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# Erythema gyratum repens-like presentation of folliculotropic mycosis fungoides

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**Keywords:** *erythema gyratum repens, folliculotropic, gamma chain, immunohistochemistry, mycosis fungoides, polymerase chain reaction, T cell receptor*

To the Editor:

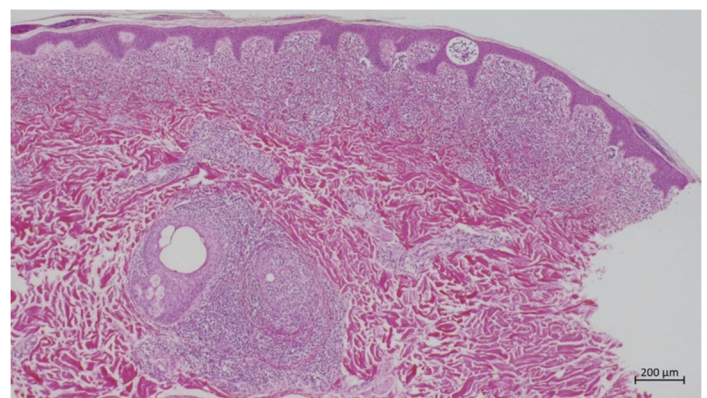
Folliculotropic mycosis fungoides (FMF) is a rare but well-defined clinicopathological variant of mycosis fungoides (MF) in which neoplastic T lymphocytes display tropism for the follicular epithelium. Folliculotropic mycosis fungoides presents with a wide variety of clinical presentations and mimics various dermatoses. Herein, we describe a patient with FMF presenting with erythema gyratum repens (EGR)-like findings. To the best of our knowledge, this is the first reported case of a unique clinical feature that mimics EGR.

A 46-year-old woman with pruritic eruption refractory to topical corticosteroid therapy was referred to our hospital. Physical examination revealed concentric erythematous plaques up to approximately 20cm in size scattered on the chest, anterior abdomen, and both thighs with a wood-grain appearance resembling EGR (**Figure 1**). Repeated microscopic examinations using potassium hydroxide and fungal cultures of the lesions ruled out mycotic infection.

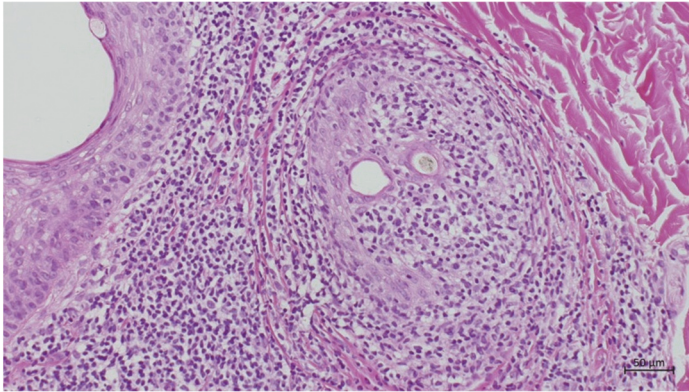
A punch biopsy of an EGR-like lesion on the chest showed follicular and band-like dermal lichenoid infiltrates of atypical lymphocytes (**Figure 2**). Careful examination of the epidermis revealed a few small foci of epidermotropism of atypical lymphocytes, similar to classic forms of MF. A higher magnification demonstrated intrafollicular and perifollicular



**Figure 1.** Concentric erythematous plaques resembling erythema gyratum repens on the chest.



**Figure 2.** Low-power view of a biopsy specimen of an erythematous plaque on the chest showed follicular and dermal lichenoid infiltrate of atypical lymphocytes. H&E.

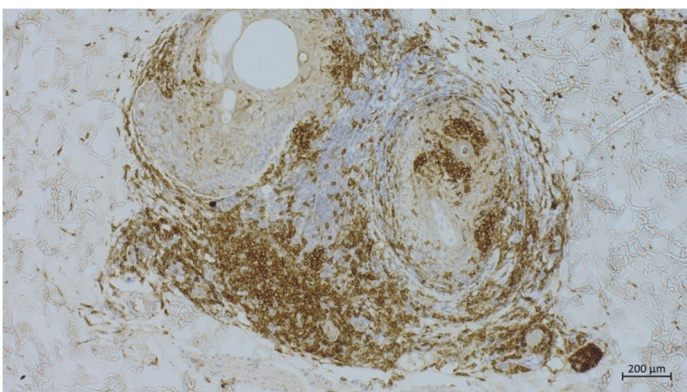


**Figure 3.** Higher magnification demonstrated perifollicular and intrafollicular infiltration of medium atypical lymphocytes without mucin deposition. H&E.

infiltration of medium-sized atypical lymphocytes (**Figure 3**). Alcian blue staining revealed no mucin deposition.

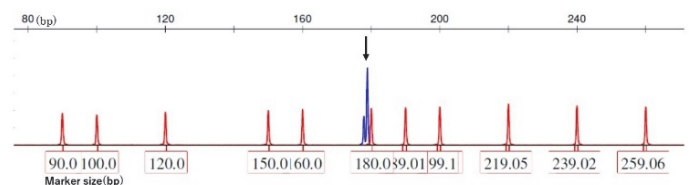
Immunohistochemically, the majority of atypical lymphocytes in the follicles, papillary dermis, and epidermis exhibited positive CD3 and CD4 staining (**Figure 4**), negative CD7 staining, and scattered CD8-positive cells. Polymerase chain reaction-based T-cell receptor (TCR)  $\gamma$  chain clonality testing using formalin-fixed paraffin-embedded samples, as described previously (van Dongen JJM et al. 2003 [1]), detected the clonality of the TCR  $\gamma$  chain gene rearrangement (**Figure 5**), suggesting the presence of clonal T-cells. These immunohistochemical and molecular features were compatible with those of FMF [2,3].

We treated the patient with narrow-band ultraviolet B phototherapy, which resulted in a partial response. We then switched to ultraviolet A1 phototherapy and most of the eruptions disappeared.



**Figure 4.** Immunohistochemical features: Perifollicular and intrafollicular atypical lymphocytes were CD4+.

Folliculotropic mycosis fungoides can present with a wide variety of clinical presentations and mimic many dermatoses. The clinical features include acneiform lesions, comedo-like lesions, milia-like cysts, prurigo-like nodules, alopecia, plaques with follicular papules, tumors, and erythroderma [2,3]. Various clinical morphologies are elicited by different histological patterns [2]. Folliculotropism of atypical lymphocytes is crucial in the diagnosis of FMF [2,3], but the following histological features were observed: epidermotropism of the non-follicular epithelium that appears in usual MF, basaloid folliculo-lymphoid hyperplasia, granulomatous changes, eosinophilic folliculitis-like presentation, formation of dilated follicular cysts, and follicular mucinosis [2]. Various clinical and histologic features can not only make diagnosis more difficult but also influence disease prognosis, because of delayed diagnosis. Host responses among patients can differ and contribute to various histological features. The diversity of reactive inflammatory cells makes the histology more diverse. In addition to differences in host response between patients, various subsets of CD4-positive neoplastic T cells may influence histological features and clinical morphologies. Immunohistochemically, neoplastic T cells in FMF express the CD4 antigen, but reactive inflammatory cells obscure the predominance of CD4-positive cells [2,3]. The most important diagnostic finding was that CD4-positive atypical lymphocytes were observed primarily in the infundibular and isthmic epithelia [2,3]. In this case, there were a few small nests in the epidermis and papillary dermal lichenoid infiltration of atypical lymphocytes, but marked infiltration by CD4-positive atypical lymphocytes to the follicular epithelium was also present. Hence, we diagnosed this case as FMF. Gerami and Guitart detected clonal expansion in 16 of 18 patients with FMF using PCR of



**Figure 5.** Polymerase chain reaction analysis for rearranged TCR  $\gamma$  chain gene revealed clonal amplification of 178 base pairs (arrow).

the TCR  $\gamma$  gene [2]. PCR-based TCR  $\gamma$  clonality testing using formalin-fixed paraffin-embedded samples is useful to confirm the diagnosis of FMF after biopsy.

A case of an EGR-like eruption associated with FMF was not reported in a PubMed article search. However, seven cases of EGR-like eruptions associated with MF have been reported [4-10]. Three of these were dermatophyte-induced EGR-like eruptions [4,6,10]. Mycosis fungoides has been known as "The great imitator," as MF mimics a large number of dermatoses [11]. Folliculotropic mycosis fungoides may also be a great imitator owing to the variety of clinical presentations. Based on this case, we must be aware that FMF can resemble EGR.

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The disease course of FMF is generally more aggressive and has a poorer outcome than that of conventional MF [12]. Regarding the treatment of FMF, Jang reported the efficacy of ultraviolet A1 phototherapy in early-stage FMF [13]. They treated 11 patients with early-stage FMF using ultraviolet A1 and reported excellent results (eight: complete remission, 3: partial response), [13]. This treatment was also effective in our case. Early diagnosis of FMF is the most important and challenging task.

## Potential conflicts of interest

The authors declare no conflicts of interest.