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Folliculotropic and syringotropic mycosis fungoides mimicking basal cell carcinoma

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Abstract

Mycosis fungoides (MF) is characterized by a clonal proliferation of skin-homing mature T cells with special predilection for involving the epidermis. Folliculotropic and syringotropic MF typically present with erythematous papules, patches, and plaques, with punctate accentuation that is folliculocentric in the former. We report a 67-year-old woman, with an extensive history of allergic contact dermatitis, who was referred to the Mohs surgery clinic with a large pink plaque extending from the nasal bridge to the right upper medial cheek concerning for basal cell carcinoma. An outside punch biopsy showed benign basaloid follicular neoplasm. The patient was found to also have indurated erythematous plaques of the bilateral upper arms and erythematous scaly patches of bilateral arms and legs, abdomen, lateral trunk, buttocks, and groin. Owing to concern for possible cutaneous lymphoma, punch biopsies were performed which revealed the diagnosis of folliculotropic and syringotropic MF. Of note, folliculotropic and syringotropic MF are often, but not uniformly, characterized by a more aggressive disease course. This case highlights the importance of a high index of suspicion and awareness of all clinical and histopathologic pitfalls to avoid misdiagnosis of MF.

Keywords: folliculotropic, mycosis fungoides, syringotropic

Introduction

Mycosis fungoides (MF) is the most common cutaneous T-cell lymphoma. Folliculotropic MF is classified as a subtype characterized by the presence of follicular-based lesions and folliculotropism, with or without follicular mucinosis [1]. Syringotropic MF, a rare variant, is predominantly observed in patients who also exhibit folliculotropic MF. We report a patient with folliculotropic and syringotropic MF who presented with a large, raised plaque of her nose along with erythematous scaly patches and indurated plaques at multiple locations on her body. Punch biopsies were conducted to address concerns of potential cutaneous lymphoma, ultimately confirming the diagnosis. Her treatment regimen included topical corticosteroids, radiation therapy, and narrowband ultraviolet B phototherapy, resulting in a favorable response.

Case Synopsis

A 67-year-old woman with a history of extensive contact allergies for ten years was referred from an outside clinic to the Mohs surgery clinic for a large lesion of her nose concerning for basal cell carcinoma. The lesion began as an eczematous rash believed to be related to her contact allergies but had become more raised in the past several months. An outside punch biopsy showed a benign basaloid follicular neoplasm. Physical examination revealed a large pink plaque extending from the nasal bridge to the right upper medial cheek (**Figure 1**) along with erythematous scaly patches of bilateral arms and legs, abdomen, lateral trunk, buttocks, and groin. Indurated erythematous plaques of bilateral upper arms were also noted.

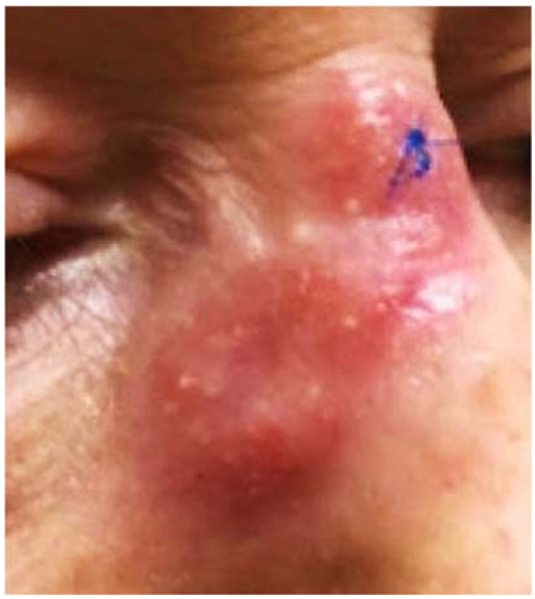

Figure 1. *Large pink plaque extending from the nasal bridge to the right upper medial cheek.*

Owing to concern for possible cutaneous lymphoma, punch biopsies were performed on the nasal bridge plaque, left upper arm, and left thigh. The nasal bridge biopsy demonstrated a dense, nodular lymphoid infiltrate in the superficial and deep dermis composed of small-to-medium sized, atypical lymphocytes. The follicular structures were dilated and distorted with follicular plugging. There were some atypical lymphocytes present within the follicular epithelium (**Figure 2**). T cell receptor gamma and beta gene rearrangement studies by PCR were positive for clonality. Subsequent biopsies of the left upper arm and left thigh were performed for further evaluation of the rash that was previously considered to represent allergic contact dermatitis. The left upper arm biopsy showed a diffuse infiltrate of small-to-medium sized, atypical lymphocytes with some concentration around the secretory eccrine units in the deep dermis, consistent with plaque-stage syringotropic mycosis fungoides (**Figure 3**). CD3 and CD4 were positive in the atypical lymphocytes throughout the dermis and CD30 expression was <5% of total infiltrate. A subset of epidermotropic CD3+ lymphocytes were CD4/CD8 negative. The left thigh biopsy showed mild spongiosis and an epidermotropic infiltrate composed of hyperchromatic lymphocytes with irregular nuclear contours, forming scattered Pautrier microabscesses within the epidermis. Systemic therapy in combination with skin directed therapy were recommended. However, the patient declined to initiate any systemic therapy and was treated with topical triamcinolone and betamethasone for the lesions on her trunk and extremities and underwent radiation therapy to her nasal area. She was further maintained with

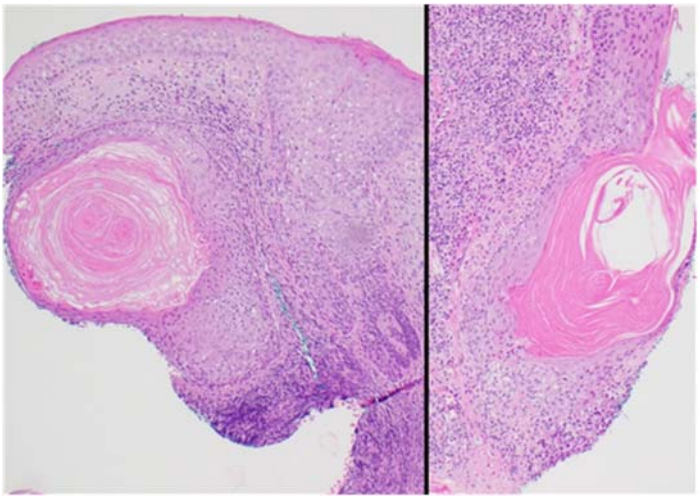

Figure 2. *H&E histopathology of nasal bridge showing a dense, nodular lymphoid infiltrate in the superficial and deep dermis composed of small to medium-sized, atypical lymphocytes. The follicular structures are dilated and distorted with follicular plugging. There are some atypical lymphocytes present within the follicular epithelium, 10×.*

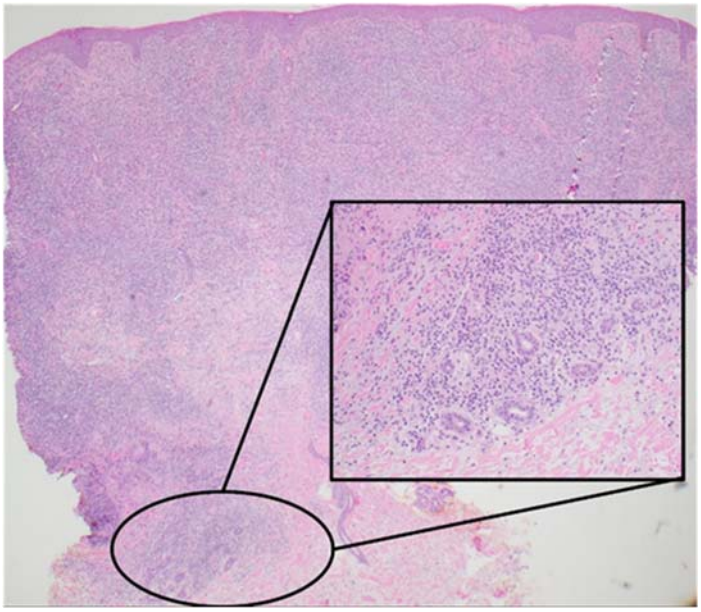

Figure 3. *H&E histopathology of left upper arm demonstrating a diffuse infiltrate of small-to-medium sized, atypical lymphocytes with some concentration around the secretory eccrine units in the deep dermis, consistent with plaque stage syringotropic mycosis fungoides, 2× and 40×.*

narrowband ultraviolet B phototherapy with surprisingly good response. The patient has not had further progression of disease after three years of follow-up care.

Case Discussion

Mycosis fungoides is characterized by clonal proliferation of skin homing mature T cells with special predilection for involving the epidermis [1]. Early stage MF typically presents as inflammatory erythematous patches or plaques [2]. Folliculotropic MF is a subtype defined as the presence of follicular-based lesions and folliculotropism with or without follicular mucinosis [1]. Syringotropic MF is a rare variant of MF seen most frequently in patients who also have folliculotropic MF. Histopathologically, syringotropic MF has a prominent involvement of the eccrine glands, often associated with folliculotropism [1,2]. Folliculotropic and syringotropic MF typically present with erythematous papules, patches, and plaques with punctate accentuation that is folliculocentric in the former; alopecia overlying the lesions is seen in approximately 63%-70% of cases.

Early-stage MF treatment focuses on controlling skin lesions mainly by skin-directed therapies, such as topical therapies, phototherapies, and radiotherapies [3]. For advanced-stages, systemic treatment with biological or targeted therapies including bexarotene and interferon are typically first-line, with more immunosuppressive chemotherapies being reserved for refractory or rapidly progressive disease. Moreover, recent improvements in biological or targeted therapies include brentuximab vedotin and mogamulizumab [3]. Most patients with early-stage MF have a good prognosis, whereas the advanced-stage prognosis is poor [4]. Of note, folliculotropic and syringotropic MF, are often, but not uniformly, characterized by a more aggressive disease course. Accordingly, systemic therapy sometimes in combination with skin directed therapy, is the preferred treatment.

Conclusion

Mycosis fungoides exhibits the capacity to imitate a wide array of both prevalent and less frequent inflammatory skin conditions, showcasing clinical and histopathological similarities [2]. At certain stages, our patient's presentation raised suspicions of both contact dermatitis and basal cell carcinoma, emphasizing the diverse nature of the clinical manifestations of MF. Given the potential aggressive nature of this condition, a high index of suspicion and awareness of all clinical and histopathologic pitfalls are necessary to avoid misdiagnosis of MF [2].

Potential conflicts of interest

The authors declare no conflicts of interest.

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