Case 14.1

A 28-year-old female

Chief Complaint: Multiple hypopigmented patches for 6 months.

Present Illness: She developed hypopigmented patches at both thighs for 5 years without any symptoms. No history of erythema prior to hypopigmented lesions. During the past 6 months, she noticed that the lesions gradually increase in size and seems to be extent. Then, she came to Ramathibodi Hospital. KOH examination was done and the result was negative.

Past History: Healthy

Family History: There was no family member experienced with similar skin lesion.

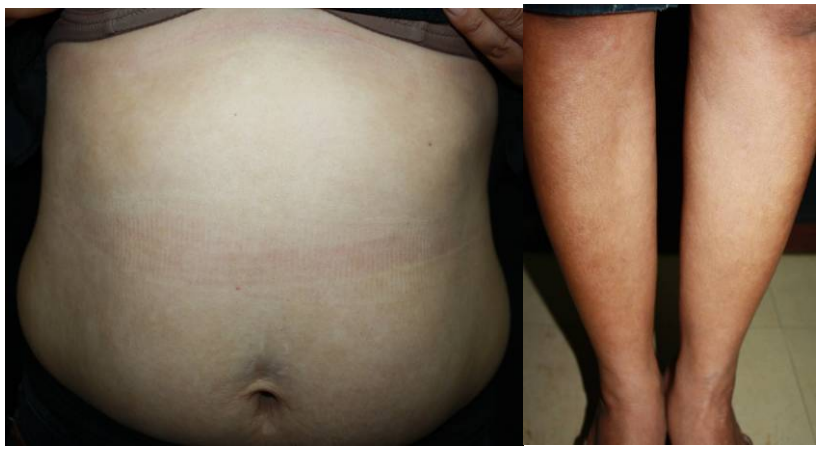
Physical Examination: Unremarkable

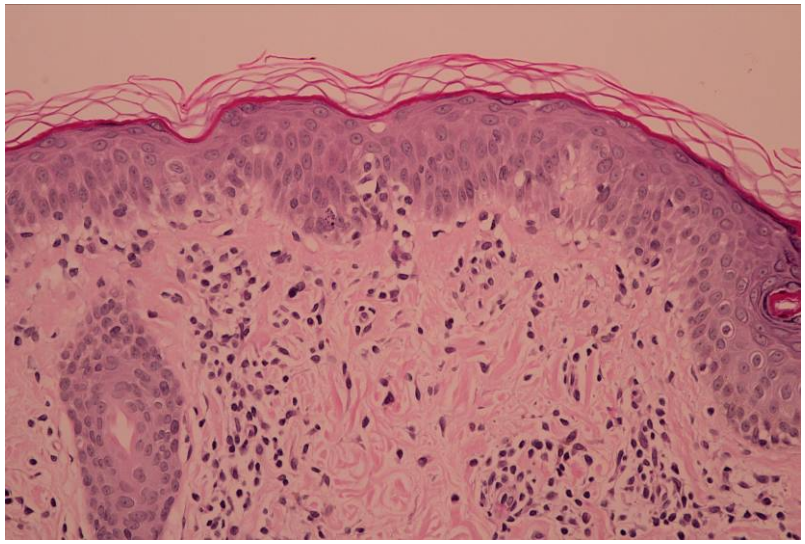
Dermatological Examination: Generalized discrete well-defined non-scaly hypopigmented patches on trunk and all extremities.

Histopathology: (s11-4554A,B)

Dense superficial perivascular patchy lichenoid infiltrate of lymphocytes in the thickened papillary dermis

Some lymphocytes show atypical nuclei and exocytosis in the epidermis (epidermotropism)





Diagnosis: Hypopigmented Mycosis Fungoides (patch stage)

Treatment: Topical corticosteroid, Narrowband UVB

Case 14.2

A 35-year-old female

Chief Complaint: Multiple hypopigmented patches for 5 months.

Present Illness: She developed hypopigmented patches at both thighs for 5 months. She notices that some lesions occurred as small erythematous macules then gradually increased in size and turned to be a large hypopigmented patches scattered all trunk and extremities without any symptoms.

Past History: Healthy

Family History: There was no family member experienced with similar skin lesion.

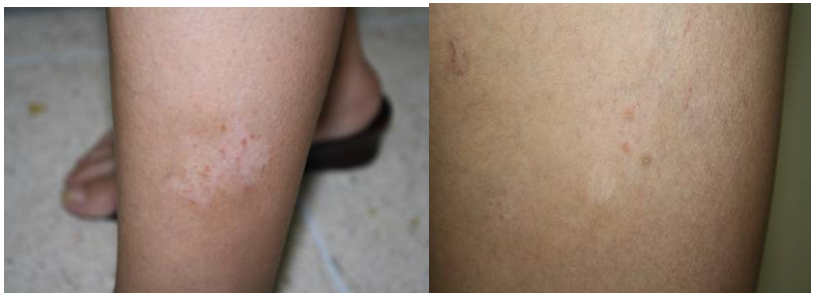
Physical Examination: Unremarkable

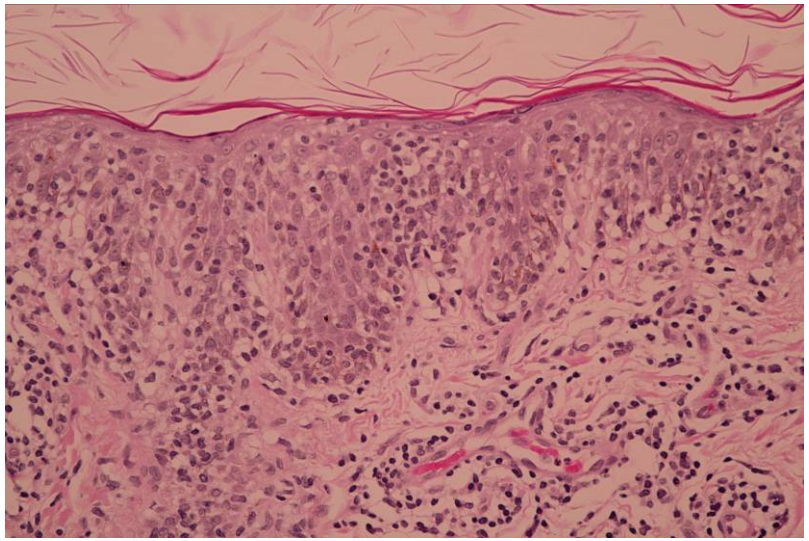
Dermatological Examination: Generalized discrete non-scaly hypopigmented patchs with some erythematous macules on top of the lesions at trunk and extremities.

Histopathology: (S11-011907A)

Superficial and deep perivascular and lichenoid infiltrate of lymphocytes with marked epidermotropism

Some lymphocytes show atypical nuclei with hyperchomatic and convolutend nuclei, predominantly in the epidermis (epidermotrophism)





Diagnosis: Hypopigmented Mycosis Fungoides

Treatment: Topical corticosteroid, Narrowband UVB

Presenter: Sunatra Nitayavardhana, M.D.

Consultant: Poonkiat Suchonwanit, M.D.

Discussion:

Cutaneous T-cell lymphomas (CTCL) are non-Hodgkin lymphomas characterized by dominant skin-homing T-cell clone and mycosis fungoides (MF) is the most common type of CTCL. MF typically affects mid to late adulthood with male predominance (2:1). MF is characterized by slow progress over years or decades from patch stage to plaque stage and finally to tumor stage. The risk of extracutaneous disease depends on the extent and type of the skin lesion. Tumors tend to be more aggressive form.[1, 2]

Hypopigmented MF is a rare clinical variant of early-stage MF which is commonly observed in dark skin, probably because of clinically obvious. Actually MF is primarily a disease of adulthood but this variant seems to be more frequent in children.[3,4,5] The natural history of this disease is similar with conventional MF, asymptomatic/mildly pruritic, variable in size, hypopigmented or erythematous, finely scale lesions can be observed. The mixed clinical pattern (hypopigmented and erythematous) is observed in Caucasian patients.[6,7] The distributions are usually in non-exposed sites. Hypopigmented MF may resemble the clinical features of other skin diseases such as vitiligo, atopic dermatitis, pityriasis lichenoides chronica, pityriasis alba, leprosy or tinea versicolor so these should be in differential diagnosis.[8,9]

The pathogenesis of hypopigmented MF is still unclear. Hypopigmentation may be due to the effect of neoplastic T lymphocytes, which have capability to destroy melanocytes in affected skin.[11, 12]

Histopathology shows superficial band-like infiltrates mainly lymphocytes and mostly confined in the epidermis (epidermotrophism). The present of intraepidermal nest of atypical cells (Pautrier's microabscess) is pathognomonic.

Immunophenotype of hypopigmented MF is usually CD8+ T-cell which is different from conventional MF is that CD4+. However, Ardigo et al. reported that 57% of cases who were Caucasians show predominant phenotype of T-helper cells. It may be speculated that CD8+ is typical for hypopigmented MF in children and darker skin but less frequent in Caucasian patients.[13, 14]

For the treatments, NBUVB and PUVA therapy are safe and effective modalities for early stage of the disease. The previous studies show that the complete remission rates were similar between those phototherapy (70-80%) and relapse-free interval were almost the same (22-24 months).[15, 16] In refractory or advance stage, combination with systemic treatments mainly interferon and retinoid might be valuable.

By clinical and histological finding in our patients, the disease was categorized in stage 1B by TNMB classification. The clinical presentation in the first case, the lesions occurred at hypopigmented patch but in contrast, the second case started with erythematous macules prior to hypopigmented patches which can be found in some case. They had received topical corticosteroids and also NBUVB. All the lesions response well. However, regular follow up is necessary.

References

1. Bolognia Jean L, Jorizzo Joseph, Rapini Roald P, editors. Dermatology 2nd edition, Elsevier: Mosby;2008.
2. Klaus W, Lowel G, Katz S, Barbara A, Paller A, Leffell D, editor. Fitzpatrick's Dermatology in general medicine 7th edition, McGrawhill; 2008.
3. Di Landro A, Marchesi L, Naldi L, Motta T, Cainelli T. A case of hypopigmented mycosis fungoides in a young Caucasian boy. Pediatric Dermatol 1997; 14:449-52.
4. Grunwald MH, Amichai B. Localized hypopigmented mycosis fungoides in a 12 year-old Caucasian boy. J Dermatol 1999; 26:70-1.
5. Ratnam KV, Pang BK. Clinico-pathological study and five-years follow-up of 10 cases of hypopigmented mycosis fungoides. J Eur Acad Dermatol Venereol 1994; 3:505-10
6. El Shabrawi-Caelen L, Cerroni L, Medeiros LJ, McCalmont TH.Hypopigmented mycosis fungoides. Frequent expression of a CD8+ T-cell phenotype. Am J Surg Pathol 2002; 26:450-7.
7. Ardigo M, Borroni G, Muscardin L, Kerl H, Cerroni L. Hypopigmented mycosis fungoides in Caucasian patients: A clinicopathology of 7 cases. J AM Acad Dermatol 2003; 49(2):264-70.

8. Robert C, Moulonguet I, Baudot N, Flageul B, Dubertret L. Hypopigmented mycosis fungoides in a light-skinned woman. Br J Dermatol 1998; 139:341-3.
9. Lambroza E, Stevan R, Cohen R, Phelps R, Lebwohl M, Braverman IM, et al. Hypopigmented variant of mycosis fungoides: demography,histopathology, and treatment of seven cases. J Am Acad Dermatol 1995; 32:987-93.
10. Neuhaus IM, Ramos-Caro FA, Hassanein AM. Hypopigmented mycosis fungoides in childhood and adolescence. Pediatric Dermatol 2000; 17:403-6.
11. Whitmore SE, Simmons-O'Brien E, Rotter FS. Hypopigmented mycosis fungoides. Arch Dermatol 1994; 130:476-80.
12. Palermo B, Campanelli R, Garbelli S, Mantovani S, Lantelme E, Brazzelli V, et al. Specific cytotoxic T lymphocyte responses against Melan-A/MART1, tyrosinase and gp100 in vitiligo by the use of major histocompatibility complex/peptide tetramers: the role of cellular immunity in the etiopathogenesis of vitiligo. J Invest Dermatol 2001; 117:326-32.
13. Whittam LR, Calonje E, Orchard G, Fraser-Andrews EA, Woolford A, Russel-Jones R. CD8-positive juvenile onset mycosis fungoides: an immunohistochemical and genotypic analysis of six cases. Br J Dermatol 2000; 143:1199-204.
14. Castanet J, Ortonne J. Pathophysiology of vitiligo. Clin Dermatol 1997; 15:845-51.
15. Diederen PVMM, van Weelden H, Sanders CJG, Toonstra J, Van Vloten WA. Narrowband UVB and psoralen-UVA in the treatment of early-stage mycosis fungoides: a retrospective study. J Am Acad Dermatol 2003; 48: 215–219.
16. Franz Trautinger. Review phototherapy of mycosis fungoides: Photoimmunology & Photomedicine 2011; 27:68-74.