Case 1

A 17-year-old Thai woman from Bangkok

Chief complaint: Recurrent pruritic erythematous eruptions with residual reticulated hyperpigmentation on her back, chest, and abdomen for 4 years.

Present illness: The patient presented with a 4 year-history of recurrent pruritic lesions over her back, chest, and abdomen which were triggered by emotional stress, menstrual cycle, and swimming. The pruritic lesions usually persisted for 4 days leaving a net-like hyperpigmentation after. The patient had been treated with oral steroid without any improvement.

Past history:

No history of underlying disease

Family history:

None of her family had history of similar skin lesion.

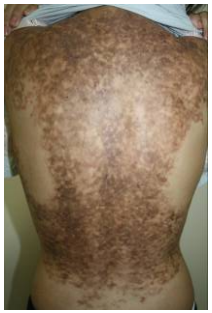
Skin examination:

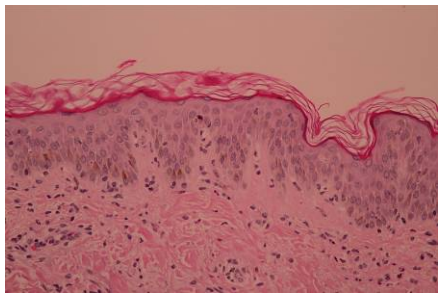
Generalized discrete erythematous papules distributed over the chest, abdomen, and back with large areas of reticulated hyperpigmented patches.

Histopathology (S11-07797)

Perivascular and interstitial inflammatory-cell infiltrate of lymphocytes, admixed with numerous neutrophils and some melanophages in the superficial and mid dermis.

Scattered necrotic keratinocytes with some exocytosis of lymphocytes.





Diagnosis: Prurigo pigmentosa

Treatment: Doxycycline 100mg daily

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Discussion:

Prurigo pigmentosa (PP) is a rare inflammatory dermatosis of unknown aetiology. It was first reported in 1971 by M. Nagashima from Japan, where it is most commonly found. This disease is rare in western countries. The clinical features may include symmetrical and intensely pruritic erythematous papules, papulovesicles and vesicles appearing in a reticular pattern with a predilection for the upper part of the back, neck, shoulders, lumbosacral region and abdomen. The lesions involute in a matter of days leaving reticulate pigmentation[1].

The histopathological findings vary at the different stages. In the early stage, a superficial perivascular infiltration of neutrophils appears and spreads into the edematous papillary dermis. The neutrophils move swiftly through the epidermis, with the accompanying of spongiosis, necrotic keratinocytes or intraepidermal abscess. In fully developed lesions, lymphocytes and eosinophils predominate over neutrophils in both the dermis and epidermis, which resemble as a patchy, lichenoid pattern. Spongiosis and vacuolar degeneration of the basal layer may lead to intraepidermal vesiculation or subepidermal blisters. In the later stage, as lymphocytes become predominant with perivascular infiltration, the epidermis becomes hyperplastic and parakeratotic. The melanophages appear in the papillary dermis causing hyperpigmentation in the final stage.[1-3]

The aetiology of PP remains unclear. The environmental triggering factors may include sunlight, sweating and friction from clothing. It has also been suggested that the disease is a result of allergic reactions from chemical contactants such as para-amino compounds, trichlorophenol or chromium[4]. In addition to the above mentioned exogenous factors, some cases are reported to be related to ketosis resulting from diabetes mellitus, fasting, dieting or anorexia nervosa. Atopy[5], pregnancy and menstruation[6] have also been reported to be associated with PP. The Worsening PP during menstruation (as in this case), especially in young females and pregnancy suggests a possible correlation between hormone and the disease. Hence the role of oestrogen in patient with PP warrants further investigation.

The differential diagnosis during the active stage may include dermatitis herpetiformis, linear IgA dermatosis, and acute lupus erythematosus. The reticulate pattern of pigmentation in its final stage must be distinguished from confluent and reticulate papillomatosis of Gougerot and Carteaud.

Minocycline, doxycycline, dapsone, sulfamethoxazole, and macrolides are the effective treatments for PP. IL-6 in PP skin lesions may explain the effects of these drugs in terms of their anti-inflammatory properties[7]. Low dose oral isotretinoin may improve both the erythematous lesions in the acute stage, and the hyperpigmentation in the later stage[8]. Oral antihistamine, oral and topical steroids have failed to show any treatment benefit[7].

References

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