Solitary mycosis fungoides treated with photochemotherapy: A case report

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ABSTRACT:

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Mycosis fungoides (MF) is the most frequent diseases among cutaneous T-cell Lymphoma (CTCL). MF is categorized as being patch, plaque, or tumor stage, but patients may concurrently have more than one type of the lesion. Solitary or unilesional mycosis fungoides is a variant of MF characterized by a single lesion involving less than 5% of total body surface skin area. Clinical feature is single erythematous plaque occurs anywhere on the body such as scalp, face, trunk, upper and lower extremities. Histopathology is similar to MF which has atypical lymphoid infiltrate with epidermotropism, possible adnexal involvement. Many cases report presented relationship among MF, koebner phenomenon and cutaneous infection. However solitary MF usually has an indolent course and good prognosis. There are various therapeutic approaches to solitary MF such as radiotherapy, surgical excision, photodynamic therapy and topical corticosteroid. We report an 84-year-old woman presented with solitary scaly hyperkeratotic erythematous plaque with minimal erosions on left palm for 1 year. She had history of thorn injury at left palm before the lesion developed. The histopathological study, immunohistochemistry and T-cell receptor (TCR) gene rearrangement were compatible with MF. She was treated by topical psoralen plus UVA (PUVA) with resolved of lesion after 6 months of treatment without any side effect.

Key words: unilesional, solitary, mycosis fungoides

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Introduction

Mycosis fungoides (MF) is the most frequent diseases among cutaneous T-cell lymphoma (CTCL) and accounts for more than 40% of all primary CTCL. The solitary MF or solitary CTCL is a unique form of MF that present as an isolated lesion with similar histopathologic features, immunocytochemistry with typical patch and plaque-stage MF.

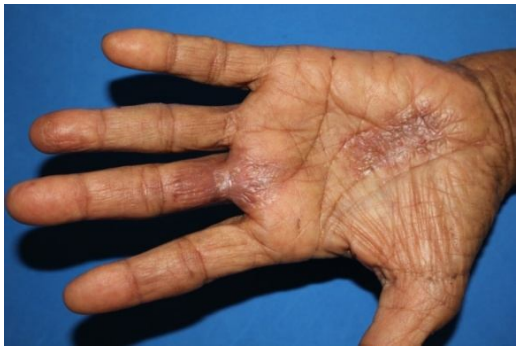

Figure 1 Solitary scaly hyperkeratotic erythematous plaque with minimal erosions on left palm

Case report

An 84-year-old Thai woman presented with solitary scaly hyperkeratotic erythematous plaque with minimal erosions on left palm for 1 year. She had history of thorn injury at left palm for 1 month before the lesion occurred. She received short course of oral antibiotic but the lesion gradually expanded. She went to primary care hospital, the skin biopsy was done and histopathological feature was compatible with MF. She was referred to our hospital for an appropriate management. Her underlying disease was hypertension. Dermatological examination revealed solitary scaly hyperkeratotic erythematous plaque with minimal erosions on left palm. (Figure 1) Systemic examination revealed no abnormality.

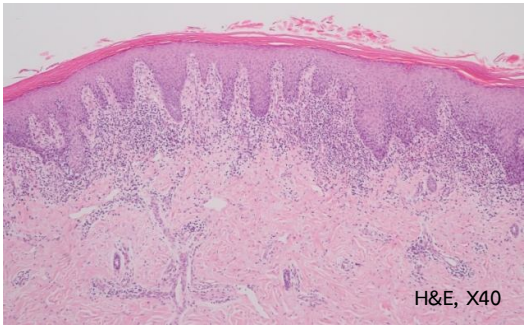

Figure 2 The histopathological feature showed epidermal hyperplasia and hyperkeratosis in the epidermis. There was dense lichenoid cells infiltrate in the dermis

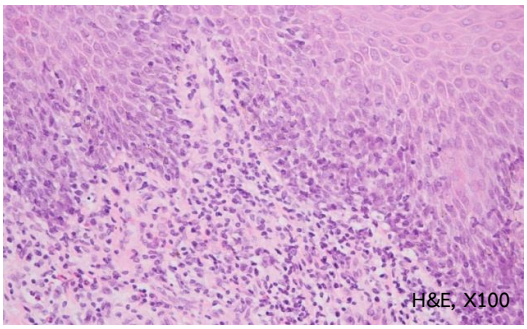

Figure 3 There were epidermotropism of atypical lymphocytes in the epidermis and dense lichenoid cells infiltrate in the dermis composed of mainly large convoluted dense nucleolar lymphocytes

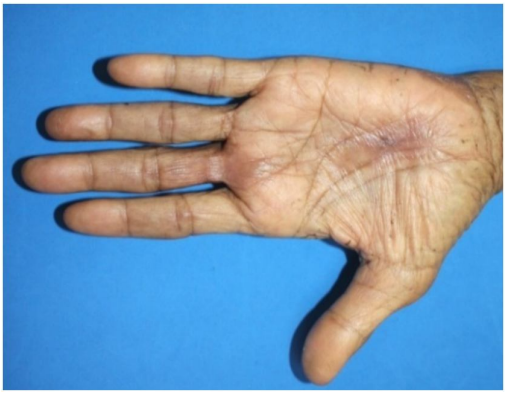

Figure 4 Post inflammatory hyperpigmentationon left palm after 58 sessions of local PUVA therapy

The histopathological feature showed epidermal hyperplasia and hyperkeratosis in associated with epidermotropism of atypical lymphocytes. There was dense lichenoid cells infiltrate composed of mainly large convoluted dense nucleolar lymphocytes. (Figure 2 and 3) Immunohistochemistry from skin showed CD3+, CD4+, CD5+, CD8-, CD20- and BF1+. T-cell receptor (TCR) gene rearrangement from skin showed monoclonal TCR gene rearrangement. Skin culture for fungus and polymerase chain reaction (PCR) for18s ribosomal ribonucleic acid (rRNA) were positive for *Candida parapsilosis*. Skin culture for *Mycobacterium* spp., PCR for *Mycobacterium* spp., and 16s rRNA were also negative. Other investigations include complete blood count, liver function test, *lactate dehydrogenase* (LDH) and chest X-ray revealed no abnormality. She was diagnosed as solitary MF with cutaneous candidiasis on left palm.

She was treated with oral fluconazole 200 mg/day for 2 weeks followed by topical psoralen (1:50) plus UVA (PUVA) 1J/cm², 2 times/week for 23 sessions, then 1.5J/cm² 2 times/week for 20 sessions, 1.5J/cm² 1 time/week for 10 sessions, and 1.5J/cm² 1 time/2weeks for 5 sessions, respectively. The lesion was gradually resolved with post inflammatory hyperpigmentation (Figure 4) after 58 sessions of PUVA therapy. Topical corticosteroid was not applied in this case.

Discussion

The solitary MF or unilesional MF was first described by Jones and Chu in 1981[1] which defined as a single area of involvement by MF covering less than 5% of the body surface area. It clinically manifests as a solitary patch, plaque or nodule that may vary in size and location that usually arising in any age group[2]. The solitary MF or unilesional MF histopathology includes an atypical lymphoid infiltrate with epidermotropism, with possible adnexal involvement which is indistinguishable from classic MF.

Pathogenesis of MF is T cells residing in the skin underlies a significant potential for neoplasia. Genetic factors, such as individual's HLA type, may influence someone to develop CTCL by

inappropriate activation and accumulation of T cells via antigen presentation[3]. The environment may decontrol tumor suppressor or pro-oncogenic pathways include viral or other organism pathogens (Epstein-Barr virus, herpes simplex virus, *Staphylococcus aureus*, dermatophytes, *Mycobacterium leprae*, and *Chlamydia pneumoniae*)[4,5]. Drug triggers (antihistamines, antiepileptics, antihypertensives, and selective serotonin reuptake inhibitors)[6], occupational (exposure to aromatic hydrocarbons)[7] and nutritional association (vitamin D deficiency)[8] support an environmental role in the evolution of CTCL.

Clinically, MF is categorized as being in the patch, plaque, or tumor stage, but patients may concurrently have more than one type of the lesion. Other MF variants are folliculotropic MF, hypopigmented MF, pagetoid reticulosis, granulomatous slack skin and unilesional MF (UMF).

Solitary MF or unilesional MF is a variant of MF which has an excellent prognosis. Since 1981, only 166 solitary cases of MF have been described with one case of cutaneous spreading and three cases of large cell transformation. The patients were followed for a minimum of 12 months (range, 12 to 96 months)[9,10].

Ally and Robson suggest a useful algorithm to define the diagnosis of solitary MF which begin with epidermotropism of significant cytological atypia of lymphocyte in combination with loss of T-cell antigen and detection of monoclonal TCR gene rearrangement. Treatment modalities are radiotherapy, surgical excision, photodynamic therapy and topical therapy (e.g., potent corticosteroids, calcineurin inhibitors, imiquimod, carmustine, and nitrogen mustards)[9,10].

Our patient was treated by topical PUVA with resolved of lesion after 6 months of treatment without any side effect or recurrence after 6 months of follow up. One case report, 9 years old boy was diagnosed unilesional folliculotrophic mycosis fungoides and treated with topical PUVA. Complete response was achieved after 30 sessions[11].

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