Case Report of Hypopigmented Mycosis Fungoides in a Child

Nicha Jenmanachaiyakun MD.

ABSTRACT:

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PRANANGKLAO HOSPITAL, MINISTRY OF PUBLIC HEALTH, NONTHABURI, THAILAND.

Mycosis fungoides (MF) is a cutaneous T-cell lymphoma, which some variants manifest as generalized hypopigmentation of non-sun exposed area of skin. Classic MF presents with the typical evolution stages of patches, plaques and tumors. MF is common in older adults but rarely occurs in children.

We reported a case of 9-year-old Thai boy patient who presented with generalized hypopigmented patches and macules on face, trunk and both extremities. The histopathology was compatible with MF. There was no systemic involvement. The TNM staging was stage IB. The patient was treated with 28 sessions of narrowband UVB (NB-UVB) phototherapy with complete response, however recurrence occurred after 7 months of NB-UVB discontinuation.

Key words: Child, cutaneous T-cell lymphoma, hypopigmented mycosis fungoides

From: Pranangklao Hospital, Ministry of Public Health, Nonthaburi, Thailand.

Corresponding author: Nicha Jenmanachaiyakun MD., email: mzyynj@hotmail.com

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Case report

A 9-year-old Thai boy patient presented with 4 months history of generalized hypopigmented patches and macules on face, trunk and both extremities. He denied history of any systemic symptoms. No family members experienced similar condition. Physical examination revealed skin phototype IV with multiple hypopigmented patches and macules on face, trunk and both extremities with 22% body surface area involvement. Neither hepatosplenomegaly nor lymphadenopathy was observed. Complete blood count, liver function tests, renal function tests, Lactate dehydrogenase were unremarkable. Film chest x-ray was normal. Ultrasound whole abdomen worked up no retroperitoneal or as seen mesenteric adenopathy. A 4-mm punch biopsy from the right thigh was performed. The epidermis showed parakeratosis, spongiosis and irregular acanthosis. The mononuclear cells were scattered both single-cells and in nests in the epidermis. Some cells had large hyperchromatic nuclei with perinuclear halo. There were also atypical mononuclear cells with perinuclear halo arranged in linear pattern along the basal cell layer. The dermis showed interstitial, superficial and deep perivascular infiltration with mononuclear cells. Thus, the final diagnosis was consistent with hypopigmented mycosis fungoides (MF) ($T_2N_0M_0$; stage IB). The treatments of stage IB were narrowband UVB (NB-UVB) with overall of 28 sessions and 0.1% triamcinolone cream for 3 months with complete response, but recurrence occurred after 7 months.

Discussion

MF is the most common type of cutaneous T-cell lymphoma (CTCL). MF accounts for 50% of all primary cutaneous lymphomas[1]. Primary cutaneous lymphomas are more common than secondary lymphomas in childhood[2,3]. MF often occurs in older adults with the median age at diagnosis is 55-60 years. However, MF is rare in children and adolescents. The mean age of presentation of hypopigmented MF is between 11 and 14 years[1,4-6]. The study of 23 patients under the age of 18 years reported the onset of skin lesions is 9 years, and the diagnosis is 11 years (median age)[7].

One of the variants of MF is characterized as generalized hypopigmentation in hyperpigmented skin type. The initial skin lesions are on the buttocks and in non-sun exposed area of trunk and limbs[1]. Hypopigmented MF is the most common skin manifestation in children, but rarely in adults (Table 1)[8].

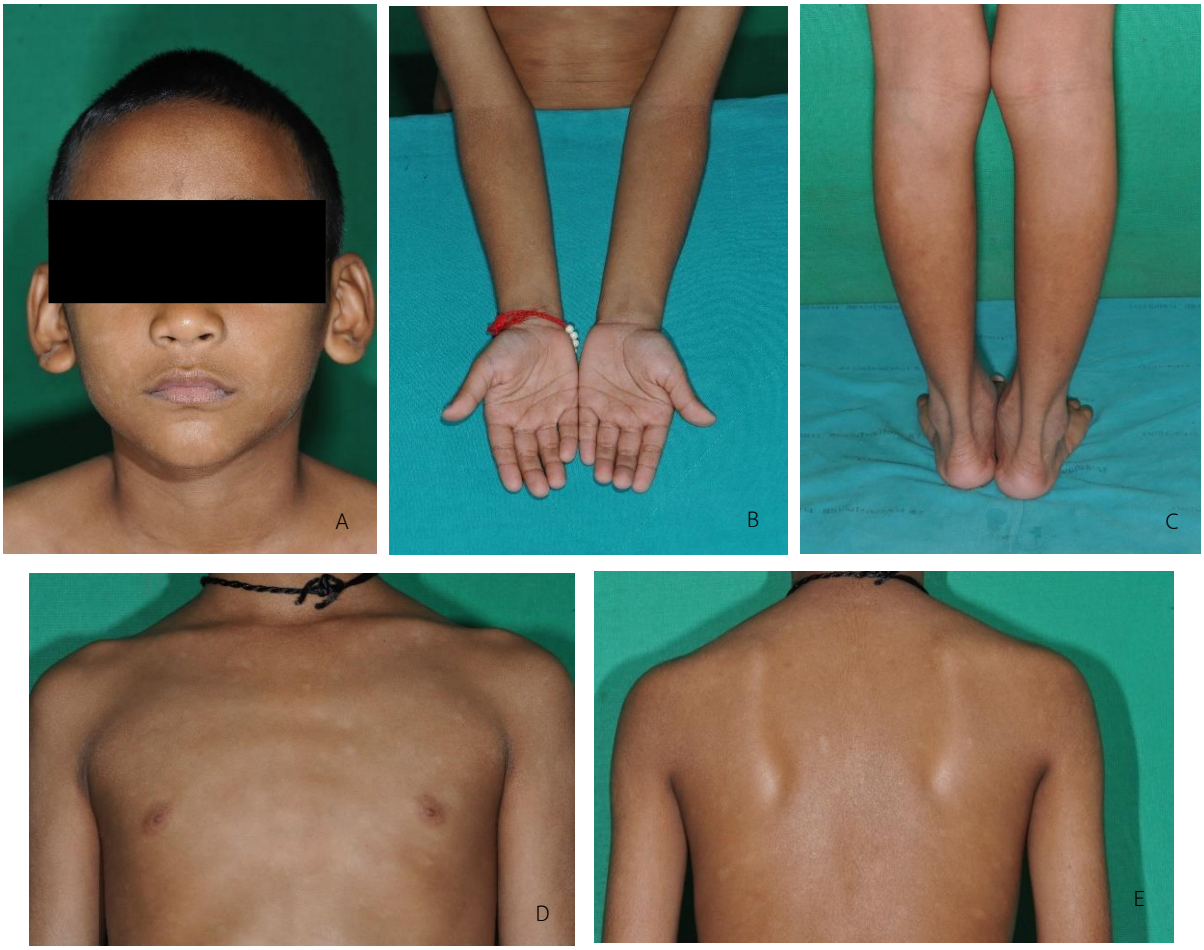

Figure 1 Multiple hypopigmented patches and macules on face (A), both extremities (B, C) and trunk (D, E)

Table 1 Variants of mycosis fungoides in adults versus children

Mycosis fungoides variants	Adults	Children
Classic	+++++	+
Hypopigmented	+	+++++
Folliculotropic	+	++
Unilesional	+	++

The diagnosis of MF is delayed due to similar clinical skin manifestations as pityriasis alba, pityriasis versicolor, vitiligo and atopic dermatitis[9]. More than 20% of children is initially diagnosed

and treated as other skin diseases[10]. MF is diagnosed by pathology. The characteristic features of MF are the presence of epidermotropism, intraepidermal nests of atypical cells (Pautrier microabscesses), mild or absent spongiosis[1].

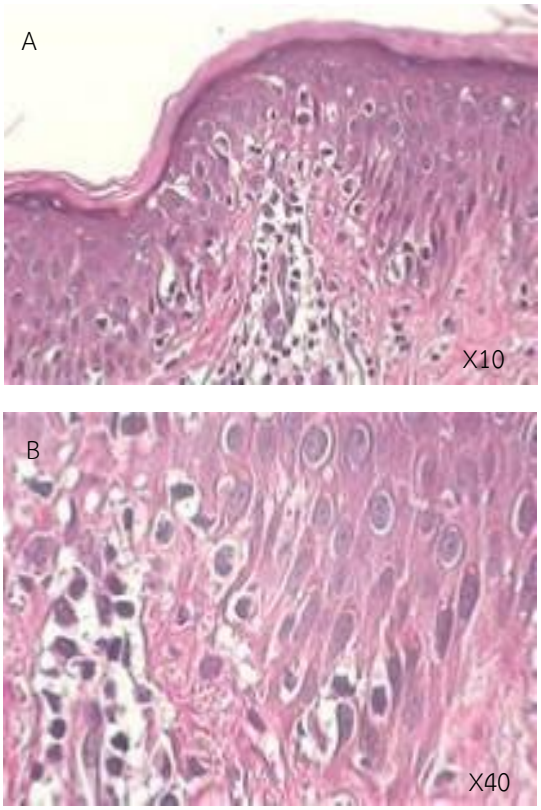

Figure 2 (A) The epidermis showed parakeratosis, spongiosis and irregular acanthosis. The mononuclear cells were scattered both single-cells and in nests in the epidermis
(B) Some cells had large hyperchromatic nuclei with perinuclear halo. There were also atypical mononuclear cells with perinuclear halo arranged in linear pattern along the basal cell layer

Treatment of MF is depending on TNM staging. Stage IB is the most common representation in children and adolescents, rarely seen in stage II to IV[7]. There are no specific management guidelines of MF in children. The children mostly receive the same management as adults depending on the severity of the disease. Treatments of stage IA-IIA consist of topical corticosteroids and phototherapy, which are NB-UVB, psoralen-UVA (PUVA) and UVA 340-400 nm (UVA-1)[11,12]. PUVA is more potential in complete response than NB-UVB in the management of early stage MF. Complete response rate of PUVA is 74%, while NB-UVB is 62%. Although, there is no difference in general response between PUVA and NB-UVB. However, PUVA has more side effects than NB-UVB with the hazard ratio 1.93[13]. Adverse effects of PUVA in children are cataract[14]. PUVA is not recommended in children younger than 12 years[15]. The treatment response to NB-UVB is lower for the folliculocentric MF. Therefore, PUVA is more appropriate for the folliculocentric MF variant[10]. Rates of complete remission are comparable in the different skin phototypes (81.2%, 71.4%, and 75% in skin phototypes III, IV, and V) respectively[16]. Comparing between light to dark skinned patients, the results suggest that NB-UVB is almost equally effective in treating early stage of MF, but more treatment sessions and higher cumulative NB-UVB doses are needed, especially in skin phototype V[16].

The study of 23 children and adolescents reported the disease did not progress to advanced stages in patients who remained follow-up. The longest follow-up was 10 years[7]. However, the disease progression was 5% at 5 years and 29% at 10 years in a retrospective study of 34 patients with juvenile-onset MF[17].

The prognosis of MF is good in children, with survival rates of 95% and 93% at the age of 5 and 10 years, respectively[17]. The progression factor of MF is clinical variants of MF. The hypopigmented or poikilodermatous MF have less disease progression than other variants[18].

We reported a case of hypopigmented MF in which the clinical features and histology fit into the diagnosis. However, the patient had rapid recurrence in 7 months. Hypopigmented MF is a common variant, but delayed diagnosis is also common. Therefore, early diagnosis improves outcomes.

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