Erythema Gyratum Repens–like Eruption in Sézary Syndrome: Evidence for the Role of a Dermatophyte

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PRACTICE **POINTS**

- Erythema gyratum repens (EGR) presents as rapidly advancing, erythematous, concentric bands that can be figurate, gyrate, or annular, with fine trailing scale.
- Although EGR typically is associated with underlying malignancy, it is not an obligate paraneoplastic syndrome. There are numerous cases that are not associated with underlying neoplasms.
- An EGR-like eruption may be observed in Sézary syndrome, and an overlying superficial dermatophyte infection may play a role.

Erythema gyratum repens (EGR) is a rare and poorly understood dermatosis. The relationship of superficial dermatophytic infection to EGR-like eruptions in mycosis fungoides (MF) is unclear. We present a case of an EGR-like eruption in a patient with Sézary syndrome (SS). Histopathologic examination revealed both a superficial dermatophyte (*Trichophyton rubrum*) and cutaneous T-cell lymphoma (CTCL) in biopsies of the skin, regardless of whether those biopsies showed EGR-like lesions or erythroderma clinically. On 2 occasions, treatment of the superficial dermatophytic infection led to resolution of the EGR-like eruption and associated pruritus but not to resolution of the erythroderma. This case supports a role for dermatophytic superinfection in an EGR-like eruption in SS. Further investigation is necessary to fully understand the impact of dermatophytic infection in this clinical setting.

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Case Report

A 65-year-old woman presented with stage IVA2 mycosis fungoides (MF)(T4N3M0B2)/Sézary syndrome (SS). A peripheral blood count contained 6000 Sézary cells with cerebriform nuclei, a $CD2^{+/-}CD3^{+}CD4^{+}CD5^{+/-}CD7^{+}CD8^{-}CD26^{-}$ immunophenotype, and a highly abnormal CD4 to CD8 ratio (70:1). Positron emission tomography and computed tomography demonstrated hypermetabolic subcutaneous nodules in the base of the neck and generalized lymphadenopathy. Lymph node biopsy showed involvement by T-cell lymphoma and dominant T-cell receptor γ clonality by polymerase chain reaction.

On initial presentation to the Cutaneous Lymphoma Clinic at the University of Wisconsin-Madison, the patient was erythrodermic. She also was noted to have undulating wavy bands and concentric annular, ringlike, thin, erythematous plaques with trailing scale, giving a wood grain, zebra hide–like appearance involving the buttocks, abdomen, and lower extremities (Figure 1). Lesions were markedly pruritic and were advancing rapidly. A diagnosis of erythema gyratum repens (EGR)–like eruption was made.

Biopsy of an EGR-like area on the leg showed a superficial perivascular and somewhat lichenoid lymphoid infiltrate (Figure 2). Lymphocytes were lined up along the basal layer, occasionally forming nests within the epidermis. Nearly all mononuclear cells in the epidermis and dermis exhibited positive CD3 and CD4 staining, with only scattered CD8 cells. These features were compatible with cutaneous involvement in SS. A concurrent biopsy from diffusely erythrodermic forearm skin, which lacked EGR-like morphology, showed similar histopathologic and immunophenotypic features.

Periodic acid–Schiff (PAS) with diastase stain revealed numerous septate hyphae within the stratum corneum in both skin biopsy specimens (Figure 3). Fungal culture of EGR-like lesions was positive for a nonsporulating

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filamentous fungus, identified as *Trichophyton rubrum* by DNA sequencing.

A diagnosis of EGR-like eruption secondary to tinea corporis in SS was made. The possibility of tinea incognito also was considered to explain the presence of dermatophytes in the biopsy from skin that exhibited only erythroderma clinically; however, the patient did not have a history of corticosteroid use.

Interferon alfa-2b and methotrexate therapy was initiated. Additionally, oral terbinafine (250 mg/d) was initiated for 14 days, resulting in complete resolution of the EGR-like eruption; nevertheless, diffuse erythema remained. Subsequently, within 3 months of treatment, the cutaneous T-cell lymphoma (CTCL) improved with continued interferon alfa-2b and methotrexate. Erythroderma became minimal; the circulating Sézary cell count decreased by 50%. The patient ultimately had multiple relapses in erythroderma and progression of SS. Erythema gyratum repens–like lesions recurred on multiple occasions, with a temporary response to repeat courses of oral terbinafine.

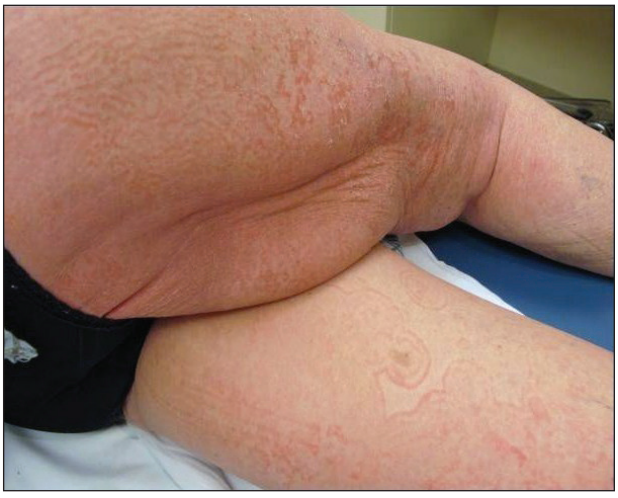

FIGURE 1. Erythema gyratum repens–like eruption on the legs.

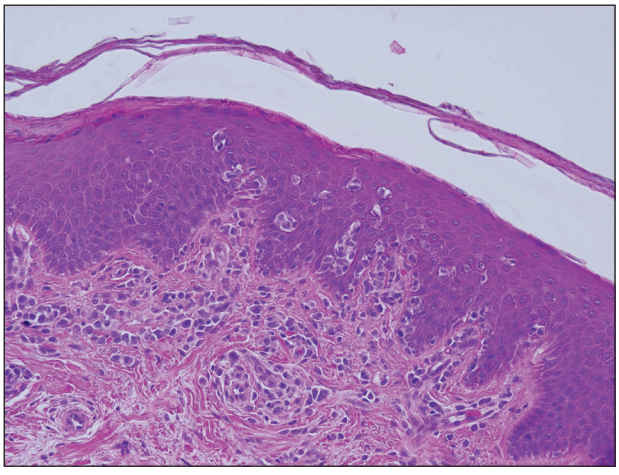

FIGURE 2. Histopathology revealed a superficial perivascular and somewhat lichenoid lymphoid infiltrate, consistent with mycosis fungoides (H&E, original magnification ×20).

Comment

Defining True EGR vs EGR-like Eruption—Sézary syndrome represents the leukemic stage of CTCL, which is defined by the triad of erythroderma; generalized lymphadenopathy; and neoplastic T cells in the skin, lymph nodes, and peripheral blood. It is well known that CTCL can mimic multiple benign and malignant dermatoses. One rare presentation of CTCL is an EGR-like eruption.

Erythema gyratum repens presents as rapidly advancing, erythematous, concentric bands that can be figurate, gyrate, or annular, with a fine trailing edge of scale (wood grain pattern). The diagnosis is based on the characteristic clinical pattern of EGR and by ruling out other mimicking conditions with biopsy.[1] Patients with the characteristic clinical pattern but with an alternate underlying dermatosis are described as having an EGR-like eruption rather than true EGR.

True EGR is most often but not always associated with underlying malignancy. Biopsy of true EGR eruptions show nonspecific histopathologic features, with perivascular superficial mononuclear dermatitis, occasional mild spongiosis, and focal parakeratosis; specific features of an alternate dermatosis are lacking.[2] In addition to CTCL, EGR-like eruptions have been described in a number of diseases, including systemic lupus erythematosus, erythema annulare centrifugum, bullous dermatosis, erythrokeratodermia variabilis, urticarial vasculitis, leukocytoclastic vasculitis, and neutrophilic dermatoses.

Prior Reports of EGR-like Eruption in Association With MF—According to a PubMed search of articles indexed for MEDLINE using the terms *erythema gyratum repens in mycosis fungoides, mycosis fungoides with tinea,* and *concentric wood grain erythema,* there have been 6 other cases

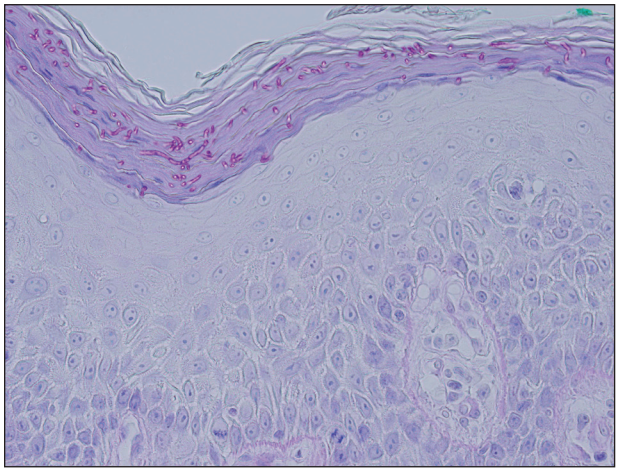

FIGURE 3. Periodic acid–Schiff with diastase stain revealed septate hyphae within the stratum corneum (original magnification ×20).

of an EGR-like eruption in association with MF (Table). Poonawalla et al[3] first described an EGR-like eruption (utilizing the term *tinea pseudoimbricata*) in a 55-year-old man with stage IB MF (T2N0M0B0). The patient had a preceding history of tinea pedis and tinea corporis that preceded the diagnosis of MF. At the time of MF diagnosis, the patient presented with extensive concentric, gyrate, wood grain, annular lesions. His MF was resistant to topical mechlorethamine, psoralen plus UVA, and oral bexarotene. The body surface area involvement decreased from 60% to less than 1% after institution of oral and topical antifungal therapy. It was postulated that the widespread dermatophytosis that preceded the development of MF may have been the persistent antigen leading to his disease. Preceding the diagnosis of MF, skin scrapings were floridly positive for dermatophyte hyphae. Fungal cultures from the affected areas of skin grew *T rubrum*.[3]

Moore et al[4] described an EGR-like eruption on the trunk of a 73-year-old man with stage IA MF (T1N0M0B0). Biopsy was consistent with MF, but no fungal organisms were seen. Potassium hydroxide preparation and fungal cultures of the lesions also were negative for organisms. The patient was successfully treated with topical betamethasone.[4]

Comparison of Case Reports of EGR-like Eruption in Association With MF

Reference (Year)	Age, y/ Sex	MF Disease Stage at Presentation	PAS and KOH Findings	Fungal Culture Results	Disease Progression
Poonawalla et al[3] (2006)	55/M	Stage IB (T2N0M0B0)	PAS-negative tissue; KOH preparation positive	*Trichophyton rubrum*	EGR-like eruption cleared with antifungals; MF with 3.5% BSA stable patches and plaques at 1-year follow-up
Moore et al[4] (2008)	73/M	Stage IA (T1N0M0B0)	No mention of PAS; KOH preparation negative	Fungal culture negative	Not mentioned
Jouary et al[5] (2008)	77/M	Stage III (erythrodermic variant [T4N1M0B0])	PAS-positive tissue	*T rubrum*	MF progressed to SS following EGR-like eruption
Cerri et al[6] (2010)	61/M	Stage I	KOH preparation and culture negative; no mention of PAS staining	No culture obtained	EGR-like eruption preceded exacerbation of MF
Holcomb et al[7] (2012)	75/M	Stage IIB (large-cell transformation [T3N0M0B0])	PAS stain and KOH preparations repeatedly negative	No culture obtained	No mention of disease progression following appearance of EGR-like eruption
Nagase et al[8] (2014)	73/M	Stage IB (T2N0M0B0)	Microscopy negative for mycotic infection	Culture negative for mycotic infection	Partial remission with PUVA combined with topical corticosteroids; no major changes in skin lesions following surgical resection of the lung cancer
Current report	65/F	Stage IVA2 (T4N3M0B2)	PAS-positive and culture-positive tissue	*T rubrum*	Antifungal treatment cleared the EGR-like lesions but not erythroderma; multiple relapses in erythroderma and progression of SS

Abbreviations: EGR, erythema gyratum repens; MF, mycosis fungoides; PAS, periodic acid-Schiff; KOH, potassium hydroxide; M, male; BSA, body surface area; SS, Sézary syndrome; PUVA, psoralen plus UVA; F, female.

Jouary et al[5] described an EGR-like eruption in a 77-year-old man with stage III erythrodermic MF (T4N1M0B0). Biopsy showed mycelia on PAS stain. Subsequent culture isolated *T rubrum*. Terbinafine (250 mg/d) and ketoconazole cream 2% daily were initiated and the patient's EGR-like rash quickly cleared, while MF progressed to SS.[5]

Cerri et al[6] later described a case of EGR-like eruption in a 61-year-old man with stage I MF and an EGR-like eruption. Microscopic examination of potassium hydroxide (KOH) preparations and fungal culture of the lesions failed to demonstrate mycotic infection. There was no mention of PAS stain of skin biopsy specimens. In this case, the authors mentioned that EGR-like lesions preceded exacerbation of MF and questioned the prognostic significance of the EGR-like eruption in relation to MF.[6]

Holcomb et al[7] reported the next case of a 75-year-old man with stage IIB MF (T3N0M0B0) with $CD25^+$ and $CD30^+$ large cell transformation who presented with an EGR-like eruption. In this case, PAS stain and KOH preparations were repeatedly negative for mycotic infection. Disease progression was not mentioned following the appearance of the EGR-like eruption.[7]

Nagase et al[8] most recently described a case of a 73-year-old Japanese man with stage IB (T2N0M0B0) $CD4^-CD8^-$ MF and lung cancer who developed a cutaneous eruption mimicking EGR. Microscopy and culture excluded the presence of a mycotic infection. The patient achieved partial remission with photochemotherapy (psoralen plus UVA) combined with topical corticosteroids. No major changes in the patient's skin lesions were noted following surgical resection of the lung cancer.[8]

Dermatophyte Infection—It is known that conventional tinea corporis can occur in the setting of CTCL. However, EGR-like eruptions in CTCL can be distinguished from standard tinea corporis by the classic morphology of EGR and clinical history of rapid migration of these characteristic lesions.

Tinea imbricata is known to have a clinical appearance that is similar to EGR, but the infection is caused by *Tinea concentricum*, which is limited to southwest Polynesia, Melanesia, Southeast Asia, India, and Central America. Although *T rubrum* was the dermatophyte isolated by Poonawalla et al,[3] Jouary et al,[5] and in our case, whether *T rubrum* infection in the setting of CTCL has any impact on prognosis needs further study.

Our case of an EGR-like eruption presented in a patient with SS and tinea corporis. Biopsy specimens showed CTCL and concomitant dermatophytic infection that was confirmed with PAS stain and identified as *T rubrum*. Interestingly, our patient's EGR-like eruption cleared with oral terbinafine therapy, consistent with findings described by Poonawalla et al[3] and Jouary et al[5] in which treatment of the dermatophytic infection led to resolution of the EGR-like eruption, suggesting a causative role.

However, testing for dermatophytes was negative in the other reported cases of EGR-like eruptions in patients with MF, despite screening for the presence of fungal microorganisms using KOH preparation, PAS staining, or fungal culture, or a combination of these methods,[3-8] which raises the question: Do the cases reported without dermatophytic infection represent false-negative test results, or can the distinct clinical appearance of EGR indeed be seen in patients with CTCL who lack superimposed dermatophytosis? In 3 prior reported cases of EGR-like eruptions in MF, the eruption was preceded by immunosuppressive therapy.[5-7]

Further investigation is needed to correlate the role of dermatophytic infection in EGR-like eruptions. Our case and the Jouary et al[5] case reported dermatophyte-positive EGR-like eruptions in MF and SS detected with histopathologic analysis and PAS stain. This low-cost screening method should be considered in future cases. If the test result is dermatophyte positive, a 14-day course of oral terbinafine (250 mg/d) might induce resolution of the EGR-like eruption.

Conclusion

The role of dermatophyte-induced EGR or EGR-like eruptions in other settings also warrants further investigation to shed light on this poorly understood yet striking dermatologic condition. Our patient showed both MF and dermatophytes in skin biopsy results, regardless of whether those sites showed erythroderma or EGR-like features clinically. On 3 occasions, antifungal treatment cleared the EGR-like lesions and associated pruritus but not erythroderma. Therefore, it appears that the mere presence of dermatophytes was necessary but not sufficient to produce the EGR-like lesions observed in our case.

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