Case 13

A 42-year-old Thai female from Bangkok

Chief complaint: Multiple progressive confluent reticulated erythematous patches and thin plaques with hypo- & hyperpigmented and atrophic patches on both axillae, middle abdomen and both groins for 2 years

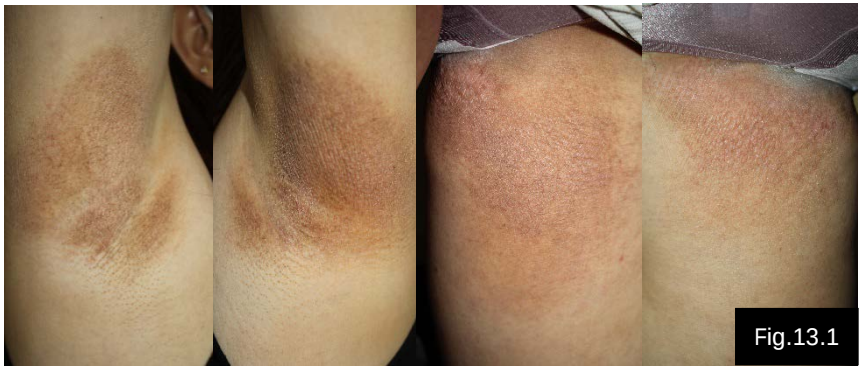

Fig.13.1

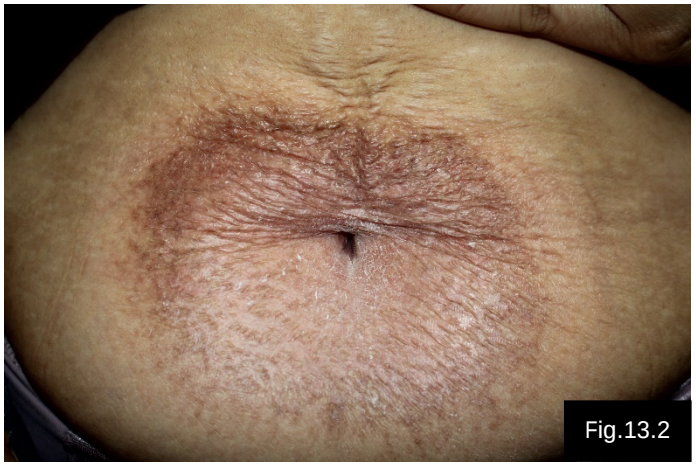

Fig.13.2

Present illness: Thirteen years ago, she was diagnosed with mycosis fungoides (MF) stage IIB and treated with PUVA phototherapy for 2 years and then she loss follow-up due to pregnancy. 8 years ago, the rash progressed and skin biopsy was done for confirm diagnosis of MF. After treatment with PUVA phototherapy, she also loss follow-up again. Now a day, she visits Ramathibodi hospital due to multiple progressive widespread confluent reticulated erythematous patches and thin plaques with hypo- & hyperpigmented and atrophic patches, admix with wrinkle surface and fine scales on both axillae, middle abdomen and both groins for 2 years.

Past history: No underlying disease

Family history: No family history of skin disease

Physical examination:

- V/S: BT 37.3°C, BP 104/65 mmHg, PR 77/min, RR 20/min
- HEENT: No pale conjunctivae, no icteric sclerae
- Heart: Normal S1 S2, no murmur
- Lung: Normal and equal breath sound
- Abdomen: Soft, no mass, no hepatosplenomegaly
- Lymph node: Impalpable

Dermatologic examination:

- Multiple progressive widespread confluent reticulated erythematous patches and thin plaques with hypo- & hyperpigmented and atrophic patches, admixed with wrinkle surface and fine scales on both axillae, middle abdomen and both groins (Fig.13.1, 13.2)

Histopathology (S18-037008, abdomen):

- Band like infiltration of atypical lymphocytes with epidermotropism (Fig.13.3)

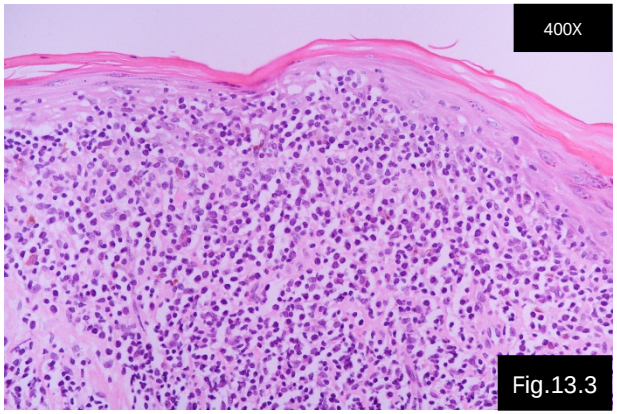

Fig.13.3

Immunochemistry: Not done

Investigation:

- CBC: Hb 13.3 g/dL, Hct 41.4%, Plt 359,000 /mm^3, WBC 5,620 /mm^3 (N 49%, L 39%, M 7%, E 5%)
- AST/ALT: 26/27 U/L, ALP/GGT: 64/30 U/L, TB/DB: 0.6/0.2 mg/dL
- BUN/Cr: 10/0.5 mg/dl

Diagnosis: Poikilodermatous mycosis fungoides

Treatment:

- PUVA phototherapy twice weekly
- Long-term follow-up for monitoring systemic involvement and recurrent disease

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Discussion:

Poikilodermatous mycosis fungoides is a rare distinct clinical variant of cutaneous T-cell lymphoma (CTCL), formerly referred to as poikiloderma vasculare atrophicans or parapsoriasis variegate.[1] This complex dermatologic disease characterized by telangiectasia, pigmentation and atrophy which typically presents on non-sun exposed areas.[2-4]

Poikilodermatous MF is usually characterized by the development of large plaques or generalized skin involvement.[5-7] However, at the onset of the disease the lesions may present either small plaques or papules arranged in a net-like pattern. The typical patches often show a predilection to the major flexural areas & trunk and present with erythema, mild scaling, mottled dyspigmentation (hypo- and hyperpigmentation) with atrophy and telangiectases. As opposed to classic MF, poikilodermatous lesions are generally asymptomatic or mildly pruritic and are usually stable or slowly increasing in size.[8,9] The first manifestation of poikilodermatous MF usually occurs at an earlier age than that of classic MF and a male predominance was reported for both forms.

Histopathology of poikilodermatous lesions discloses an atypical T-cell infiltrate in the papillary dermis often with epidermotropism.[10,11] However, Pautrier microabscesses are not as common in comparison to classic MF. Melanophages and melanin incontinence are also observed and admix with telangiectasia & epidermal atrophy.

For immunohistological staining commonly shows either a revalence of the CD4+, CD8− pattern or CD8+, CD4− immunophenotype.

In our case, the lesions and histopathological finding are typically poikilodermatous MF. The evaluation of the patient was done by physical examination and laboratory investigation which show limited cutaneous involvement without lymphadenopathy. So CT scan was not done in this case and skin-directed treatment which was PUVA phototherapy for twice weekly was prescribed in this case. Now a day, the lesions are improved and PUVA phototherapy was gradually taper off by once a week.

References:

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