Prurigo Pigmentosa: A Report of Two cases In Malaysia

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Introduction
Prurigo pigmentosa (PP) is a rare inflammatory dermatosis of unknown etiology described first by Nagashima et al in 1971. It is characterized by a network of pruritic, erythematous papules that evolves into reticulated hyperpigmentation with predilection for the neck, chest and back. The disease is rarely diagnosed outside Japan. Here we present two patients with prurigo pigmentosa in Malaysia.

Case 1
An 47 year old Malay lady presented with non-pruritic progressive hyperpigmented patches over her anterior neck and cubital fossa for 6 months. On examination, there were well-defined reticulated hyperpigmented patch on her neck extending up to the chin and cubital fossa with no surface changes (Figure 1a-c ). Our differential diagnoses were prurigo pigmentosa and lichen planus pigmentosus. Fasting blood sugar was normal. Skin biopsy was done and histopathological examination showed mild epidermal acanthosis with hyperkeratosis, foci of mild spongiosis with basal cells vacuolar degeneration. Lymphocytes exocytosis was also present. The superficial dermis showed moderate perivascular lymphocytic cells infiltration with melanophages and melanin incontinence. The features were in keeping with prurigo pigmentosa. She was treated with tablet doxycycline 100mg OD for 3 months.

Case 2
An 18 year old Malay student presented with itchy, erythematous papules that progressed to hyperpigmented reticulated patches over her back for 2 years. It was aggravated by heat, sunlight and sweating. She had no systemic symptoms. On examination, there were scattered erythematous papules with reticulate hyperpigmented patches over her back (Figure 2a-d). Differential diagnoses were prurigo pigmentosa, reticular erythematous mucinosis, and subacute cutaneous lupus erythematosus. Investigations showed negative ANA, and other results (FBC, RP, LFT, FLP, FBS, and ESR) were normal. Skin biopsy showed sections of focal parakeratosis, exocytosis, spongiosis and hyperkeratosis with formation of colloid bodies. The upper dermis and perivascular area also showed some lymphohistiocytic infiltration and occasional eosinophils. There was no mucin deposition and immunofluorescence was negative. The features were compatible with prurigo pigmentosa. She was treated with tablet doxycycline 100mg OD for 10 months. After initiation of treatment, she has no new lesions.

Discussion
Prurigo Pigmentosa (PP) is a rare inflammatory disease characterized by recurrent pruritic eruptions of erythematous papules that evolve into urticarial plaques and eventually leaving reticulate hyperpigmentation. The pathogenesis of PP remains unclear. Several hypotheses have been proposed, including exogenous factors like friction with clothes and allergic reaction to chemical agents like chromium. Recent reports have demonstrated the associations of this disease with diabetes mellitus, ketosis, fasting, dieting, and pregnancy.

PP displays stage specific histopathological features, with an initial superficial perivascular neutrophils infiltration, followed by scattered spongiosis, necrotic keratinocytes and epidermal ballooning. Lymphocytic infiltrates predominates and distributes in patchy lichenoid pattern in fully developed lesions. There would be profound liquefactive degenerations of basal layer with pigmentary incontinence. In later stage, the epidermis becomes hyperplastic with parakeratosis. Biopsy finding alone are not specific enough to make the diagnosis, thus, correlations of clinical and histopathological are important.

The mainstays of treatment are tetracycline class of antibiotics and dapsone. Doxycycline at doses of 50mg to 100mg BD and dapsone at doses of 25mg to 100mg OD has been used with great efficacy. Both treatments are thought to be effective by inhibiting neutrophils chemotaxis. Topical and systemic corticosteroids or antihistamines are not beneficial in treating PP.

Conclusion
Due to the rarity of its presentation, underdiagnosis or misdiagnosis of this condition remains a distinct possibility. In view of the rarity of this condition, a high index of suspicion is required to make the diagnosis of prurigo pigmentosa. It should be considered in any patient presenting with acquired pigmentary disorder. Correlation with clinical and histopathological findings is essential to make the correct diagnosis.

Reference