A Rare Case of Poikilodermatous Mycosis Fungoides

Preema Sinha, Durga Madhab Tripathy, Divya Shelly, Shekhar Neema

Abstract
Poikilodermatous mycosis fungoides (PMF) is a rare clinical variant of early-stage mycosis fungoides with peculiar histological features and with low risk of disease progression. Since poikilodermia can coexist with classical mycosis fungoides lesions, PMF can only be considered when poikilodermatous lesions are predominant (>50% of lesions). We here report one such rare case of PMF with poikilodermatous lesions covering almost 70% of the body surface and with characteristic clinical, histopathological, dermoscopic, and immunohistochemical findings.

Key Words: Dermoscopic changes, mycosis fungoides, poikilodermatous mycosis fungoides

Introduction
Cutaneous lymphoma can be broadly divided into T- and B-cell lymphomas depending on the nature of the neoplastic cells. The most common variant of cutaneous T-cell lymphoma which represents approximately 80% of total cutaneous lymphomas is mycosis fungoides (MF) which sometimes is also referred to as “Alibert-Bazin’s disease.”[1] It presents as erythematous patches and plaques with or without scaling. There are numerous variants of MF, among which poikilodermatous MF is a rare variant constituting 1%-2% of the total cases.[2] Herein, we report one such rare case.

Case History
A 56-year-old male symptomatic since past 5 years in the form of generalized burning sensation over the body with multiple mildly pruritic dark-colored lesions over the trunk and lower extremities presented to us. There was no history of any fever, weight loss, night sweat, joint pain, photosensitivity, any known triggering factor, or significant family history.

The general and systemic examinations were unremarkable without any significant lymphadenopathy. Dermatological examination revealed diffuse poikilodermia over the trunk involving abdominal folds and upper limbs along with multiple discrete violaceous papules over the thighs and gluteal region involving nearly 70% of body surface area [Figures 1 and 2]. Dermoscopic examination revealed reticular brown pigment pattern on a pink white background. Unevenly distributed grey dots were present throughout the pigment pattern. Linear and glomerular vessels were also visualized [Figure 3].

All routine hematological and biochemical parameters were normal. Antinuclear antigen panel for ruling out connective tissue disorders was also negative. Computed tomography scans of the thorax, abdomen, and pelvis, as well as aspiration of the sternal bone marrow for cytological assessment were performed to determine the stage of the disease. Two skin biopsies performed from the right thigh and back revealed atrophy of epidermis along with focal hyperkeratosis and parakeratosis especially over the tissue obtained from the violaceous papules over the right thigh. A band-like inflammatory infiltrate was observed in the dermoepidermal junction [Figure 4]. Focal epidermotropism of lymphocytes with hyperchromatic nuclei and irregular nuclear membranes was noted. No Pautrier’s microabscesses were seen, but pigmentary incontinence was noted. Immunohistochemistry showed an overwhelming population of CD3 T cells along with the focal loss of CD7, restriction of CD4, and no loss of CD6 [Figure 5a-c]. Overall, the findings were consistent with poikilodermatous mycosis fungoides (PMF).

The patient was managed as a case of Stage T2N0M0B0 MF with ultraviolet (UV) A therapy to which he had shown a good response with resolution of the lesions to a great extent. Till the time of reporting, the patient was under regular follow-up.

Access this article online
Quick Response Code:
Website: www.e-ijd.org
DOI: 10.4103/ijd.IJD_145_19

How to cite this article: Sinha P, Tripathy DM, Shelly D, Neema S. A rare case of poikilodermatous mycosis fungoides. Indian J Dermatol 2020;65:417-9.
Received: February, 2019. Accepted: March, 2019.

This is an open access journal, and articles are distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 4.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as appropriate credit is given and the new creations are licensed under the identical terms.

For reprints contact: reprints@medknow.com
Sinha, et al.: A rare case of poikilodermatous mycosis fungoides

Discussion

Initially described as poikiloderma vasculare atrophicans, PMF is a rare clinical variant of cutaneous T-cell lymphoma. It was first described by an American pediatrician Abraham Jacoby in 1908 and also referred at times as lichenoid variant of MF. Initially considered as a variant of large plaque parapsoriasis, it was previously called “parapsoriasis variegata,” but now the terminology has become obsolete and the entity is referred to as an early stage of MF.[1]

In spite of remarkable similarity in histopathological and immunohistochemical findings of poikilodermatous MF with classical MF, clinical presentations vary. The patches show features of poikiloderma in the form of both hypopigmentation and hyperpigmentation along with atrophy and telangiectasia. The patient usually complains of burning sensation over the body rather than frank pruritus in contrast to classical MF which is an extremely pruritic condition. Various other poikilodermatous conditions, such as lupus erythematosus, dermatomyositis, and poikiloderma of Civatte affect predominantly the photo-exposed areas in contrast to poikilodermatous MF which has a bathing-suit distribution over the trunk involving the folds as was seen in our patient.[2]

PMF has a predilection to affect the younger age group and has a better prognosis with good response to therapy as opposed to classical MF.[3]

Histopathologically, there is evidence of epidermotropism along with atypical T-cell infiltrate in the papillary dermis. Pautrier’s microabscesses, which are very specific for classical MF, are rarely seen in the poikilodermatous variant. Pigmentary incontinence along with a band-like inflammatory infiltrate is usually observed in the dermoeidermal junction in poikilodermatous MF. Cytotoxic T-cell preponderance has been mentioned in literature to be commonly

Figure 1: Diffuse poikiloderma noted over the trunk

Figure 2: Multiple violaceous papules noted on a background of diffuse poikiloderma over both thighs

Figure 3: Dermoscopic examination revealed reticular brown pigment pattern on a pink white background. Unevenly distributed grey dots are present throughout the pigment pattern. Linear and glomerular vessels can also be visualized (3Gen DermLite DL4 polarized dermatoscope)

Figure 4: Mildly atrophic epidermis with a band-like inflammatory infiltrate and melanin incontinence at places (H and E, ×100)
Sinha, et al.: A rare case of poikilodermatous mycosis fungoides

Indian Journal of Dermatology | Volume 65 | Issue 5 | September-October 2020

419

associates with poikilodermatous MF. Presence of epidermotropism and characteristic immunohistochemical findings differentiates it from other poikilodermatous conditions. Moreover, the absence of spongiosis and lack of mucin/elastin in the dermis are characteristics of this variant of cutaneous lymphoma.\(^3\,4\)

Xu and Tan found dermoscopic features in the form of multiple polygonal structures consisting of the lobules of white storiform streaks with septa of pigmented dots.\(^5\)

However, we found a reticular brown pigment pattern on a pink white background with unevenly distributed grey dots present throughout the pigment pattern.

Treatment modalities are similar to classical MF and include skin-directed and systemic therapy based on the stage of the disease. Skin-directed therapies include topical corticosteroids, bexarotene, carmustine, mechlorethamine, narrowband ultraviolet B (UVB), PUVA and radiation. New treatment options for MF have recently expanded to include UVA1 and excimer laser. Topical corticosteroids are usually ineffective especially in cases of diffuse cutaneous involvement. The skin-directed therapy-resistant disease can be managed with oral retinoids (acitretin, isotretinoin, and bexarotene) and interferon alpha.\(^6\,8\) Novel systemic agents in the form alemtuzumab (anti-CD 52 antibodies), interferon-12, and histone deacetylase inhibitors are restricted to tumor stage or visceral/nodal involvement in the disease.\(^6\,8\)

Conclusion

PMF is a unique clinical entity with a different presentation in comparison to classical MF with an excellent prognosis in the form of slow progression of the disease and good response to both skin-directed and systemic agents with a period of remission extending up to 10 years. The dermoscopic examination also aids in quick clinical diagnosis.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Pankratov O, Gradova S, Tarasevich S, Pankratov V. Poikilodermatous mycosis fungoides – Clinical and histopathological analysis of a case and literature review. Acta Dermatovenereol Alp Pannonica Adriat 2015;24:37-41.
2. Mataix J, Bañuls J, Lucas A, Belinchón I, Betlloch I. Poikilodermatous mycosis fungoides. Int J Dermatol 2007;46:950-1.
3. Farley-Loftus R, Mandal R, Latkowski JA. Poikilodermatous mycosis fungoides. Dermatol Online J 2010;16:8.
4. Bloom B, Marchbein S, Fischer M, Kamino H, Patel R, Latkowski JA. Poikilodermatous mycosis fungoides. Dermatol Online J 2012;18:4.
5. Xu P, Tan C. Dermoscopy of poikilodermatous mycosis fungoides (MF). J Am Acad Dermatol 2016;74:45-7.
6. Lindae ML, Abel EA, Hoppe RT, Wood GS. Poikilodermatous mycosis fungoides and atrophic large-plaque parapsoriasis exhibit similar abnormalities of T-cell antigen expression. Arch Dermatol 1988;124:366-72.
7. Wain EM, Orchard GE, Blattler SJ, Spittle M, Sc MF, Russell-Jones R. Outcome in 34 patients with juvenile-onset mycosis fungoides: A clinical, immunophenotypic, and molecular study. Cancer 2003;98:2282-90.
8. Olsen EA, Hodak E, Anderson T, Carter JB, Henderson M, Cooper K, et al. Guidelines for phototherapy of mycosis fungoides and Sézary syndrome: A consensus statement of the United States Cutaneous Lymphoma Consortium. J Am Acad Dermatol 2015;74:27-58.