Case 22

Rash with residual reticulated hyperpigmentation

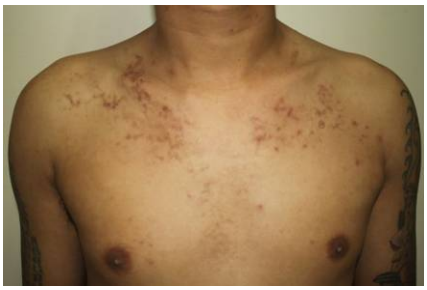
Patlada Ingkaninanda, M.D.
Penpun Wattanakrai, M.D.

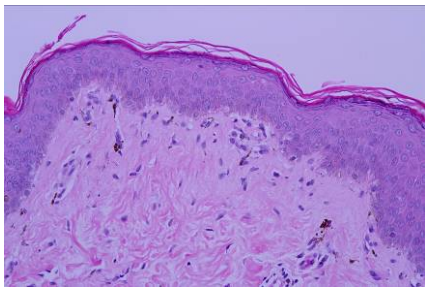
Patient: A 28-year-old man from Bangkok

Chief complaint:
Recurrent erythematous papules on the chest and upper back for 5 years

Present illness:
The patient had a 5-year history of asymptomatic recurrent erythematous papules on his chest and upper back. The lesions spontaneously subsided without treatment with residual reticulated hyperpigmentation.

Past history: No underlying disease





Skin examination:
- Multiple symmetrical reticulated erythematous to brownish macules and papules coalescing to form patches and plaques on chest and upper back

Histopathology: (S14-16793, upper chest)
- Superficial inflammatory cell infiltrate of numerous melanophages in the dermis

Diagnosis: **Prurigo pigmentosa**

Treatment:
- Doxycycline 200 mg/day

Discussion:
Prurigo pigmentosa (PP) is a rare inflammatory skin disease first described by Nagashima in 1971.[1] PP is characterized by a markedly pruritic eruption of erythematous papules and papulovesicles arranged in reticular pattern distributed symmetrically on the back, neck, chest, and lumbosacral region.[2] Crops of inflammatory lesions develop rapidly and then involute within a week, leaving macular reticulated hyperpigmention. The disease has a fluctuating course with exacerbations and recurrences. Recurrences tend to occur at the same site, and lesions in multiple stages are often evident. Pruritus is usually severe in early lesions, but resolving lesions may be devoid of

symptoms. PP is common in young adults, with a female:male ratio of ≥ 2:1. The majority of reported patients have been from Japan, followed by Korea.[1-4]

The cause and pathogenesis of PP are still unclear. Many factors have been implicated in the pathogenesis including; sunlight, sweating, friction, contact allergens such as para-amino compounds, trichlorophenol, chromium and nickel, drug use (bismuth subsalicylate-containing antacid), weight loss (by fasting, dieting, anorexia nervosa). Other conditions reported in PP are insulin dependent diabetes mellitus, ketosis, pregnancy, *Helicobacter pylori* infection, primary biliary cirrhosis with Sjögren's syndrome, and atopic diathesis but none of them showed consistent association.[2-5]

The differential diagnosis of PP includes confluent and reticulated papillomatosis of Gougerot and Carteaud. However, keratosis is not a clinical feature of prurigo.[6]

Three histopathologic stages have been identified in PP. (1) Early lesion demonstrates neutrophilic exocytosis, spongiosis, papillary dermal edema and a superficial perivascular infiltrate of neutrophils. (2) Fully developed lesion, an intra-/subepidermal vesiculation, necrotic keratinocytes and a patchy lichenoid infiltrate of predominantly lymphocytes. (3) Late-stage PP shows parakeratosis and dermal melanophages.[2]

Treatment with oral minocycline, doxycycline or dapsone is usually effective for the inflammatory component of PP but it is not effective for the pigmentation. These agents inhibit migration and function of neutrophils and they are helpful in patients with improvement of pruritus and inflammatory papules usually evident within a few days.[2,5] Treatment with a mean 2.4 week course of doxycycline at a dose of 200 mg/day has been reportedly effective in most cases both in initial and recurrent episodes of the disease.[5] Low-dose isotretinoin 0.3 mg/kg/day (20 mg/day) has been reported to improve erythematous lesions, and also helps resolve the reticular hyperpigmentation of PP.[7]

Our patient was diagnosed with PP from clinical presentation and histopathological features. He was treated with doxycycline 200 mg/day with good response. Although far more prevalent in Japanese and young females, PP can occur in Thai males and may be asymptomatic as demonstrated in our patient. Physicians should consider a diagnosis of PP for patients presenting with a reticulated pruritic eruption with hyperpigmentation symmetrically distributed over the trunk.

References:

1. Nagashima M. Prurigo pigmentosa--clinical observations of our 14 cases. J Dermatol. 1978; 5(2): 61-7.
2. Böer A, Misago N, Wolter M, Kiryu H, Wang XD, Ackerman AB. Prurigo pigmentosa: a distinctive inflammatory disease of the skin. Am J Dermatopathol. 2003; 25(2): 117-29.
3. Shin JW, Lee SY, Lee JS, Whang KU, Park YL, Lee HK. Prurigo pigmentosa in Korea: clinicopathological study. Int J Dermatol. 2012; 51(2): 152-7.
4. Kim JK, Chung WK, Chang SE, Ko JY, Lee JH, Won CH, et al. Prurigo pigmentosa: clinicopathological study and analysis of 50 cases in Korea. J Dermatol. 2012; 39(11): 891-7.
5. Lu PH, Hui RC, Yang LC, Yang CH, Chung WH. Prurigo pigmentosa: a clinicopathological study and analysis of associated factors. Int J Dermatol. 2011; 50(1): 36-43.
6. Asgari M, Daneshpazhooh M, Chams Davatchi C, Böer A. Prurigo pigmentosa: an underdiagnosed disease in patients of Iranian descent? J Am Acad Dermatol. 2006; 55(1): 131-6.
7. Akoglu G, Boztepe G, Karaduman A. Prurigo pigmentosa successfully treated with low-dose isotretinoin. Dermatology. 2006; 213(4): 331-3.