

Case: 1

A 14-year-old Thai boy from Phrae province

Chief complaint:

Skin lesions on the chest and back for 1 month

Present illness:

The patient presented with mild pruritic skin lesions that spread from his back to the chest for 1 month. He had been treated with 0.1% Triamcinolone acetonide cream and ketoconazole cream without improvement. The lesions progressed and leaving hyperpigmentation.

Past illness:

No history of underlying disease.

Family history:

None of his family had history of similar skin lesion

Physical examination

Skin : large area of excoriated, erythematous papules confluent to plaque and few papulovesicles on the upper back, interscapular, nuchae chest and clavicular area. The lesions turned to hyperpigmentation in a reticular pattern. (Fig. 1.1, 1.2)



Fig. 1.1



Fig. 1.2

Laboratory investigation:

KOH preparation: negative for fungus

Histopathology : (S09-006073A)

- Necrotic keratinocytes, spongiosis, with intraepidermal vesicle and pustule in the epidermis.
- Dense superficial perivascular infiltrate of lymphocytes, neutrophils and eosinophils in the dermis and exocytosis to the epidermis.

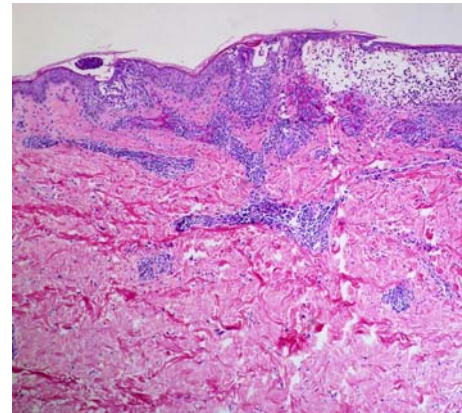


Fig. 1.3

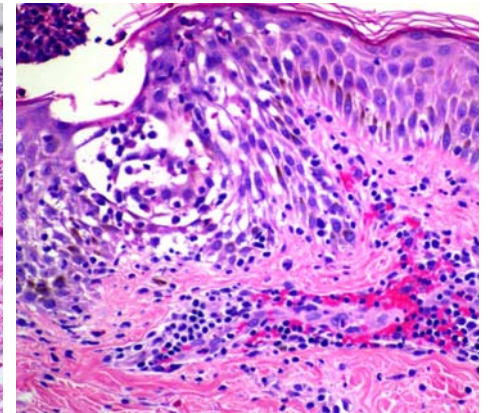


Fig. 1.4

DIF: non-specific feature

Diagnosis : Prurigo pigmentosa

Presenter : Suthida Chaithirayanon

Consultant : Amornsri Chunharas

Treatment : Doxycycline 100 mg OD., Ceterizine 10 mg OD.

Discussion:

Prurigo pigmentosa is a distinctive inflammatory disease first described by the Japanese dermatologist Masaji Nagashima in 1971. Pathogenesis is unknown. It is characterized by an inflammatory phase with pruritic erythematous papules, papulovesicles, vesicles and a resolution phase with reticulated hyperpigmentation. However, the bullous form has been reported in literature.¹ The distribution of the lesions is on the back, including the upper back and scapular regions, nuchae, clavicular regions, and chest. However, the lesions may occur on other sites, such as the abdomen, lumbosacral regions, antecubital fossae, limbs, and forehead. The mucous membranes, hair and nails are spared.² It is more common among young adult females.^{3,4} Mostly occur in the spring and summer.⁴

Differential diagnosis in the early stage is dermatitis herpetiformis and linear IgA dermatosis. The resolving stage is lichen pigmentosus, pigmented contact dermatitis, erythema dischroicum perstans and reticulated papillomatosis of Gougerot and Carreaud, but in contrast to those lesions, pigmented macules of prurigo pigmentosa are not keratotic.⁵

Histology study in the early stage shows a sparse superficial perivascular and interstitial dermatitis infiltrate with neutrophils in the epidermis to poorly formed microabscesses. The degree of spongiosis ranges from slight to marked. Necrotic keratinocytes are present in conjunction with scattered neutrophils and spongiosis in the epidermis. Lymphocytes are sprinkled along the dermo-epidermal junction at the same time when neutrophils dominated the upper layers of the epidermis. In a fully developed lesion, a patchy lichenoid pattern infiltrate with lymphocytes and eosinophils more than neutrophils. Ballooning is more prominent than spongiosis. In a resolving lesion, parakeratosis and scale crust at the cornified layer. Few to many melanophages in the papillary dermis and the upper part of the reticular dermis. Direct immunofluorescence studies gave non-specific results.^{5,6}

Treatment is usually with oral dapsone with dosage schedules as low as 25 mg daily and as high as 100 mg daily.⁷ Sulfamethoxazole or minocycline is effective due to the mechanism of inhibit migration and function of neutrophils. Doxycycline has been reported to be effective too.⁵ Low-dose isotretinoin in addition helps resolve the reticular hyperpigmentation of prurigo pigmentosa.⁸ However, oral antihistamine and topical corticosteroid show no effect as in this case.

References:

1. Yumigo K, Tetsuya, Juichiro N. Bullus prurigo pigmentosa and diabetes. *Eur J Dermatol* 1998; 8:439-41
2. B-aykal C, Buyukbabani N, Akinturk S, Saglik E. Prurigo pigmentosa: not an uncommon disease in the Turkish population. *Int J Dermatol* 2006; 45(10): 1164-8
3. Boer 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.
4. Gur-Toy G, Gungor E, Artuz F, Aksoy F, Alli N. Prurigo pigmentosa. *Int J Dermatol* 2002; 41(5):288-91.
5. Boer A, Asgari M. Prurigo pigmentosa: An underdiagnosed disease? *Indian J Dermatol Venereol Leprol* 2006; 72(6):405-9.
6. Boer A, Misago N, Wolter M, Kiryu H, Wang XD, Ackerman AB. Prurigo pigmentosa: A distinctive inflammatory of the skin. *Am J Dermatopathol* 2003; 25(2):117-29.
7. Joyce AP, Horn TD, Anhalt GJ. Prurigo pigmentosa. Report of a case and review of the literature. *Arch Dermatol* 1989; 125(11):1551-54.
8. Akoglu G, Boztepe G, Karaduman A. Prurigo pigmentosa successfully treated with low-dose isotretinoin. *Dermatology* 2006; 213(4):331-3.