CASE 14

Patient: A 46-year-old Thai male from Bangkok

Chief Complaint: Polymorphic asymptomatic skin eruptions for 3 weeks

Present History: The patient presented with 3-weeks of polymorphic asymptomatic skin eruptions, which initially occurred at trunk and rapidly increased in number extend over the entire body. He had not febrile or any illness preceded the onset of rash. The patient denied recent medicine ingestions.

Past History: He had neither underlying disease nor other medication.

Family History: His family history was unremarkable for dermatologic disease.

Dermatological Examination (Figure 14.1-4)**:** Multiple scattered, round to ovoid, erythematous papules covered with fine scale, vesicles, and superficial erosion with hemorrhagic crust. The lesions extended cover the entire body.

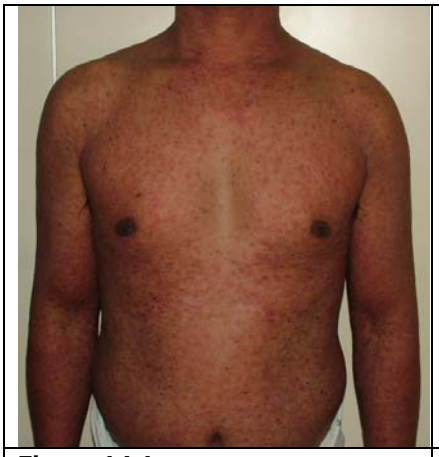

Figure 14.1

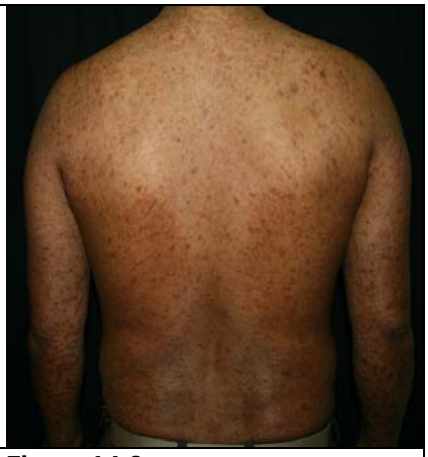

Figure 14.2

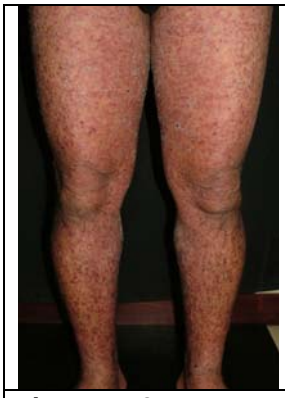

Figure 14.3

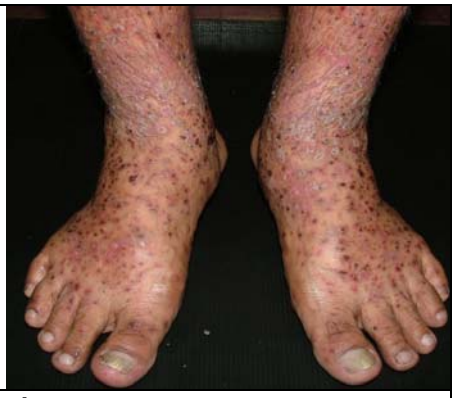

Figure 14.4

Physical examination: Unremarkable

Histolopathology (S10-1478B) (Figure 14.5)**:**

- Mounds of parakeratosis, occasional necrotic keratinocytes lymphocytic infiltrate obscuring the dermoepidermal junction and within epidermis
- Superficial and deep perivascular and focal lichenoid infiltrate of lymphocytes and some melanophages in the dermis

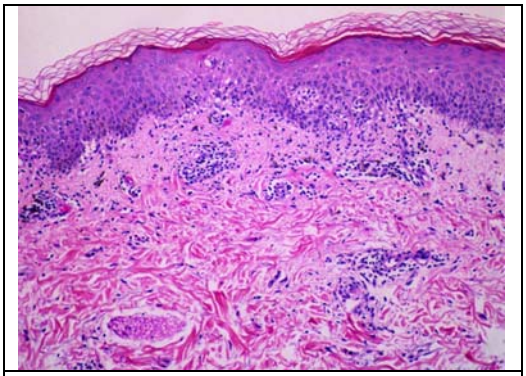

Figure 14.5, H&E 100x

Investigations: Routine laboratory test were within normal limits, except for an elevated ESR. Anti-HIV was negative.

Diagnosis: Pityriasis lichenoides et varioliformis acuta (PLEVA)

Treatment: 0.1% triamcinolone cream apply lesion, bid
Tetracycline 500 mg/day orally
Prednisolone orally
Methotrexate 7.5-10 mg/week orally
Narrow-band UVB 3 times/week

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Discussion:

Pityriasis lichenoides is a unique group of inflammatory skin disorders that include pityriasis lichenoides et varioliformis acuta (PLEVA) and pityriasis lichenoides chronic (PLC). It is generally accepted that PLEVA and PLC represent two end of a continuous disease spectrum. Therefore, it is not uncommon to observe both acute and chronic lesions in the same person.[1] Pityriasis lichenoides is more common in children and young adults with male predominance of 1.5:1 to 3:1.[2] The etiology of pityriasis lichenoides is unknown. It is speculated to be an inflammatory reaction triggered by certain infectious agents, an inflammatory response secondary to T-cell dyscrasia, or an immune complex-mediated hypersensitivity.[1]

PLEVA is characterized by 2-3 mm. diameter erythematous macules that quickly evolved into papules with a fine micaceous scale. The papule often has a central punctum that becomes vesiculopustular, hemorrhagic necrosis, and ulceration. The eruption is polymorphous, as lesions exist in all stage of development. Varioform scars and postinflammatory hyper- and hypopigmentation may result. PLEVA most often

occurs at the trunk, extremities, and flexural areas, but diffuse and generalized patterns as seen in our patient may also occur.[3] The more severe ulcerative variant of PLEVA is known as “Febrile ulceronecrotic Mucha-Habermann disease”.[4]

Histopathologic finding of PLEVA revealed focal parakeratosis, dyskeratosis, epidermal spongiosis and exocytosis of lymphocytes in the malpighian layer, as well as vacuolar degeneration of the basal layer, occasional epidermal vesicles, focal epidermal necrolysis. The dermis showed a moderately dense lymphohistiocytic perivascular infiltrate, often wedge-shaped and obscuring the dermoepidermal junction. Extravasation of erythrocytes and lymphocytic vasculitis can exhibit.[5] Immunohistochemistry of PLEVA shows a predominance of $CD8^+$ T cells.[6]

In the majority of PLEVA patients, the prognosis is good. The disorder may resolved spontaneously within a few months or, less commonly, persist for years. PLEVA usually has a shorter duration than PLC. PLEVA may transition into the chronic form, PLC.[7] However, despite the benign nature of PLEVA, long-term follow up is recommended because some rare cases progressively evolved to mycosis fungoides, particularly coexistent with poikiloderma.[8]

Several treatment modalities have been used. First-line therapy includes topical corticosteroids, antibiotic (erythromycin, tetracycline, minocycline), phototherapy (sunbathing, UVA+UVB, NUVB). Topical tacrolimus, prednisolone, methotrexate, photherapy (UVA1, PUVA), cyclosporine or oral retinoids are recommended as a second-line treatment. [9, 10, 11]

Our patient was initially treated with topical corticosteroids and tetracycline 500 mg/day orally for 2 weeks that proved insufficient to crease disease evolution.

Prednisolone 30-40 mg/day and methotrexate 7.5 mg/week were replaced. Response to this combination scheme was satisfactory, with partial clearance of the new lesions. Unfortunately, the disease was worsening when dosage of prednisolone was reduced. Additional therapy with narrow-band UVB (3 times per week) was started. After 4 weeks of NUVB therapy, prednisolone was gradually decreased to 5 mg/day, while methotrexate was continued at dose 10 mg/week. At present, treatment combination of NUVB, low dose prednisolone, and methotrexate can control disease without evolution of new lesion.

References

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