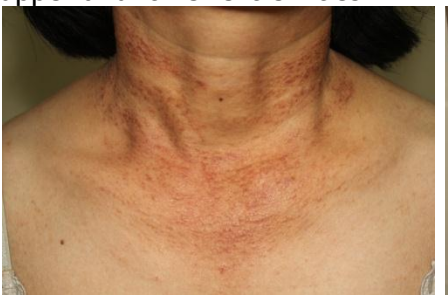
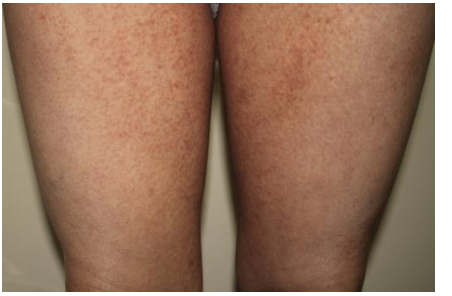
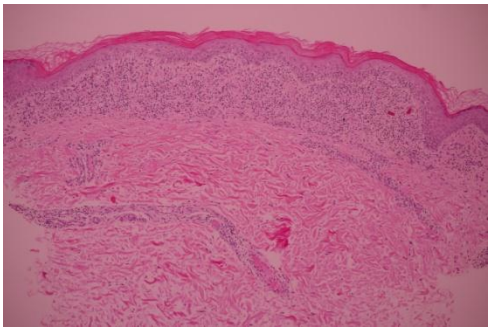
Case 21
A 50-year-old woman from Chonburi.
Chief complaint: Asymptomatic rash on neck, trunk, both upper and lower extremities for 2 years.
Present illness: The patient has developed asymptomatic scaly erythematous papules, hyper- and hypopigmentation on neck, both upper and lower extremities for 2 years. She denied the history of fever, weight loss, or other systemic symptoms.
Past history: No underlying disease.

Physical examination:
LN: cannot be palpated
GI: no hepatosplenomegaly
Skin examination:
Multiple slightly raised, erythematous papules with scale, on atrophic, mottled hyper- and hypopigmented patches, in association with telangiectasia on her neck and the inner side of both proximal upper and lower extremities.

Histopathology: (S13-010171, left arm)
There was hyperkeratosis with focal parakeratosis dense superficial lichenoid infiltration of lymphocytes admixed with some melanophages in the mid thicken papillary dermis with some exocytosis with poral edema vascular atrophic. Positive staining for CD3, CD4, and CD8.



Investigation:

- CBC: WBC 6,830 cells/mm^3(N 60%, L 29%, M 8%, E 3%), Hb 9.9 g/dL, Hct 30.1%, MCV 57.8, Plt 489,000 cells/mm^3
- Hb typing: EA (HbA 73.2%, HbE 26.8%)
- Renal and liver function tests: normal

Diagnosis: Poikilodermatous mycosis fungoides, stage IB
Treatment: PUVA
0.1% Triamcinolone acetonide milk lotion

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Discussion

Mycosis fungoides(MF) is the most common type of cutaneous T-cell lymphoma (CTCL). Neoplastic cells, that result from the clonal expansion of CD4+ memory T-cells, demonstrate a predilection for skin.[1, 2] MF is usually arising in mid to late adulthood, with the median age at diagnosis about 55-60 years and a male predominance.

Classic MF is characterized by erythematous, atrophic, and wrinkled patches, with fine scale on non-sun-exposed skin. Plaques may arise de novo or from the progression of patch-stage disease. Further progression may result in nodules and tumors, which often

ulcerate. Clinical variants of MF are less common including granulomatous slack skin, pagetoid reticulosis, hypopigmented MF, poikilodermatous MF, and folliculotropic MF.[2]

Poikilodermatous MF, originally termed poikiloderma atrophicans vasculare described by Jacobi in 1907, it initially was considered to be a premycotic eruption that would eventually progress to MF.[3-5] In the past, it was classified into the group of large-plaque parapsoriasis, but the current data support that this form represents a clinical variant, and is not a precursor of MF.[6,7] Poikilodermatous MF is a rare clinicopathologic variant of MF clinically characterized by localized or diffuse patches, which consist of telangiectasia, mottled hyper- and hypopigmentation, and atrophy.[1] Lesions often are noted on breasts, hips, buttocks, and flexural areas. It may coexist with patches of classic MF in some patients.[8] Patches may be asymptomatic or mildly pruritic.[2] Patients with poikilodermatous MF often describe a stinging sensation rather than pruritus. Similar to classic MF, poikilodermatous MF usually presents at an earlier stage (IA-IIA) at time of diagnosis with a male predominance. However in comparison with classic MF, poikilodermatous MF has an earlier age of onset (median age of 44 years). Poikilodermatous MF is also frequently associated with lymphomatoid papulosis (18%).[9]

The differential diagnosis of poikilodermatous MF includes other conditions in which poikiloderma is prominent, such as dermatomyositis, lupus erythematosus, poikiloderma of Civatte, overuse of topical glucocorticoids, radiation dermatitis, graft-versus-host disease, and genodermatoses, such as Rothmund-Thompson syndrome.[1,8]

Histopathology from poikilodermic areas shows a markedly flattened epidermis, covered with hyperkeratosis or parakeratosis. Epidermotropism is a constant feature and, occasionally, mycosis/Sézary cells may be seen. In the dermis, there is a band-like or perivascular lymphohistiocytic infiltrate. Dilated vessels,

pigmentary incontinence, and loss of rete ridges are usually evident. The superficial dermis is scarred and elastic fibers are characteristically absent.[8] The atypical lymphocytes are commonly CD4+, which is similar to classic MF. However, several recent studies suggest a predominance of a CD8+, CD4- immunophenotype.[9]

Staging in patients with MF is crucial as it will determines the management, treatment and prognosis. Recently, a revised clinical staging system for MF has been proposed, which is based on the TNM(tumor–node–metastasis) classification system and takes into account the type and extent of skin lesions as well as the presence or absence of nodal, visceral and peripheral blood involvement.[10]

Treatment for poikilodermatous MF is similar to that of classic MF. Initial management depends on the stage, age, and general health of the patient. Treatment modalities for MF range from skin-directed to systemic therapies. Treatment with NBUVB and PUVA are safe and effective. Other skin-directed therapies reported to be helpful include topical glucocorticoids, topical retinoids, topical cytotoxic agents, and radiation therapy. Topical glucocorticoids are not as effective in poikilodermatous MF as they are in classic MF. If the patient fails cutaneous therapies or presents at an advanced stage, systemic therapies such as oral retinoids and alpha interferon are preferably used.[11]

Poikilodermatous MF has an excellent prognosis. From a retrospective study over a mean follow-up period of more than 11 years, 96% of patients remained at the same stage.[9]

In summary, our patient was diagnosed poikilodermatous MF stage IB based on clinical, histopathology, and immunohistochemistry findings. She received topical corticosteroid treatment and PUVA phototherapy with good response. However, regular follow up is necessary.

References

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