Dermoscopy of folliculotropic mycosis fungoides of the scalp: a pediatric case report with comparative trichoscopic analysis

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Abstract

Folliculotropic mycosis fungoides (FMF) is an uncommon variant of mycosis fungoides (MF) that primarily affects hair follicles and often misdiagnosed due to its similarity with other scalp conditions. The median age at diagnosis is 60 years, while it is rare in childhood and adolescence. This case presents a 10-year-old boy with alopecic, indurated plaques on the scalp, unresponsive to antifungal therapy. Dermoscopy revealed key features, including perifollicular hyperkeratosis, broken hairs, irregularly distributed white halos, and features resembling inflammatory alopecias. Histopathology confirmed the FMF diagnosis. This case highlights the helpfulness of dermoscopy in FMF differential diagnosis, even in a rare pediatric case. It underscores the need for further studies to define dermoscopic aspects of FMF, ensuring more timely and effective treatment.

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Introduction

Folliculotropic mycosis fungoides (FMF) is a rare and aggressive variant of mycosis fungoides (MF) that selectively affects hair follicles rather than the epidermis.[1] The median age at diagnosis is 60 years, while it is rare in childhood and adolescence.[2] Scalp involvement often presents as indurated, alopecic plaques, which may be misdiagnosed as discoid lupus erythematosus (DLE), folliculitis decalvans (FD), lichen planopilaris (LPP), or tinea capitis.[3] While incisional biopsy and histological examination remain the gold standard for diagnosis, dermoscopy is emerging as a crucial tool to differentiate FMF from other inflammatory scalp disorders, guiding early biopsy and assuring appropriate treatment.[4]

Case Report

A 10-year-old boy presented with a three-year history of non-pruritic, alopecic plaques on the nuchal and right retroauricular scalp, gradually enlarging despite multiple topical and systemic antifungal therapies. The lesions were indurated, with perifollicular hyperkeratosis and mild erythema (Figure 1 A,B). The absence of pruritus, scarring, and anamnesis of recurrent pustular episodes reduced the likelihood of tinea capitis, LPP, or FD diagnosis, prompting further investigations.

Dermoscopic evaluation showed perifollicular hyperkeratosis with a patchy background erythema, broken hairs at various lengths, hair casts, scattered perifollicular pustules, irregularly distributed horn plugs, and white halos (Figure 2).

Given these atypical trichoscopic findings, a biopsy was performed at a site of perifollicular hyperkeratosis; histopathology showed dense perifollicular infiltration of atypical $CD4^+$ lymphocytes with folliculotropism and mucin deposition, establishing the diagnosis of FMF.[3]

The patient was then sent to the Santobono-Pausilipon Hospital and assessed for staging by the Pediatric Oncohematology Unit. Systemic involvement was ruled out through PET-CT imaging, ultrasound assessments, and comprehensive blood tests, including complete blood count and CD4 and CD8 T-cell counts. Given the exclusive cutaneous localization, the patient has been prescribed a therapy consisting of acitretin at 10 mg daily combined with narrow-band UVB phototherapy sessions three times weekly. The patient has been treated for two months so

far. Clinical and dermoscopic follow-up is performed every 15 days; preliminary observations suggest an initial trend toward reduction of skin inflammation and infiltration.

Discussion

Dermoscopy is increasingly recognized as an adjunctive tool in the diagnosis of cutaneous lymphomas affecting the scalp.[4] While some trichoscopic features overlap with other inflammatory alopecias,[1] specific distribution patterns and follicular alterations appear to be key diagnostic clues.

A recent case series of 18 patients with scalp FMF identified six main trichoscopic features: a decreased number of pilosebaceous units (single hair) (83.3%), dotted dilated vessels (77.8%), dystrophic/broken hairs (66.7%), vellus hairs (61.1%), sperm-like vessels (55.6%), and yellow dots (55.6%).[1] Additional findings included follicular openings dilation, scale-crust formations, purpuric dots, short hairs with double tips, tail-like hairs, perifollicular hyperkeratosis, milky-white globules, black dots, and white dots/lines, as well as follicular ostia absence.[1] The median age of the enrolled patients was 71 (range from 51 to 97 years),[1] including the most frequent age at diagnosis (60 years of age).[2]

In our case, dermoscopy revealed a unique combination of features (Figure 2): in fact, we observed perifollicular hyperkeratosis, broken hairs at various lengths, and irregularly distributed white halos, partially overlapping with the main features described in the case series. However, dotted dilated vessels, sperm-like vessels, and yellow dots were not evident in our patient. Moreover, trichoscopic features more common in inflammatory alopecias, like hair casts, horn plugs, perifollicular pustules, and a patchy background erythema, were present, although with peculiar characteristics: i)

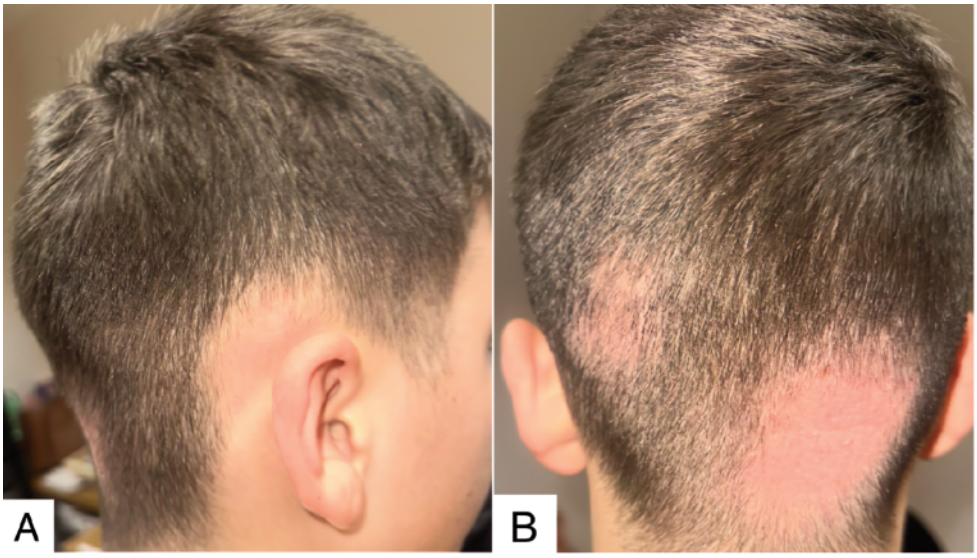

Figure 1. A, B) Clinical presentation at first evaluation: indurated lesions with perifollicular hyperkeratosis and mild erythema.

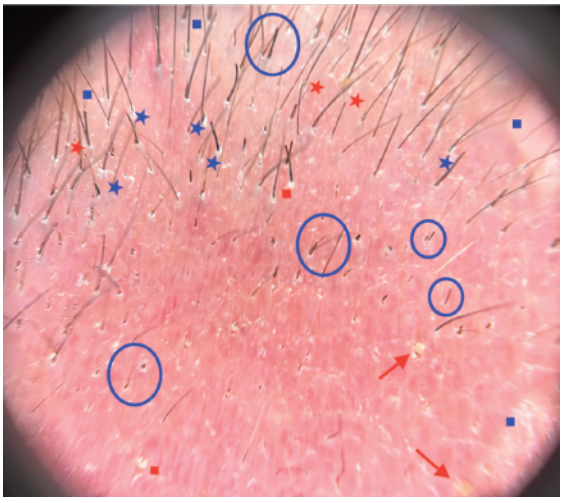

Figure 2. Dermoscopy: patchy background erythema along with perifollicular hyperkeratosis (blue star) + broken hairs at various lengths (blue circle) + irregularly distributed white halos (blue square); hair casts (red star) associated with perifollicular scaling + scattered horn plugs (red arrow) and perifollicular pustules (red square).

patchy background erythema, unlike LPP, was perifollicular and sharply demarcated; ii) hair casts, commonly seen in LPP, were associated with perifollicular scaling rather than perifollicular erythema; iii) horn plugs, resembling those seen in DLE, were irregularly distributed, whereas in DLE, they are more uniform and symmetrical; iv) in our case, perifollicular pustules, similar to FD, were scattered rather than confluent and not associated with tufted hairs. These particular dermoscopic aspects could possibly be due to an earlier disease stage or a different disease expression in pediatric patients. The comparison between our case and the 18-case series suggests that perifollicular hyperkeratosis, broken hairs at various lengths, and irregularly distributed white halos may be common features of FMF. Still, additional signs like sperm-like vessels, dilated vessels, and yellow dots could vary depending on disease progression and/or patient age. In addition, we observed that in FMF, inflammatory trichoscopic features like patch background erythema, perifollicular pustules, hair casts, and horn plugs, when present, appear to show distinctive features; searching for them could be an additional trichoscopic evaluation step. These findings emphasize the need for further studies to validate FMF-specific trichoscopic hallmarks in both adult and pediatric populations, possibly showing differences in populations and at different disease stages. Further studies are needed, especially in the pediatric population, given the rarity of the condition and the subsequent absence of dermoscopic literature regarding this age.

Conclusions

This case highlights distinct trichoscopic features of FMF that differentiate it from other inflammatory scalp conditions. Perifollicular hyperkeratosis, broken hairs at various lengths, and irregularly distributed white halos may serve as early clues to FMF, warranting biopsy. Additional trichoscopic features appear to be the ones more common in inflammatory alopecia, like hair casts, horn plugs, scattered perifollicular pustules, and a patchy background erythema, when presenting unusual distribution patterns. Recognizing these subtle trichoscopic patterns can facilitate earlier diagnosis and treatment, potentially improving patient outcomes. More extensive studies are needed to refine trichoscopic criteria for FMF and to possibly identify distinct patterns regarding patients' age, further validating its role in clinical practice.

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