Dermatology Practical & Conceptual

Folliculotropic Mycosis Fungoides Mimicking Pityriasis Rubra Pilaris and Lichen Spinulosus: A Dermoscopic and Histopathologic Study

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Introduction

Folliculotropic mycosis fungoidesis (FMF) a rare variant of cutaneous T-cell lymphoma characterized by the presence of folliculotropic neoplastic infiltrates, leading to follicular-based lesions that often mimic various inflammatory dermatoses [1]. The term "follicular mycosis fungoides" was introduced by Kim in 1985, emphasizing the involvement of hair follicles in the lymphoproliferative process rather than the epidermis, which is often spared [2]. The clinical spectrum of FMF overlaps with several benign conditions, making histopathologic confirmation essential for diagnosis.

Case Presentation

A 60-year-old woman presented with a six-year history of refractory cutaneous lesions. Clinical examination revealed erythematous plaques with follicular hyperkeratosis, primarily distributed over the trunk and mammary region, sparing the peri-areolar area, a distribution pattern commonly associated with pityriasis rubra pilaris (Figure 1 A). On the arms and upper back, the lesions resembled lichen spinulosus, manifesting as small, follicular keratotic papules (Figure 1 B). Dermoscopy played a critical role in the differential diagnosis. Examination revealed follicular keratotic plugs

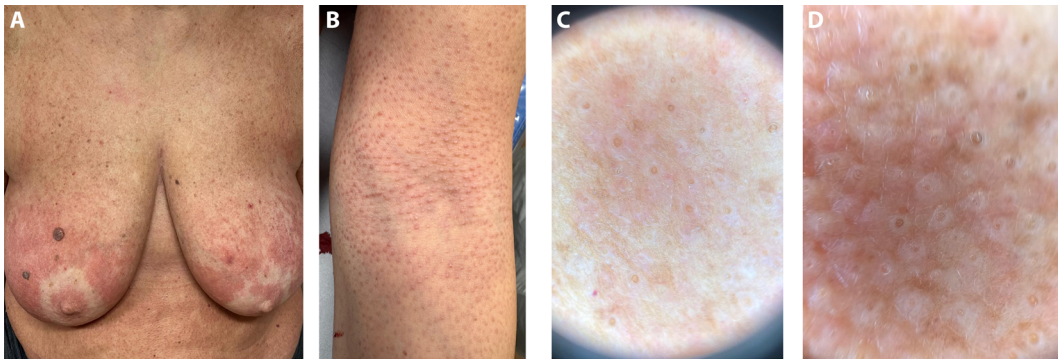

Figure 1. (A) erythematous plaques with follicular hyperkeratosis distributed over the trunk and mammary region, sparing the peri-areolar area; (B) clinical image of the affected arm, where the lesions resemblen lichen spinulosus, manifesting as small, follicular keratotic papules; (C,D) dermoscopic images showing follicular keratotic plugs surrounded by a white halo on an uneven erythematous background.

surrounded by a white halo on an uneven erythematous background (Figure 1 C,D), consistent with previously descrive dermoscopic patterns of pityriasis rubra pilaris and lichen spinulosus, which feature perifollicular hyperkeratosis and background erythema. However, recent studies on FMF dermoscopy have highlighted overlapping characteristics, including perifollicular accentuation seen as a white halo around follicles, comedo-like openings, white structureless areas, and dotted or fine linear vessels [3]. Histological analysis of two skin biopsies, one from the mammary region where lesions resembled pityriasis rubra pilaris and another from the upper arm where lesions mimicked lichen spinulosus, revealed a dense folliculotropic lymphoid infiltrate. The epidermis was largely spared, while the dermis exhibited a partial lichenoid infiltrate with mild fibrosis. Immunohistochemical analysis demonstrated a predominance of CD4+ T cells, with a CD4/CD8 ratio consistent with FMF.

Conclusion

The literature describes the dermoscopic features of FMF ashighly variable, with patterns mimicking a variety of follicle-based dermatoses, including infectious conditions such as bacterial folliculitis and tinea capitis, as well as non infectious disorders like lichen spinulosus, keratosis pilaris, acne vulgaris, and certain forms of alopecia [4]. Although research on the topicis limited, several dermoscopic features have been reported, including perifollicular accentuation, seen as a white halo around follicles, comedo-like openings, white structureless areas, lack of hair, and dotted or fine linear vessels. Additional findings such as milky-white globules, black dots, broken hairs, and keratotic cone-shaped spicules have been described in patients with extensive scalp alopecia due to FMF and in cases of spiky mycosis fungoides. These dermoscopic features indicate that FMF can closely resemble pityriasis rubra pilaris and lichen spinulosus, leading to potential diagnostic pitfalls. Pityriasis rubra pilaris typically presents with follicular plugs and diffuse erythema but lacks perifollicular halos, whereas lichen spinulosusis characterized by keratotic follicular projections on a homogeneous erythematous background. In this patient, the heterogeneous erythema combined with perifollicular white halos and follicular keratotic plugs distinguished FMF from these inflammatory conditions (Table1). Given the significant overlap in clinical and dermoscopic findings, early biopsy is essential in distinguishing FMF from benignf ollicular disorders, ensuring timely diagnosis and appropriate management.

Table 1. Comparative table FMP vs PRP vs LS.

Feature	Folliculotropic Mycosis Fungoides (FMF)	Pityriasis Rubra Pilaris (PRP)	Lichen Spinulosus (LS)
Typical Age of Onset	Middle-aged to older adults	Bimodal (children and adults)	Children and young adults
Course	Chronic, progressive	Acute or subacute; often self-limited	Often self-limiting
Distribution	Face, trunk, scalp, sometimes breasts or limbs	Trunk, extremities, scalp; may show 'islands of sparing'	Extensor surfaces of arms, thighs, sometimes trunk
Clinical Features	Erythematous plaques with follicular hyperkeratosis; alopecia may be present	Salmon-colored plaques with follicular plugs, fine scaling, and diffuse erythema	Clusters of spiny, follicular keratotic papules
Dermoscopic Features	Perifollicular white halos, follicular plugs, comedo-like openings, dotted/fine linear vessels, white structureless areas	Follicular plugs, perifollicular erythema, dull red background; no perifollicular white halo	Spiky keratotic projections on pink/red background, no white halos or vessels
Histopathology	Folliculotropic CD4+ T-cell infiltrates, epidermal sparing, perifollicular fibrosis	Psoriasiform hyperplasia, alternating ortho/parakeratosis, follicular plugging, sparse dermal inflammation	Follicular hyperkeratosis with minimal inflammation
Response to Topical Therapy	Poor; often refractory to corticosteroids	Moderate response to keratolytics and topical corticosteroids	Usually good response to emollients or keratolytics
Immunophenotype	CD4+ predominant; high CD4/CD8 ratio	No atypical lymphocytes; normal CD4/CD8 ratio	No atypia; normal T-cell profile
Key Diagnostic Clue	Perifollicular lymphoid infiltrate with epidermal sparing	Follicular plugs with diffuse erythema and scaling	Symmetrical, grouped follicular keratotic papules

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