Annals of Clinical Case Reports

Case Report
Published: 03 Jan, 2024

The Weirdest Presentation of CTCL (Cutaneous T-Cell Lymphoma) of All Time: A Widespread Comedonal Eruption in Association with Kaposi Sarcoma

*Balighi K[1], Lajevardi V[1], Tootoonchi N[1], Ghanadan A[1], Sharifi M[1], Gholibeigian Z[1] and Akhdar M[1,2]**

[1]Department of Dermatology, Razi Hospital, Tehran University of Medical Sciences, Iran

[2]Department of Dermatology, Nabatieh Governmental University Hospital, Lebanon

Abstract

Background: Folliculotropic Mycosis Fungoides (FMF) is a rare and distinctive variant of MF (Mycosis Fungoides) which is found in approximately 10% of MF patients, and those patients can present with folliculotropic lesions such as comedones, cysts and acneiform lesions.

Methods: Histopathologic examination and immunohistochemistry staining were done for different skin biopsies. Flow cytometry and TCR gene rearrangement were also carried out.

Results: H&E and IHC along with the clinicopathologic algorithm for the diagnosis of early MF were in favor of FMF.

Conclusion: Early diagnosis can be critical for patients with FMF and clinicians should consider such rare presentations and consider folliculotropic MF as a differential diagnosis.

Introduction

Cutaneous T-Cell Lymphoma (CTCL) is a T-cell- homing skin malignancy that has a wide range of clinical presentations, histopathologic features, prognosis and immunophenotypes. Over the years, Mycosis Fungoides and Sezary syndrome were the only known forms of CTCL, however, in recent years, other new forms of CTCL have been identified. The most common type of CTCL is Mycosis Fungoides accounting for about 50% of all the cases [1]. Folliculotropic Mycosis Fungoides (FMF) is a rare and distinctive variant of MF which is found in approximately 10% of patients [2]. Those patients can present with folliculotropic lesions such as comedones, cysts and acneiform lesions [1]. To our best knowledge the comedonal presentation is extremely rare, and to date, there is no such case reported with extensive comedonal eruptions. Hence, a CTCL case is introduced here, presenting with extensive widespread comedonal lesions all over the patient's body.

Case Presentation

A 65-year-old male patient who had lichen planopilaris-like lesions in the past 2 years ago, presented with giant milia lesions on the head and neck, predominantly on the periauricular area, and wide-spread comedonal lesions extensively distributed on the head (temporal area), upper back, lateral arms, buttocks, posterior thighs, axilla, and inguinal areas (Figure 1a, 1b). The onset of these lesions was 15 years ago. The patient also had scarring alopecia on the scalp, and papules with a violaceous hue on his legs (Figure 2a). The lesions were occasionally pruritic. On the Trunk and upper extremities, areas of erythema and papulosquamous lesions were noticed later on (Figure 2b). The patient didn't have any history of contact with chemicals or other materials that may induce chloracne. His habitual history included smoking and opium use.

The patient underwent assessment 2 years ago; 3 biopsy specimens were taken from different sites, where lichen planopilaris was reported for the scalp and face specimens for which he was treated with oral Hydroxychloroquine (200 mg daily) and Clobetasol lotion, after which the hair loss was partially improved. During this time, milia and comedones lesions kept on erupting. The patient was first treated with oral Isotretinoin 20 mg every other day, topical Benzoyl Peroxide 5% and had several sessions of comedones extractions but there was no significant improvement. The patient was then recommended to do biopsies from different body sites. The biopsy specimen of the neck was reported as milia formation, where on histology there were sections with milia-like structures in superficial

OPEN ACCESS

Correspondence:
Marwa Akhdar, Department of Dermatology, Nabtieh Governmental University Hospital, Nabatieh, Lebanon, Tel: 009617510362

Received Date: *14 Dec 2023*
Accepted Date: *29 Dec 2023*
Published Date: *03 Jan 2024*

Citation:
Balighi K, Lajevardi V, Tootoonchi N, Ghanadan A, Sharifi M, Gholibeigian Z, et al. The Weirdest Presentation of CTCL (Cutaneous T-Cell Lymphoma) of All Time: A Widespread Comedonal Eruption in Association with Kaposi Sarcoma. Ann Clin Case Rep. 2024; 9: 2551.

ISSN: 2474-1655.

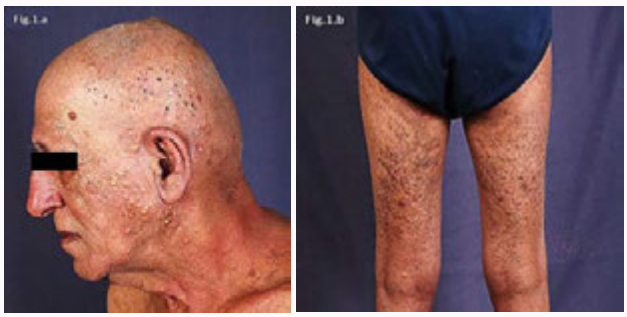

Figure 1a: Giant milia lesions on the head and neck, predominantly on the periauricular area, and wide-spread comedonal lesions extensively distributed on the head. **Figure 1b:** Extensive comedonal and milia lesions on the posterior thighs.

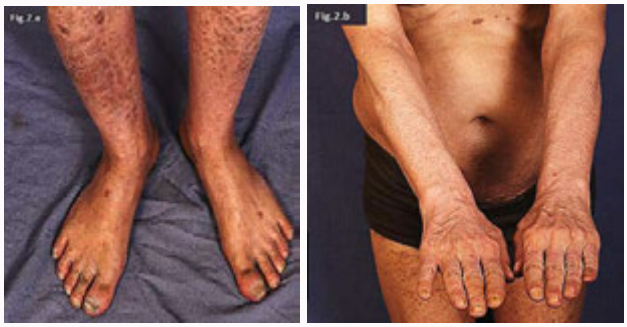

Figure 2a: Papules with a violaceous hue on the legs. **Figure 2b:** Erythema and papulosquamous lesions on the trunk and upper extremities.

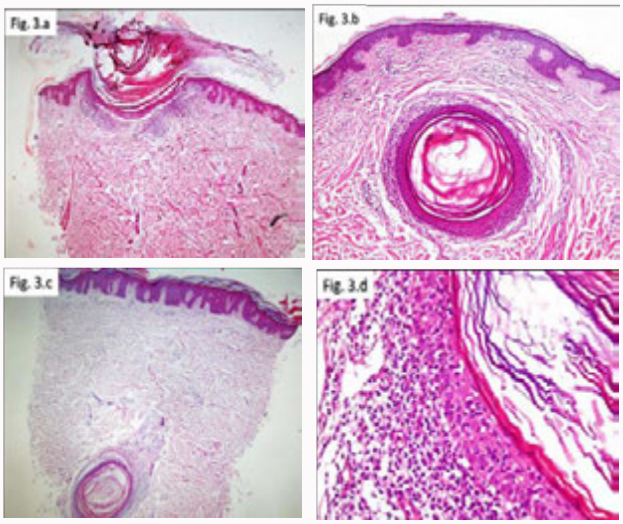

Figure 3a-3d: Follicular structures with dilated follicular infundibulum (a), isthmus (b) and lower part of follicle (c) surrounded by many lymphocytes with permeation into follicular epithelium (d). (H&E, original magnification x40, x100, x40, x400).

dermis lined by thin and flat epithelial cells in conjunction with eccrine glands surrounded by some lymphocytes. The other site was reported as follicular lichen planus with an interface lichenoid reaction pattern with follicular involvement, so the patient was treated with topical emollients and corticosteroids. On the third visit, there was no much improvement, however the erythematous and papulosquamous background of the lesions were getting more prominent. Therefore, the patient underwent a biopsy once more with probable differential

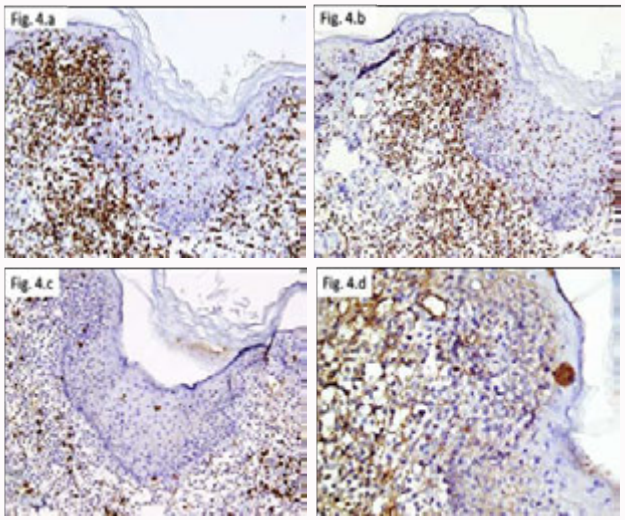

Figure 4a-4d: Immunohistochemistry staining reveals positive immunoreaction for CD3 (a), CD4 (b), and few CD8+ lymphocytes (c) in epidermis and follicular structures, in conjunction with negative immunoreaction for CD 7 (d) and CD5 (not show).

diagnosis were mycosis fungoides, chloracne and lichen planus were. In addition, some new red papules had been found on the feet and legs of the patient, which suggested the diagnosis of Kaposi's sarcoma. The first reports of the neck, buttock and back biopsies were compatible with chloracne, where a mild psoriasiform reaction was seen with cysts and dilated atrophic milia-like follicular structures. The report of leg biopsy was compatible with Kaposi sarcoma, where microscopic description showed a focus of infiltrative proliferation of spindle endothelial cells with extensive compressed slit-like vascular channels dissecting through dermal collagen with extravasation of red blood cells. After further investigation, Immunohistochemistry staining revealed positive immunoreaction for CD3 and CD4 in most of intraepidermal lymphocytes and few CD8+ lymphocytes in epidermis. No immunoreaction for CD7 and CD5 was identified (Figure 3a-3d). And after reconsidering the biopsy specimens, little foci of epidermotropism and folliculotropic lymphocytic infiltration were seen. There were also some follicles with dilated canal and keratin-filled infundibular plugging, resembling milia-like structures surrounded by some lymphocytes with permeation into basal layer and collection into follicular infundibulum (Figure 4a-4d).

The patient's laboratory data were otherwise normal except for leukocytosis with white blood cell count 11,900/mm^3 and triglyceride level was 251 mg/dl. All viral markers were negative.

The final report after taking into consideration the patient's clinical features and the IHC results concluded that the diagnosis was most compatible with cutaneous T cell lymphoma (Mycosis Fungoides). TCR gene rearrangement was also done but the result showed a polyclonal pattern. Flow cytometry showed higher CD4 count (46.3%) than CD8 count (14.6%).

Discussion

Mycosis Fungoides is known to be the most common type of cutaneous T-cell lymphoma, where the Folliculotropic variant (FMF) as a rare variant, represents less than 10% of the mycosis fungoides cases [2]. Patients of this variant may commonly present with patches and plaques associated with follicular prominence and alopecia, and rarely with comedo-like lesion, nodules and cysts [3]. Acneiform

Table 1: Clinicopathologic algorithm for the diagnosis of early MF.

Criteria	Major (2 points)	Minor (1 point each)
Clinical Persistent and/or progressive patches/thin plaques plus	Any two	Any one
1. Non-sun-exposed location		
2. Size/shape variation		
3. Poikiloderma		
Histopathologic Superficial lymphoid infiltrate plus	Both	Either
1. Epidermotropism		
2. Atypia		
Molecular/biological	—	Any
Clonal TCR gene rearrangement		
Immunopathologic	—	Any one
1. 0<50% $CD2^+$,3^+,5^+ T cells		
2. <10% $CD7^+$ T cells		
3. Epidermal/dermal discordance of CD2, CD3, CD5 or CD7		

A total of 4 points is required for the diagnosis of mycosis fungoides based on any combination of points from the clinical, histopathologic, molecular, and immunopathologic criteria. Adapted from Pimpinelli N, Olsen EA, Santucci M, et al. Defining early mycosis fungoides. J Am Acad Dermatol. 2005;53:1054.

lesions (comedones, cysts) are most pronounced in the head and neck area [2].

Generally, the comedo-like and milia-like presentation of FMF is very rare, and when presented, it is usually localized. Nikoo et al. in the year 2016 reported a 37-year-old patient which presented with erythematous patches and plaques on his trunk and followed by a crusted nodule with comedonal changes just on his forehead [4]. Shahriari et al. in the year 2016 also reported a 50-year-old patient with FMF which presented acneiform lesions only on the face, postauricular scalp and chest [5]. Fraser-Andrews et al. in the year 1999 reported a 45-year-old patient with FMF who presented with comedo- and cyst-like lesions on the face, trunk and limbs [6]. Furthermore, in the textbook of Skin Lymphoma [7] it has been mentioned that there's a rare presentation of MF with a widespread eruption of infundibular cysts and/or comedones, as in our case.

The immunohistochemistry study of Folliculotropic MF shows that neoplastic T cells have CD3+, CD4+ and CD8- (as in classical MF), and an admixture of CD30+ blast cells [1]. The same was seen in the present case, however CD8+ cells were very few. According to the article published by Olsen in 2015 [8], a clinicopathologic algorithm for the diagnosis of MF was introduced [adapted from Pimpinelli et al. [9] (Table 1), according to which our patient scored 4 (Persistent and progressive patches/thin plaques in non-sun exposed locations with variation in size and shape-superficial lymphoid infiltrate and epidermotropism without spongiosis- <10% CD7 + T cells) and the diagnosis of MF was confirmed.

What made our case unique is the extensive and wide distribution of the comedo-like and milia-like lesions all over the body, which was later on followed by the onset of erythematous and papulosquamous and scaly lesions on the upper extremities, and what made it even more unique is its association with Kaposi sarcoma. To date, there is not a single case of FMF in literature reported with such an extensively and widely distributed comedo-like lesions. However, the association of Kaposi's sarcoma and MF is very rare and its reason has not been well established yet, and very few cases have been reported to date. In the year 2016, Bariani et al. reported a case of Mycosis Fungoides and Kaposi's sarcoma in an HIV-negative patient, the patient was a 53-year-old male who developed Kaposi's sarcoma nodular lesions on his limbs (HHV-8 positive) after 3 years of diagnosing MF and 2 years of treatment with 138 NB-UVB phototherapy sessions [10]. It has been postulated that impaired cell-mediated immunity by T-cell activation in MF could contribute to the development of other neoplasms [11,12]. Apparently, a study done by Rinne et al. in 2019 succeeded to link the association of MF and Kaposi's Sarcoma to genetic predispositions [13], and another study by Emadi et al. in the year 2012 found this association to be due to an exposure to an environmental factor which was sulfur mustard gas [14].

This kind of presentation of folliculotropic MF can be challenging because in most of the cases when the comedo-like lesions first appear, there is no presence of any erythematous scaly patches, and this can be misleading and cause a delay in the diagnosis and treatment. Recent studies have shown that the 5-year survival rate was 70% to 80% in patients with FMF, which was similar to that of classical tumor-stage mycosis fungoides, but worse than the classical plaque stage [1,2,15].

Previous studies have shown that FMF is less responsive to some skin-directed therapies such as PUVA and topical nitrogen mustard, and due to its aggressive clinical course, that is similar to that of tumor-stage classic MF, it should be treated accordingly, and in such cases total skin electron beam irradiation can be an effective treatment [2]. Alternatively, PUVA combined with retinoids or interferon alpha may be considered, whereas persistent tumors can be effectively treated with local radiotherapy [1].

As a conclusion, early diagnosis can be critical, and this case report highlights the clinical features that might help clinicians to consider folliculotropic MF as a differential diagnosis.

References

1. Willemze R, Jaffe ES, Burg G, Cerroni L, Berti E, Swerdlow SH, et al. WHO-EORTC classification for cutaneous lymphomas. Blood. 2005;105(10):3768-85.

2. van Doorn R, Van Haselen CW, van Voorst Vader PC, Geerts ML, Heule F, de Rie M, et al. Mycosis fungoides: Disease evolution and prognosis of 309 Dutch patients. Arch Dermatol. 2000;136(4):504-10.

3. V Vergote PW, Hauben E, Busschots AM. Folliculotropic mycosis

fungoides: A clinicopathological study of a series of 10 cases. J Dermatol Res Ther. 2016;2(2).

4. Azita Nikoo MR. Folliculotropic mycosis fungoides with comedonal-like appearances. Iran Red Crescent Med J. 2016;18(12):e16542.
5. Neda Shahriari MS, Katalin Ferenczi. A case of folliculotropic mycosis fungoides masquerading as acne. J Am Acad Dermatol. 2016;74(5): AB176.
6. Fraser-Andrews E, Ashton R, Russell-Jones R. Pilotropic mycosis fungoides presenting with multiple cysts, comedones and alopecia. Br J Dermatol. 1999;140(1):141-4.
7. Cerroni L. Skin Lymphoma-The Illustrated Guide. 2020.
8. Olsen EA. Evaluation, diagnosis, and staging of cutaneous lymphoma. Dermatol Clin. 2015;33(4):643-54.
9. Pimpinelli N, Olsen EA, Santucci M, Vonderheid E, Haeffner AC, Stevens S, et al. Defining early mycosis fungoides. J Am Acad Dermatol. 2005;53(6):1053-63.
10. Bariani MC, Fleury LFJ, Ribeiro AM, Carneiro SS, Pereira TA. Mycosis fungoides and Kaposi's sarcoma association in an HIV-negative patient. An Bras Dermatol. 2016;91(5 suppl 1):108-10.
11. Väkevä L, Pukkala E, Ranki A. Increased risk of secondary cancers in patients with primary cutaneous T cell lymphoma. J Invest Dermatol. 2000;115(1):62-5.
12. Brownell I, Etzel CJ, Yang DJ, Taylor SH, Duvic M. Increased malignancy risk in the cutaneous T-cell lymphoma patient population. Clin Lymphoma Myeloma. 2008;8(2):100-5.
13. Rinne SJ, Sipilä LJ, Sulo P, Jouanguy E, Béziat V, Abel L, et al. Candidate predisposition variants in kaposi sarcoma as detected by whole-genome sequencing. Open Forum Infect Dis. 2019;6(10):ofz337.
14. Emadi SN, Babamahmoodi F, Poursaleh Z, Sayad-Noori SS, Soroush MR, Maleki AR, et al. Sézary syndrome, Kaposi sarcoma and generalized dermatophytosis 15 years after sulfur mustard gas exposure. J Dermatol Case Rep. 2012;6(3):86-9.
15. Bonta MD, Tannous ZS, Demierre MF, Gonzalez E, Harris NL, Duncan LM. Rapidly progressing mycosis fungoides presenting as follicular mucinosis. J Am Acad Dermatol. 2000;43(4):635-40.