OLGU SUNUMU CASE REPORT

DOI: 10.5336/dermato.2016-51209

Folliculotropic Mycosis Fungoides Associated with Alopecia in a Case

Alopesi ile Seyreden Folikülotropik Mikozis Fungoidesli Bir Olgu

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Geliş Tarihi/*Received:* 03.03.2016
Kabul Tarihi/*Accepted:* 04.11.2016

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ABSTRACT Mycosis fungoides (MF) is the most common type of cutaneous lymphoma and characterized by proliferation of small to medium T lymphocytes with cerebriform nuclei in most cases. However, the diagnosis of MF may be very difficult in certain cases, particularly in those variants of MF such assyringotropic MF, granulomatous MF and folliculotropic MF. The classic histopathologic feature is the presence of atypical T-cells with a tropism to hair follicle epithelium. The clinical presentation of folliculotropic MF often differs from the patches and plaques of classic MF and may be associated with decreased clinical suspicion for folliculotropic MF. The average time interval from onset of lesions till diagnosis of folliculotropic MF was 2 years. Folliculotropic MF displays resistance to standard treatment modalities, has an unfavourable course and diversity in the histological spectrum. Here we reported a rare case who presented to our dermatology polyclinic with a complaint of hair loss on his back firstly. On the follow-up, the case was diagnosed as folliculotropic MF and treated with interferon-α2a and PUVA successfully.

Keywords: Mycosis fungoides; alopecia areata; skin neoplasms

ÖZET Mikozis fungoides (MF); birçok olguda, küçük ve orta boyuttaki serebriform çekirdekçiklere sahip T lenfositlerinin çoğalmasıyla karakterize en sık olarak görülen kutanöz lenfoma tipidir. Bununla birlikte; özellikle asiringotropik, granülomatöz ve follikülotropik MF gibi olgularda tanı koymak güç olabilmektedir. Folikülotropik MF'de klasik histopatolojik görünüm; atipik T hücrelerinin folliküler epitele tropizmi ile karakterizedir. Folikülotropik MF'in klinik görünümü özellikle klasik MF'de görülen yama ve plaklardan farklılık arz etmektedir ve bu durum da klinik olarak folikülotropik MF öntanısını güçleştirmektedir. Hastalığın başladığı dönemle tanı arasındaki süre ortalama 2 yıldır. Folikülotropik MF, standart tedavi seçeneklerine dirençlidir, istenmeyen klinik gidişata sahiptir ve histopatolojik olarak farklılıklar arz etmektedir. Biz burada; sırtta kıl dökülen alanlar şikayeti ile polikliniğimize başvuran, takiplerinde Folikülotropik MF gelişen ve PUVA ve interferon- α2a ile başarıyla tedavi edilen oldukça nadir görülen bir olguyu sunduk.

Anahtar Kelimeler: Mikozis fungoides; alopesi areata; deri neoplazileri

Mycosis fungoides (MF) is the most common type of cutaneous T-cell lymphoma and characterized by proliferation of small to medium T lymphocytes with cerebriform nuclei in most cases. However, the diagnosis of MF may be very difficult in certain cases, particularly in those variants of MF such assyringotropic MF, granulomatous MF and folliculotropic MF.[1] Folliculotropic MF is a rare variant, which histopathologically is characterized by pronounced folliculotropism of neoplastic T cells, with or without follicular mucinosis, and clinically by an im-

paired prognosis compared to classic MF. The clinical presentation of folliculotropic MF often differs from the patches and plaques of classic MF and may be associated with decreased clinical suspicion for MF.[2] Here we reported a rare folliculotropic MF case and treated with interferon (IFN)-α2a and psoralen and ultraviolet A (PUVA) successfully.

CASE REPORT

In 2011, a 30 year-old-male presented to our tertiary dermatology unit with a 1-week history of alopesic patch of the right arm and right side of lower scapular region. When the alopesic lesion first appeared, the surface of the lesion was smooth and slightly red skin color without any skin alterations like atrophic, scaling and follicular changes. The patient was diagnosed as having alopecia areata (AA). Treatment with topical/intralesional steroid was attempted for 3 months but without success. Then slightly erythematous atrophic scale appeared within alopesic sites and we performed skin punch biopsy from the lower scapular region (Figure 1). The histopathologic examination revealed a marked and predominant involvement of hair follicles, mostly consisting on perifollicular and intrafollicular infiltration by small and medium atypical T lymphocytes (Figures 2, 3). The histopathological findings were consistent with folliculotropic MF. Within a few weeks, new alopesic patches and MF lesions apperared on body and extremities. Our patient was administered both with PUVA therapy and IFN- α2a 3x 10^6 units thrice every week for 1 year. After the clinical and histopathological improvement were achieved at 12^{th} month of treatment, IFN-α2a was stopped and PUVA therapy was then tapered gradually a period of 1 year. There was no recurrence in follow-up for 1 year. Neverthless, no hair regrowth was observed within folliculotropic MF plaques during treatment and follow-up period.

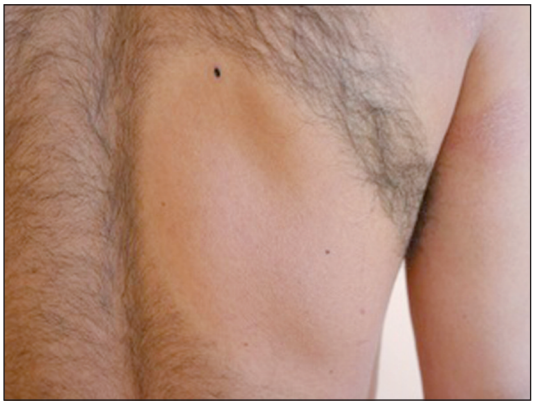

FIGURE 1: Erythematous slightly atrophic scale within alopesic site and erythematous atrophic macule on the right arm.

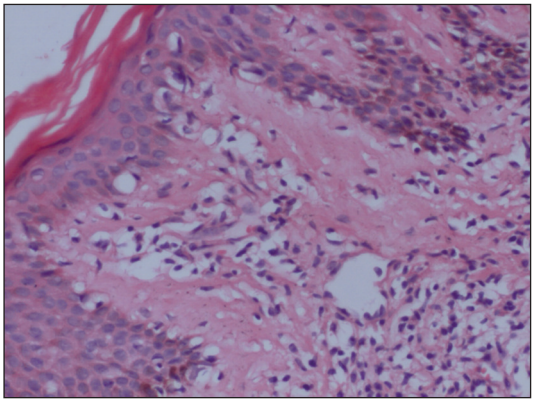

FIGURE 2: Atypical T lymphocytes in upper dermis and squamous epithelium (HE, x40).

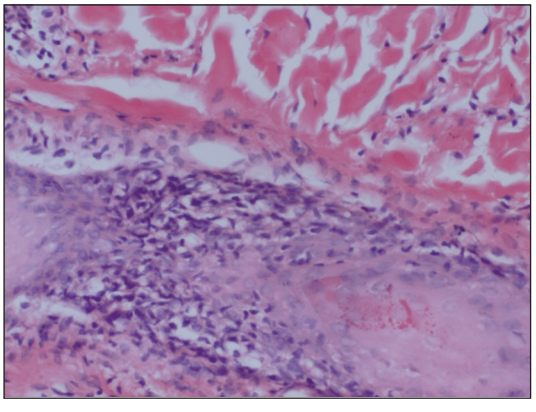

FIGURE 3: Perifollicular and intrafollicular infiltration by small and medium atypical T lymphocytes (HE, x20).

DISCUSSION

Folliculotropic MF has a broad clinical spectrum with acneiform lesions, comedones, plaques with follicular papules, alopecia, cysts and nodules, frequently on head and neck areas. Other important observations in folliculotropic MF are the occasional presence of peculiar lesions such as excoriated nodules, xanthomatous changes, or pustules which are very uncommonly found in conventional MF forms. The extent of lesions can be quite

variable.[3] Folliculotropic MF occurs mostly in adults with a male predominance, but it has also been reported in children and adolescents.[4]

The diverse cutaneous involvement delays correct diagnosis in folliculotropic MF.[5] The average time interval from onset of lesion stil diagnosis of folliculotropic MF is 2 years.[6] In our patient, the time interval was 3 months. Among the diverse cutaneous involvements, alopecia is tended to occur after the onset folliculotropic MF symptoms.[7] In the literature, there is only 1 case report include folliculotropic MF mimicking alopecia areata.[8] Contrary to limited alopesic patch in the latter case, alopesic patches and atrophic erythematous plaques were more and spreaded to other body areas very quickly in our case. In a previous study, 1550 MF patients were reviewed and only 5 patients had documented alopecia before or within 1 year of onset of skin symtoms. And additionally, it is stated that alopecia within MF lesions mostly develop more than 1 year after the onset of skin symptoms.[7] It may be difficult at least initially to distinguish alopecia areata, follicotropic MF, as in our case. Whether or not our patient first had alopecia areata and then developed folliculotropic MF later or whether this was initially undiagnosed folliculotropic MF is uncertain. It is proposed that folliculotropic MF should be kept in mind especially for patients who do not respond to alopecia area treatment.[6]

PUVA, local radiotherapy, bexarotene, total skin electronbeam, topical steroids, surgery, prednisone, chlorambucil, IFN-α2a are used in the treatment of folliculotropic MF. When IFN-α2a is used in combination with PUVA, both response and response duration are reported to be improved, with recent studies reporting overall response and complete response rates of 98% and 84%, respectively.[9] Our patient responded to PUVA and IFN-α2a treatment. Folliculotropic MF lesions were cleared and no recurrence were seen in the follow-up period. Nevertheless, no hair regrowth was observed within folliculotropic MF plaques during treatment and follow-up period.

In conclusion; this type of folliculotropic MF onset led to late diagnosis in regards of atypical localisation. The lesson to draw from this case is that folliculotropic MF is a great mimicker and may initially resemble alopecia areata. If AA resist to conventional treatments and erythematous papules and/or atrophy appear within alopesic lesions, folliculotropic MF should be kept in mind.

Conflict of Interest

Authors declared no conflict of interest or financial support.

Authorship Contributions

Idea/Concept: Constructing the hypothesis or idea of research and/or article: *Ali Balevi;* ***Design: Planning methodology to reach the conclusions:*** *Ali Balevi, Pelin Üstüner;* ***Control/Supervision: Organizing, supervising the course of progress and taking the responsibility of the research/study:*** *Mustafa Özdemir;* ***Data Collection and/or Processing: Taking responsibility in patient follow up, collection of relevant biological materials, data management and reporting, execution of the experiments:*** *Ali Balevi, Pelin Üstüner, Hatice Toy;* ***Analysis and/or Interpretation: Taking responsibility in logical interpretation and conclusion of the results:*** *Ali Balevi;* ***Literature Review: Taking responsibility in necessary literature review for the study:*** *Ali Balevi;* ***Writing the Article: Taking responsibility in the writing of the whole or important parts of the study:*** *Ali Balevi, Pelin Üstüner, Mustafa Özdemir, Hatice Toy.*

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