Dear editor,

“Poikiloderma” in dermatology is a descriptive term used for combination of cutaneous atrophy, telangiectasia, hypo or hyperpigmented macules giving mottled appearance to the skin. At many occasions fine scales, tiny lichenoid papules, and petechial hemorrhage can be associated with such lesions. There are various acquired causes of poikiloderma but dermatomyositis, systemic lupus erythematosus, systemic sclerosis, poikiloderma of Civatte, atopic dermatitis, topical misuse of steroid, etc., are prominent ones.[1] Poikilodermatous mycosis fungoides (PMF), previously termed as poikiloderma vasculare atrophicans or parapsoriasis variagata, is a rare variant of mycosis fungoides (MF) comprising of 1–2% of total MF cases. This variant is early in onset but very indolent and take longer time to diagnose as frank MF.[2-4] Our patient a 38-year-old, nonsmoker, male without any previous comorbidities became symptomatic with redness, pruritus, mottled pigmentation, and burning sensation associated with intermittent scaling of body since last 9 years. It was insidious in onset and gradually progressive from neck area to cover almost entire body over 3 years. He had multiple exacerbations with erythema and scaling during the course of disease with no remission. He was given trial of methotrexate, psoralen with ultraviolet rays therapy (PUVA), acitretin, topical steroid, and emollient without much benefit. He did not tolerate acitretin, PUVA, narrow band ultra violet rays (NBUVB) in the past. He used to get temporary relief on scaling, erythema, and pruritus with topical steroid but used to frequently relapse with severe burning sensation, superficial dermatophyte infection. There was no history of fever, joint pain, weight loss, drug intake, or any underlying systemic disease. General and systemic examinations were essentially normal. Dermatological examinations revealed diffuse poikiloderma interspersed with fine scales and tiny lichenoid papules over the face, neck, trunk, and both the extremities (upper extremities more than lower) covering 90% body surface areas [Figure 1a-d]. It was more prominent over the neck, face, trunk, and upper extremities sparing lower leg, palms, soles, nails, oral, and genital mucosa. Dermoscopic examination revealed typical features of poikiloderma [Figure 1e]. His repeated hematological, biochemical tests, viral markers (Hepatitis B&C, HIV, Herpes simplex virus, Cytomegalovirus, Epstein-Barr virus), antinuclear antibody test, VDRL, chest X-ray PA view, USG abdomen, CT chest, and abdomen were essentially normal. Skin biopsies performed at multiple occasions revealed dilation of vessels at papillary dermis along with perivascular lymphocytic infiltrate, few atypical lymphocytes without any conclusive immunohistochemistry (IHC) suggestive of mycosis fungoides. As the disease progressed, the skin biopsy revealed more number of atypical lymphocytes without loss of CD7 in upper dermis. However, last histopathological examination (HPE) and IHC performed on biopsies taken from upper back, lower back, and left arm revealed classical features of mycosis fungoides [Figure 2a-f]. On further evaluation by contrast-enhanced computed tomography chest and abdomen, positron emission tomography scan, he was given final diagnosis of erythrodermic poikilodermatous mycosis fungoides stage IIIA (T3N0M0B0). TCR gene analysis revealed T cell clonality. He was given trial of cap bexarotene in a dosage of 150 mg daily and gradually increased to 300 mg daily over 15 days. He developed extreme itching, burning sensation, and dryness of the skin so it was stopped over 3 weeks of therapy. Presently, he has been restarted on tab methotrexate along with emollient and topical steroid. There is no satisfactory relief in his skin condition. He has been planned for total skin electron beam therapy in consultation with radiation oncologist.

Discussion

The poikilodermatous form of MF even in erythrodermic presentation has better prognosis than classical MF with 100% survival rate in 5 years.[5] Poikiloderma in PMF can be generalized like erythroderma or localized mainly in folded areas or mixed, present admixed with other forms of MF or cutaneous T-cell lymphoma. Unlike pruritus of classical MF, burning sensation is more bothersome clinical presentation of PMF. Poikiloderma in covered areas of body classically known as ‘bathing suit’ type of distribution differentiates it from dermatomyositis, lupus erythematosus, systemic sclerosis, and poikiloderma of Civatte, which involve more of photo exposed areas.[5-6] Epidermotropism, Pautrier microabscesses, and “wiry” collagen which are commonly described HPE findings in classical MF are relatively uncommon in PMF. Here, epidermal atrophy, scarce epidermotropism, infiltration of upper dermis with atypical lymphocytes giving band like appearance, pigmentary incontinence, and dilatation of vessels are prominent features. It is the “lining of atypical lymphocytes” on dermoeidermal junction that alerts the dermatopathologist to follow-up such cases with repeated biopsy if initial IHC does not satisfy the criteria of MF.[5] We also found accentuation of reticular brown pigmentation pattern on a erythematous white background along with irregularly, interspersed, brown black dots and telangiectatic vessels described by Sinha et al.[4] and Bharti and Khopkar.[7]
Approach and therapeutic modalities are akin to classical MF comprising of skin-directed and systemic therapy based on the stage of the disease. Skin-directed therapies include topical corticosteroids, narrowband ultraviolet B, PUVA, bexarotene, mechlorethamine 0.01%, carmustine 0.04%, 5% 5-fluorouracil cream, total skin electron beam radiation, UVA1, and excimer laser. PMF usually respond poorly to skin-directed therapies. The skin-directed therapy-resistant disease cases require methotrexate, oral retinoids (acitretin, isotretinoin, and bexarotene), extracorporeal photopheresis, combination systemic chemotherapy (cyclophosphamide, adriamycin, vincristine, prednisone/cyclophosphamide, vincristine, prednisone), and immunotherapy like interferon alpha subsequently.[8,9] Mycosis fungoides as such take long time to diagnose. In our case methotrexate treatment, other treatment modalities like PUVA, NBUVB could have altered characteristic histopathological and immunohistochemical findings usually observed in the disease. Unlike many reports of good response to PUVA, NBUVB, acitretin our case had exacerbation of erythroderma following these modalities. Unlike previous good response to bexarotene in few cases of advanced mycosis fungoides, sezary syndrome with erythroderma in our center, this patient did not tolerate this drug. Probably earlier intolerance to similar drug like acitretin could be possible explanation. There is long journey from the onset of poikilodermatous lesions to development of frank MF. Many dermatoses clinically and histopathologically mimic this condition. Therefore, close
observation of clinical features, dermoscopy, repeated HPE and IHC, and high index of suspicion are key to diagnose PMF.

**Declaration of patient consent**

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

**Financial support and sponsorship**

Nil.

**Conflicts of interest**

There are no conflicts of interest.

**Gautam Kumar Singh, Sandeep Arora¹, Pankaj Das, Vikram Singh², Vishal Sharma³, Akanksha Gupta⁴**

Departments of Dermatology, Venereology and Leprosy, and ¹Dermatology, Base Hospital Delhi Cantt, Affiliated Faculty, Army College of Medical Sciences, Delhi, ²Department of Pathology, Army Hospital (R&R), Delhi, ³Department of Dermatology, Venereology and Leprosy, Command Hospital Western Command, Chandigarh, ⁴Department of Dermatology, Venereology and Leprosy, Base Hospital, Delhi, India

**Address for correspondence:**
Dr. Gautam Kumar Singh, Department of Dermatology, Venereology and Leprosy, Base Hospital Delhi Cantt and Army College of Medical Sciences, Delhi – 110 010, India.
E-mail: gk1june@gmail.com

**References**

1. Nofal A, Salah E. Acquired poikiloderma: Proposed classification and diagnostic approach. J Am Acad Dermatol 2013;69:e129-40.
2. Farley-Loftus R, Mandal R, Latkowski JA. Poikilodermatous mycosis fungoides. Dermatol Online J 2010;16:8.
3. Bhide AA, Singh PC, Kura MM. Poikilodermatous mycosis fungoides with erosive lesions. Indian J Dermatol 2019;64:251.
4. Sinha P, Tripathy DM, Shelly D, Neema S. A rare case of poikilodermatous mycosis fungoides. Indian J Dermatol 2020;65:417-9.
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;65:313–9.
6. Vasconcelos Berg R, Valente NYS, Fanelli C, Wu I, Pereira J, Zatz R, et al. Poikilodermatous mycosis fungoides: Comparative study of clinical, histopathological and immunohistochemical features. Dermatology 2020;236:117‑22.
7. Bharti AH, Khopkar US. Dermoscopy of poikilodermatous mycosis fungoides in pigmented skin. Int J Dermoscopy 2017;1:28-31.
8. Gilson D, Whittaker SJ, Child FI, Scarisbrick JI, Illidge TM, Parry EJ, et al. British Association of Dermatologists and U.K. Cutaneous Lymphoma Group guidelines for the management
of primary cutaneous lymphomas 2018. Br J Dermatol 2019;180:496-526.

9. Trautinger F, Eder J, Assaf C, Bagot M, Cozzio A, Dummer R, et al. European organisation for research and treatment of cancer consensus recommendations for the treatment of mycosis fungoides/Sézary syndrome-Update 2017. Eur J Cancer 2017;77:57-74.

How to cite this article: Singh GK, Arora S, Das P, Singh V, Sharma V, Gupta A. A long journey of poikilodermatous erythroderma to poikilodermatous mycosis fungoides: A case report. Indian Dermatol Online J 2022;13:663-6.

Received: 15-Oct-2021. Revised: 11-Nov-2021. Accepted: 02-Dec-2021. Published: 05-Sep-2022.