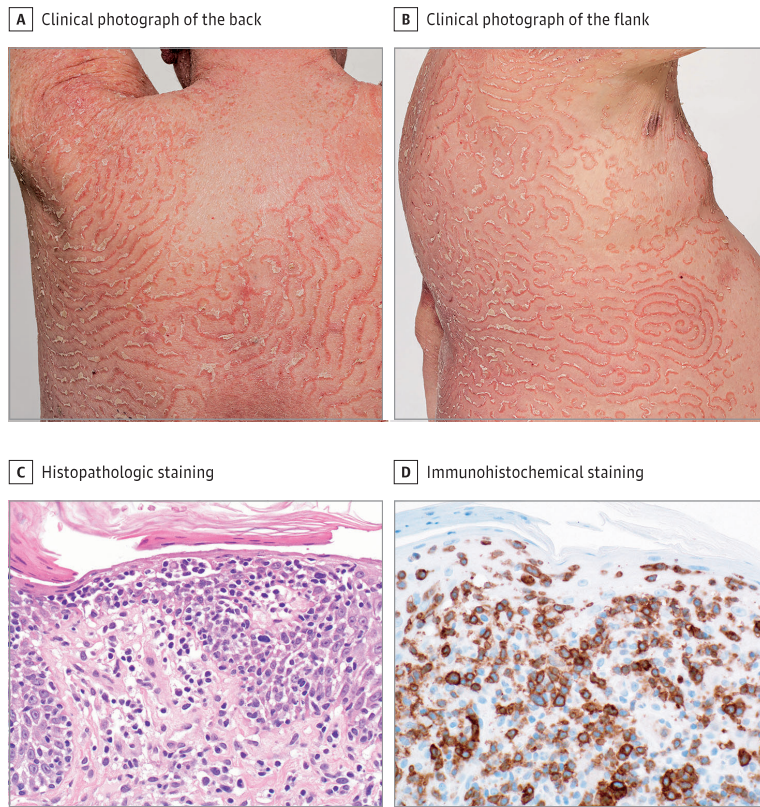


Figure. A and B, Clinical images show concentric annular, arcuate, and serpiginous red scaling plaques diffusely involving the back and right flank. C, Histopathology of skin biopsy from the chest reveals epidermal hyperplasia with spongiosis and parakeratosis (hematoxylin-eosin, original magnification $\times 400$). There is a superficial dermal and epidermotropic infiltrate of atypical lymphocytes focally distributed along the dermal-epidermal junction and aggregated in Pautrier microabscesses. D, CD30 immunohistochemical stain (original magnification $\times 400$) highlighting atypical lymphocytes.

A man in his 60s presented to the dermatology clinic with a widespread, intermittently pruritic eruption. Examination showed coalescing annular, arcuate, and serpiginous red scaling plaques in a concentric pattern diffusely involving the chest, back, and abdomen with extension to proximal extremities (Figure, A and B). Punch biopsies from the chest (Figure, C and D) and back were obtained for histopathology and tissue culture. Treatment with topical corticosteroids under wet dressings was associated with partial improvement in the eruption and pruritus.

WHAT IS YOUR DIAGNOSIS?

- A. Erythema gyratum repens associated with lung adenocarcinoma
- B. Erythema gyratum repens-like eruption associated with mycosis fungoides
- C. Erythema gyratum repens associated with pityriasis rubra pilaris
- D. Tinea imbricata

Diagnosis

B. Erythema gyratum repens-like eruption associated with mycosis fungoides

Discussion

Punch biopsies showed an epidermotropic infiltrate of small to medium atypical lymphocytes focally distributed along the dermal-epidermal junction and forming Pautrier microabscesses (Figure, C). Admixed in the infiltrate were large atypical cells, which comprised 25% to 50% of the lymphocytes depending on biopsy site. The atypical lymphocytes expressed CD2, CD3, CD4 (partial), CD30 (partial; Figure, D), and TCRαβ, and exhibited complete loss of CD7 as well as partial loss of CD5 expression. Periodic acid-Schiff stain and tissue culture were negative for fungus. Histopathologic and immunophenotypic features were consistent with mycosis fungoides (MF) with large cell transformation.

The patient had a 7-year history of refractory stage IIIA (T4N0M0B0) MF. His erythema gyratum repens (EGR)-like eruption was noted during restaging, when he also was found to have deep red to violaceous patches, plaques, and tumors of the extremities, morphologically typical for MF. Positron emission tomography-computed tomography did not show lymph node or visceral involvement. He was treated with total skin electron beam radiation.

Erythema gyratum repens presents as rapidly advancing concentric annular, serpiginous, or gyrate plaques with fine trailing scale in a wood grain pattern. EGR is associated with an underlying malignant neoplasm, most often lung, in 70% of patients.^{1,2} It also can develop in association with other skin diseases, most commonly pityriasis rubra pilaris and psoriasis. Histologic findings of EGR are non-specific and include focal parakeratosis, mild spongiosis, and perivascular lymphohistiocytic inflammation.¹ Erythema gyratum repens-like eruptions exhibit distinct histopathologic features correlating with a specific associated dermatosis, most often a bullous disease, lupus erythematosus, or MF.¹

Mycosis fungoides has been reported with an EGR-like eruption in only 8 cases, 3 with a prior history of MF³⁻⁵ and 5 heralding a new MF diagnosis.^{2,6-9} *Trichophyton rubrum* was isolated by cul-

ture in 3 patients.^{3,4,9} In the remaining patients, there was no evidence for dermatophyte by potassium hydroxide test of scale, periodic acid-Schiff histologic stain, or fungal culture.^{2,5-8} Underlying lung adenocarcinoma was discovered in 1 case.² Skin biopsies of EGR-like eruption associated with MF revealed histopathology consistent with MF alone in all but 1 case.^{2,4-9} T-cell receptor gene rearrangement analysis by polymerase chain reaction identified a clone in biopsies from 6 of 8 patients.^{2,4-8} Histopathologic features of EGR were observed in only 1 case.³

It is unclear whether an EGR-like eruption is associated with a worse prognosis in patients with cutaneous T-cell lymphoma. One patient had Sézary syndrome at onset of EGR-like eruption.⁹ One patient, besides our patient, had large cell transformation of MF.⁷ Two patients experienced MF progression after onset of EGR-like eruption, with 1 developing Sézary syndrome.^{4,5}

Erythema gyratum repens-like eruption resolved with oral terbinafine treatment in *T rubrum*-positive cases; however, MF persisted.^{3,4,9} One patient had multiple recurrences of EGR-like eruption, with each recurrence responding temporarily to terbinafine.⁹ The EGR-like eruption cleared with MF treatment in 2 patients.^{6,8} The eruption persisted after treatment of lung adenocarcinoma in 1 case.²

Dermatophyte infections are known to coincide with MF, leading to a hypothesis that MF develops secondary to a persistent cutaneous antigen stimulation.³ Tinea imbricata presents as concentric scaling plaques in association with *Trichophyton concentricum*, a dermatophyte endemic to the Pacific Islands, Southeast Asia, and Central America. Some authors have proposed that the EGR-like eruption represents tinea pseudoimbricata, a delayed hypersensitivity to *Trichophyton* species.⁹ However, association with dermatophyte was negative in most of the reported cases.

In conclusion, EGR can present as a paraneoplastic phenomenon or as an EGR-like eruption in association with another skin disease. Dermatologists and dermatopathologists should be aware that MF may manifest as an EGR-like clinical morphology, and an EGR-like eruption may represent dermatophyte superinfection in patients with classic MF histopathology.

ARTICLE INFORMATION

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