Dermoscopic Findings of Pilotropic Mycosis Fungoid: A Rare Variant

Benahmed Jihane1*, Znati2, Meziane1, Ismaili1, Benzekri1 and Senouci1

1Dermatology Department, Ibn Sina hospital, University Mohammed V, Rabat, Morocco
2Pathology Department, Ibn Sina hospital, University Mohammed V, Rabat, Morocco

*Corresponding author: Benahmed Jihane, Dermatology Department, Ibn Sina hospital, University Mohammed V, Rabat, Morocco

Introduction

Pilotropic mycosis fungoid (PMF) is a rare variant of mycosis fungoides (MF) with distinctive clinicopathological features, accounting for 1 to 5%. It is characterized by a broad spectrum of clinical features and has a more aggressive clinical course than conventional MF [1]. At the onset of its progression, pilotropic mycosis fungoides is sometimes difficult to distinguish from classical mycosis fungoides with follicular involvement. PMF diagnosis is challenging and can mimic other conditions. Dermoscopy can be a valuable clue for the clinical diagnosis and differentiation with other inflammatory conditions. We report a case of follicutropic mycosis fungoid followed up in our department, with its dermoscopic findings, masquerading as chronic eczema.

Case Report

A 38-year-old patient, working as a poultry farmer, with no relevant medical history, presented to our department with a 5 year history of erthyematous plaques located on the head and scalp treated as chronic eczema with topical corticosteroid with good improvement.

Clinical examination revealed the presence of erythematous plaques on the head and scalp, follicular papules on the trunk and the presence of a tumor firm well circumscribed with a size of 7 × 5 cm located on the occipital region.

Dermoscopy showed white and hyperpigmented halo around the follicle, broken hairs, orange pink perifollicular clods and anisotrichia.

The patient was also complaining of chronic pruritus, recurrent abscesses, and folliculitis in the lower limbs over the past 2 years. There was no alopecia and no lymph nodes were found.

A biopsy was performed, showing a lymphocytic infiltrate with perifollicular accentuation. The immunohistochemical analysis was positive for CD3, CD5 and negative for CD7.

The diagnosis of pilotropic mycosis fungoid without mucinosis was made.

Investigations carried out including full blood count, chest X-ray, CT scan, renal and liver function tests were normal. Viral screens for HIV, hepatitis B and C antibodies were also negative. He was diagnosed as early stage disease (stage IIB).

The patient received methotrexate 25 mg per week with good evolution (Figure 1 and Figure 2).

Discussion

Mycosis fungoid occurred in adulthood; the mean age of onset varies from 46 to 59 years with a male preponderance [1].

The most common clinical findings associated with FMF are patches, plaques, and tumors mainly located on the head and neck as presented in our patient and it rarely affects the trunk and extremities. In adults, pruritus is a common symptom that is often difficult
of a lymphocytic infiltrate with perifollicular accentuation. The interfollicular epidermis is frequently spared. Pautrier’s microabscesses are rarely seen. Immunochemistry stained for CD2 and CD3. Antigen loss (for example CD2, CD3, CD4) has been described and may be useful for further diagnostic differentiation. Isolated cases of CD8-positive FMF have also been reported. In addition, there may be variable expression of CD30, which is associated with large-cell transformation in some cases [9,10].

Recent studies distinguished indolent (early-stage FMF) and more aggressive (advanced stage FMP) subgroups. Clinically, patients with early-stage skin-limited FMF present with localized or follicular papules often associated with hair loss, keratosis pilaris-like and acneiform lesions or plaques with histologically peri and intrafollicular infiltrates. The diagnosis is excellent in this group with a 5-year OS of 92-94%. The treatment is based on potent topical steroid, imiquimod in case
of limited follicle-based patches (T1), PUVA therapy is indicated in patients with plaques and patches [11].

In patients with advanced stage FMP with widespread thick plaques and/or tumours, prognosis is worse than epidermotropic variants. The treatment should combine a systemic and skin directed therapy. Several options may be considered: PUVA therapy combined with local radiotherapy for most infiltrated lesions and PUVA therapy combined with IFN-α or retinoids (Isotretinoine or bexarotene). In patients with widespread skin lesions we often prefer PUVA with additional low-dose radiotherapy for thick plaques or tumours [11].

Conclusion

Folliculotropic mycosis fungoides is considered as a rare variant of mycosis fungoides with a bad response to therapy and severe prognosis. Physicians should be aware of this ‘great imitator’ which can mimic inflammatory and infectious disorders. Dermoscopy can be a tool in the diagnosis. A biopsy guided by the dermoscopy should be performed to confirm the diagnosis at an early stage when a FMF is suspected.

References

1. Van Doorn R, Scheffer E, Willemze R (2002) Follicular mycosis fungoides, a distinct disease entity with or without associated follicular mucinosis. Arch Dermatol 138: 191-198.
2. Van Santen S, Roach RE, Van Doorn R, Horvath B, Brujin MS, et al. (2016) Clinical staging and prognostic factors in folliculotropic mycosis fungoides. JAMA Dermatol 152: 992-1000.
3. Baykal C, Atci T, Ozturk Sari S, Ekinci AP, Buyukbabani N, et al. (2017) Underrecognized clinical features of folliculotropic mycosis fungoides: A large clinical series. J Dtsch Dermatol Ges 15: 289-299.
4. Muniesa C, Estrach T, Pujol RM, Gallardo F, Garcia-Muret P, et al. (2010) Folliculotropic mycosis fungoides: Clinicopathological features and outcome in a series of 20 cases. J Am Acad Dermatol 62: 418-426.
5. Monopoli A, Annessi G, Lombardo G, Balliva G, Girolomoni G (2003) Purely follicular mycosis fungoides without mucinosis: Report of two cases with review of the literature. J Am Acad Dermatol 48: 448-452.
6. Gómez Diez S, Maldonado C, Fueyo A, Vazquez Lopez F, Fresno MF, et al. (2007) Folliculotropic mycosis fungoides: Study of four cases. Actas dermosifilogr 98: 486-490.
7. Slawinska M, Sobjanek M, Olszewska B, Nowicki R, Sokolowska-Wojdylo M (2018) Trichoscopic spectrum of folliculotropic mycosis fungoides. J Eur Acad Dermatol Venereol 32: e107-e108.
8. Geller S, Rishpon A, Myskowski PL (2019) Dermoscopy in folliculotropic mycosis fungoides- A possible mimicker of follicle-based inflammatory and infectious disorders. Journal of the American Academy of Dermatology 81: e75-e76.
9. Boone SL, Guitart J, Gerami P (2008) Follicular mycosis fungoides: A Histopathologic, immunohistochemical and genotypic review. G Ital Dermatol Venerol 143: 409-414.
10. Gerami P, Guitart J (2007) The spectrum of histophatologic and immunohistochemical findings in folliculotropic mycosis fungoide. Am J Surg Pathol 31: 1430-1438.
11. Van Santen S, Van Doorn R, Neelis KJ, Daniels LA, Horvath B, et al. (2017) Recommendations for treatment in folliculotropic mycosis fungoides: report of the Dutch Cutaneous Lymphoma Group. Br J Dermatol 177: 223-228.