Case 12.1

Patient: A 23-year-old Thai man from Bangkok

Chief Complaint: 2-week-history of multiple hypopigmented patches at right check

Present Illness: A Thai man presented with non-itchy multiple hypopigmented patches on both forearms for 2 years. 2 weeks ago he developed hypopigmented patches on right check, posterior aspect of the neck and back region. His lesions cannot be relieved by 5% LCD and cold cream

Past History: Healthy

Family History: Nil

Physical Examination: Unremarkable

Dermatological Examination (Figure 12.1-4)**:** Multiple ill defined hypopigmented patches on right mandibular area, posterior aspect of neck, extensor surface of both forearms and back

Figure 12.1

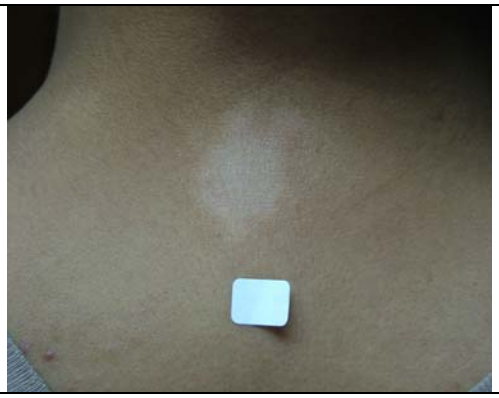

Figure 12.2

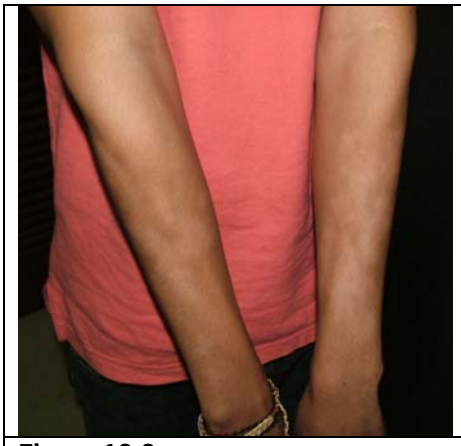

Figure 12.3

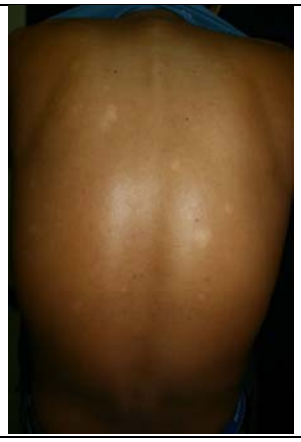

Figure 12.4

Histopathology (S09-11559) (Figure 12.5): superficial perivascular infiltrate of lymphocytes with exocytosis of some atypical lymphocytes into the epidermis

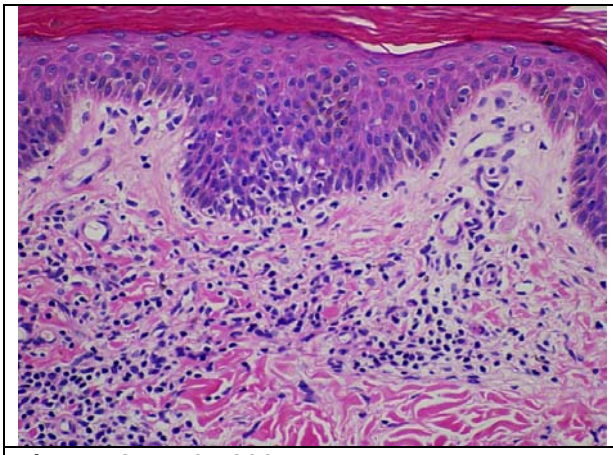

Figure 12.5, H&E 200x

Case 12.2

Patient: A 14-year-old Thai boy from Bangkok

Chief Complaint: 3-year-history of hypopigmented patches on face

Present Illness: A Thai boy presented with 3-year-hstory of non-itchy, painless hypopigmented patches at left tibia which gradually extending. 3 weeks ago, He noticed new hypopigmented patches on right tibia

Past History: ADHD (Attention Deficit Hyperactivity Disorder), learning disability, obesity

Family History: nil

Current Medication: Concerta 36 mg/day

Dermatological Examination (Figure 12.6)**:** Multiple well-defined hypopigmented round patches with fine scale at both lower legs

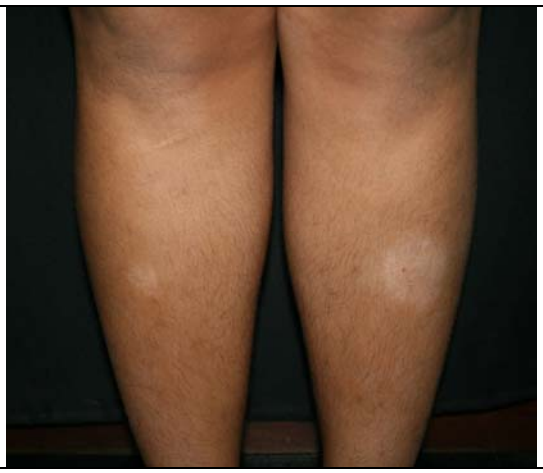

Figure 12.6

Histopathology (S10-2182) (Figure 12.7):

- superficial and deep perivascular, perifollicular and lichenoid infiltrate of lymphocytes
- infiltrate of some atypical lymphocytes in the thickened papillary dermis with epidermotropism

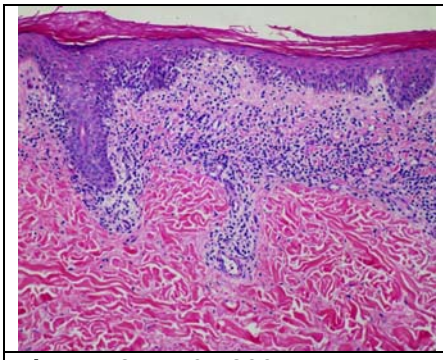

Figure 12.7, H&E 200x

Case 12.3

Patient: A 11-year-old Thai girl from Bangkok

Chief Complaint: 3-month-history of multiple hypopigmented patches on buttocks and both thighs

Present Illness: An 11-year-old Thai girl had multiple non-icthy gray-violaceous macules on trunk and both extremities for 2 years. She had been tried with oral antihistamine and topical corticosteroids twice daily which seemed to be worsened, rash extended to face. Although KOH was negative, she was treated as tinea vesicolor. Her first skin biopsy was done a year ago and compatible with Ashy's dermatosis. During the past 3 months, she noticed multiple slow growing, asymptomatic hypopigmented macules on buttocks and both thighs.

Past History: Healthy

Family History: nil

Physical Examination: unremarkable

Dermatological Examination: At first presentation (Figure 12.8-10), generalized multiple discrete gray-violaceous patches on face, trunk and both extremities. During the treatment as Ashy's dermatosis (Figure 12.11), generalized multiple discrete hypopigmented patches on buttocks and thighs.

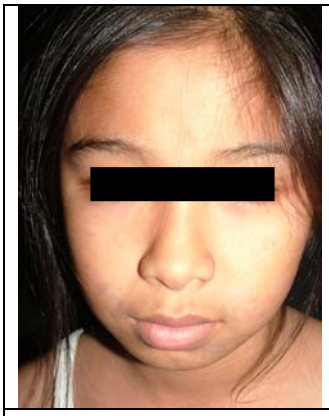

Figure 12.8

Figure 12.9

Figure 12.10

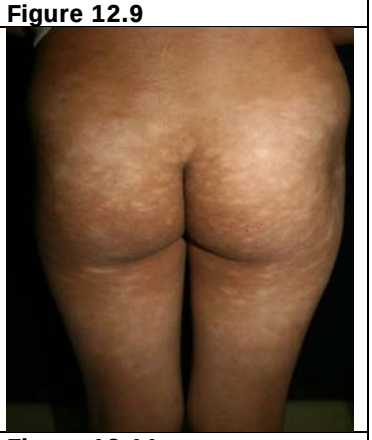

Figure 12.11

Histopathology (S10-8624) (Figure 12.12)**:** Moderately dense, superficial perivascular infiltrate of lymphocytes with some melanophages and foci of exocytosis

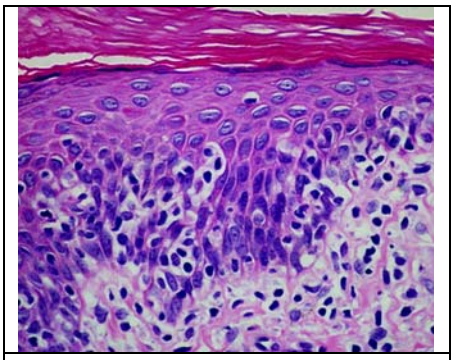

Figure 12.12, H&E 400x

Diagnosis: Hypopigmented mycosis fungoides

Presenter: Wanjarus Roongpisuthipong

Consultant: Natta Rajatanavin

Treatment: NV-UVB phototherapy
Topical corticosteroids

Discussion:

Mycosis fungoides (MF) is a malignant neoplasm of T lymphocytes that presents in the skin. The natural history is that of a chronic, slowly progressive disorder. The typical presentation consists of scaly macules, patches, plaques, nodules, tumors and erythroderma.[1]

Hypopigmented MF has been described over the last 4 decades[2,3] and has been reported over 100 cases in the literature.[4] The average age is 28-35 years at the disease onset, with an average of 5.5-5.7 years' duration of illness before presentation. Most of patients are dark-skinned individuals or Fitzpatrick skin type IV/ V and rare in Caucasians

or other skin types.[5] In a recent study in Pakistan, 21.3 percent of the MF patients[6] and 59 percent of the MF pediatric patients in Canada have the hypopigmented variety.[7] The nature history of hypopigmented MF is similar to that of conventional MF. Scattered hypopigmented macules or large patches on the trunk and the extremities are the usual presentation. Asymptomatic, nonatrophic, and nonscaly lesions with normal sensation seem to be the rule, although itching may occur. Patients are usually healthy. The differential diagnosis includes conditions such as vitiligo, pityriasis alba, leprosy, postinflammatory hypopigmentation, sarcoidosis, and pityriasis lichenoides chronic.[8]

Histology examination usually shows epidermotropism of atypical lymphocytes at dermoepidermal junction or sometimes forming Pautrier microabscess in the epidermis.[9] In contrast to conventional MF, in which the neoplastic cells are CD4+ in the vast majority of cases, the neoplastic cells in hypopigmented MF have a CD8+ T-cell phenotype. A decreased number of melanocytes and decreased expression of CD117 was presented in hypopigmented MF.[10]

The mechanism of hypomelanosis in hypopigmented MF has been unclear. It has been postulated that decreased expression of CD117 (Stem cell factor receptor/KIT protein) and its downstream events in melanocytes may be initiated by cytotoxic effects of of melanosomal-antigen-specific CD8+ neoplastic T-cell, resulting in destabilization of CD117, and leading to dysfunction and/or loss of melanocytes [10]. In contrast, Goldberg et al purpose that the loss of pigment results from a defect in melanosome transfer from melanocyte to keratinocyte.[11]

There are many treatment options such as topical corticosteroids, topical nitrogen mustard, topical

carmustine, NB-UVB, PUVA, and electron beam radiotherapy. NB-UVB is a viable, comparably safe, and easily administered option. NB-UVB is less carcinogenic than PUVA.[12]

Our patients have typical history and nature of hypopigmented MF which are young, dark-skinned and slow progression. All patients present with hypopigmented macules or patches that are confirmed by history examination for hypopigmented MF. They have been responded well to NB-UVB and topical corticosteroids except the third patient who is scheduled to receive NB-UVB during school holidays.

References

1. Chalid A and Wolfram S. Cutaneous Lymphoma In: Klaus W, Lowell AG, Stephen IK, Barbara AG, Amy SP, David JL, eds. Dermatology in General Medicine 7th ed. New York, N.Y.: McGraw-Hill, 2008: 1386-1401.
2. Ryan EA, Sanderson KV, Bartak, et al. Can mycosis fungoides begin in the epidermis? a hypothesis. Br J Dermatol 1973;88:643-9.
3. Smith NP, Samman PD. Mycosis fungoides presenting with areas of cutaneous hypopigmentation. Clin Exp Dermatol 1978;3:213-6.
4. Ratrout JT, Nazer M, Ansari N. Hypopigmented mycosis fungoides in a twenty-year-old Saudi woman with fair skin. Indian J Dermatol 2006;51(2):115-7.
5. Stone ML, Styles AR, Cockerell CJ, et al. Hypopigmented mycosis fungoides: a report of 7 cases and review of the literature. Cutis 2001;67(2):133-8.
6. Shaikh ZI, Rahman SB. Clinicopathological spectrum of mycosis fungoides type cutaneous T-cell lymphoma. J Coll Physicians Surg Pak 2006;16(3):171-4.
7. Pope E, Weitzman S, Ngan B, et al. Mycosis fungoides in the pediatric population: report from an international Childhood Registry of Cutaneous Lymphoma. J Cutan Med Surg 2010;14(1):1-6.
8. Neuhaus IM, Ramos-Caro FA, Hassanein AM. Hypopigmented mycosis fungoides in childhood and adolescence. Pediatr Dermatol 2000;17(5):403-6.
9. Richard LS, Gary RK. Pigmentary disorders of the skin In: David EE, Rosalie E, Bernerr LJJ, eds. Lever's histopathology of the skin 9th ed. Philadelohia: Lippincott Williams and Wilkins, 2005: 705-14.

10. Singh ZN, Tretiakova MS, Shea CR, et al. Decreased CD117 expression in hypopigmented mycosis fungoides correlates with hypomelanosis: lessons learned from vitiligo. Mod Pathol 2006;19(9):1255-60.
11. Goldberg DJ, Schinella RS, Kechijian P. Speculations about the mechanism of hypopigmentation. Am J Dermatopathol 1986;8(4):326-30.
12. Gathers RC, Scherschun L, Malick F, et al. Narrowband UVB phototherapy for early-stage mycosis fungoides. J Am Acad Dermatol 2002;47(2):191-7.