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Poikilodermatous Mycosis Fungoides

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Abstract
Poikilodermatous mycosis fungoides is one of the rare clinical variant of mycosis fungoides with clinical features of poikiloderma.
The variability of atypical and subtle clinical features of mycosis fungoides may delay the diagnosis. Therefore high index of suspicion and familiarity with atypical variants is helpful for early diagnosis and treatment.
We hereby report a typical case of 65 years old male presented with asymptomatic, mottled hyper and hypo-pigmentation of skin with interspread telangiectases and areas of atrophy on the trunk and extremities since 10 years. Based on the clinical manifestations and histological findings, the diagnosis of poikilodermatous mycosis fungoides was made.

Introduction

Mycosis fungoides (MF) is the most common forms of cutaneous T-cell lymphoma
Among different variants of mycosis fungoides, poikilodermatous is a rare distinct clinical variant of cutaneous T-cell lymphoma, formerly referred to as poikiloderma vasculare atrophicans.[1]
Poikilodermatous MF is characterized clinically by localized or diffuse patches consisting of telangiectasias, mottled hyper- and hypo-pigmentation, and atrophy.
The lesions are usually asymptomatic, however some patient may develop mild pruritis.
Present predominantly in males, poikilodermatous MF has a good prognosis.[2]
The lesions are usually asymptomatic, however some patient may develop mild pruritis.
Histopathologic features of poikilodermatous MF show an atypical T-cell infiltrate in the papillary dermis with evidence of epidermotropism, epidermal atrophy, dilated blood vessels in the dermis, melanophages, and melanin incontinence.[3]
The treatment is similar to that of classic MF and is skin-directed. The diagnosis usually gets delayed due to asymptomatic and subtle clinical features. The treatment is similar to that of classic MF and is skin-directed.

Case Report

A 65 years old male presented with asymptomatic pigmentary lesions over the trunk and extremities. On examination patient had mottled hyper and hypopigmentation of skin with interspread telangiectases and areas of atrophy since 10 years (figure 1&2). On the basis of clinical features, diagnosis of poikilodermatous mycosis fungoidies was kept. In the biopsy, the findings were consistent with poikilodermatous MF.

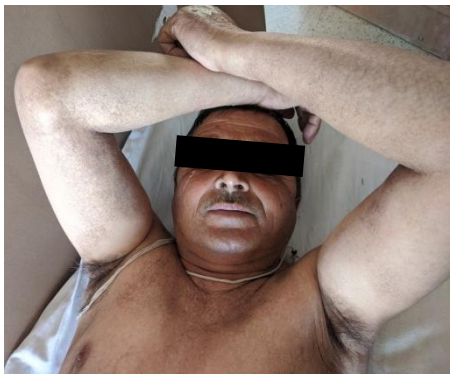

Figure 1

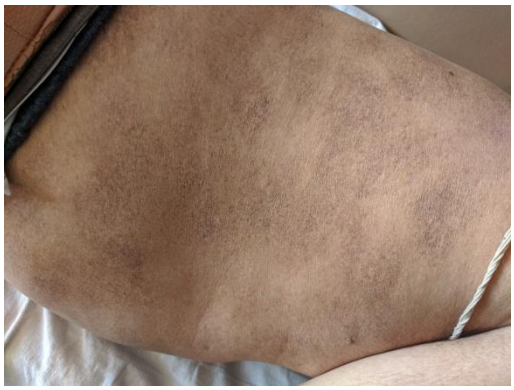

Figure 2

Figure 1 & 2 showing mottled hyper and hypopigmentation of skin with interspread telangiectases and areas of atrophy.

Discussion

Poikiloderma vasculare atrophicans is considered to be a rare variant of mycosis fungoides characterised clinically with mottled pigmentation, telangiectasias and atrophy. It usually appears on the trunk and flexural area.[4]

Similar to classic MF, poikilodermatous MF presents as an early stage (IA-IIA) at diagnosis with a male predominance.[5]

PVA has been known to show a benign course, without progression to the tumor stage of mycosis fungoides.[6]

There was a slight predominance of male patients 1.6 : 1 (30 of 49) with a median age of 44 years.[6]

The differential diagnosis includes large plaque parapsoriasis; connective-tissue diseases, such as lupus erythematosus and dermatomyositis; poikiloderma of Civatte and genodermatoses, such as Rothmund–Thomson syndrome.[7]

The treatment is similar to that of classic MF and is skin-directed. Narrowband ultraviolet B phototherapy is the first-line therapy for poikilodermatous MF.[1] Other skin-directed treatments include topical glucocorticoids, topical retinoids, topical cytotoxic agents, and radiation therapy. Topical glucocorticoids are not so effective in poikilodermatous MF as they are in classic MF.[1] If the patient fails skin-directed treatments, systemic therapies such as oral retinoids and alpha interferon are effective.[8]

Conclusion

Poikilodermatous MF is one of the rare clinical variants of MF. The asymptomatic and subtle nature of the disease may result in delayed diagnosis. The high index of suspicion and clinicopathological correlation is essential to make a precise diagnosis of MF.

Identifying the subtle features early and confirming the diagnosis with histopathology can help in early diagnosis and early treatment.

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.

Conflict of Interest: Nil

References

1. Hassan I, Keen A. Poikilodermatous mycosis fungoides: A case report. Webmed Cent Dermatol. 2011;2: WMC002124.
2. Bloom B, Marchbein S, Fischer M, Kamino H, Patel R, Latkowski JA. Poikilodermatous mycosis fungoides. Dermatol Online J. 2012;18:4.
3. Farley- NLoftus R, et al. Poikilodermatous mycosis fungoides. Dermatol Online J 2010;16(11):8
4. Kazakov DV, Burg G, Kempf W. Clinicopathological spectrum of mycosis fungoides. J Eur Acad Dermatol Venereol 2004;18:397-415.
5. Abbott RA, Sahni D, Robson A, Agar N, Whittaker S, Scarisbrick JJ. Poikilodermatous mycosis fungoides: a study of its clinicopathological, immunophenotypic, and prognostic features. J Am Acad Dermatol. 2011; 65:313–319.
6. Assaf C, Sterry W. Cutaneous lymphoma. In: Wolff K, Goldsmith LA, Katz SI, Gilchrest BA, Paller AS, Leffell DJ, editors. Fitzpatrick's dermatology in general medicine. 7th ed. New York, USA: McGraw-Hill; 2008. pp. 2154–57.
7. Assaf C, Sterry W. Cutaneous lymphoma. In: Wolff K, Goldsmith LA, Katz SI, Gilchrest BA, Paller AS, Leffell DJ, editors. Fitzpatrick's dermatology in general medicine. 7th ed. New York, USA: McGraw-Hill; 2008. pp. 2154–57.
8. Bloom, B., Marchbein, S., Fischer, M., Kamino, H., Patel, R., & Latkowski, J.(2012). Poikilodermatous mycosis fungoides. Dermatology Online Journal, 18(12).