Vesicular prurigo pigmentosa with mucosal involvement

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ABSTRACT:

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Prurigo pigmentosa (PP) is a rare inflammatory skin disease of distinct entity, characterized by sudden onset, symmetrical, pruritic erythematous papules, usually distributed over the trunk and nape of neck. The resolutions usually leave reticulated hyperpigmentation. PP were mostly reported from Japan, with only few non-Japanese cases. We report a case of Thai male with vesicular PP, a very rare pattern of PP. Our patient also had oral involvement, which has never been reported in PP.

Keyword: vesicular, bullous, prurigo pigmentosa, mucosal involvement, reticulate hyperpigmentation

บทคัดย่อ:

ณัฐพร อริยธนสุภรณ์, นุชา เนียมประดิษฐ์, อาทิตย์ นาคะเกศ รายงานผู้ป่วย PRURIGO PIGMENTOSA ชนิดตุ่มน้ำที่มีรอยโรคที่เยื่อบุปากซึ่งไม่เคยมีรายงานมาก่อน วารสารโรคผิวหนัง 2556; 29: 232-235.

แผนกผิวหนัง โรงพยาบาลพระมงกุฎเกล้า

Prurigo pigmentosa เป็นโรคผิวหนังอักเสบที่พบได้ไม่บ่อย มีลักษณะเป็นผื่นคันที่เกิดขึ้นอย่างสมมาตรที่บริเวณลำตัวและต้นคอ เมื่อหายแล้วทิ้งรอยดำมีลักษณะเป็นร่างแห ซึ่งเป็นลักษณะจำเพาะของโรคนี้ โดยผู้ป่วยที่มาด้วยผื่นเป็นลักษณะตุ่มน้ำนำมาก่อนพบได้น้อยมากและยังไม่เคยมีรายงานในประเทศไทยมาก่อน ในรายงานนี้ได้นำเสนอผู้ป่วย

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ชายไทยอายุ 21 ปี ที่ได้รับการวินิจฉัยเป็นโรค prurigo pigmentosa ชนิดตุ่มน้ำ นอกจากนี้ ยังมีรอยโรคที่บริเวณเยื่อบุปาก ซึ่งโดยปกติจะไม่พบในโรค prurigo pigmentosa

คำสำคัญ: prurigo pigmentosa, ตุ่มน้ำ, รอยโรคที่เยื่อบุปาก, รอยด่างเป็นร่างแห

Introduction

Prurigo pigmentosa (PP) is a rare inflammatory skin disease, characterized by sudden onset, symmetrical, pruritic erythematous papules, distributed over the trunk and nape of neck, healed with reticulated hyperpigmentation.[1] Vesicular or bullous lesions are very rare. We report a case of vesicular PP who also had oral involvement, which has never been reported.

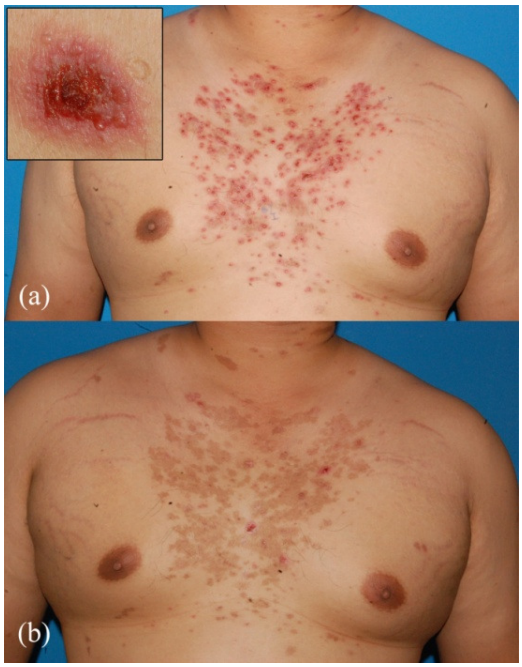

Figure 1 (a) Clinical presentation with vesicular lesions (b) Lesions healed with reticulate hyperpigmentation predominantly on the upper chest.

Case report

A 21 year-old healthy man presented with very itchy erythematous papules with small vesicles in the center and at the periphery, distributed symmetrically on mid upper chest (Figure 1a), upper back, chin, neck and concha areas of both ears for one month. A few small ulcers on erythematous base were found on the buccal mucosa with superficial erosions on the lower lip (Figure 2). Tzanck's smear of the vesicles and oral ulcers showed no acantholytic cells. Skin biopsy from the vesicles revealed subepidermal separation with numerous necrotic keratinocytes and dense band-like lymphocytic infiltration (Figure 3). Biopsy from oral ulcer was non-specific. Direct immunofluorescent examination was negative. Laboratory investigation including fasting plasma glucose, HbA1c, autoantibody profiles (ANA, anti-ds DNA) and viral profiles (EBV, CMV and HSV) were unremarkable. Treatment with topical corticosteroid and antihistamine were ineffective. New vesicles appeared while the older lesions showed necrotic crusts and healed with reticulated brownish hyperpigmentation (Figure 1b). Vesicular prurigo pigmentosa was diagnosed and doxycycline 100 mg twice daily was prescribed. After a few days, all lesions and oral ulcers were resolved.

However, relapse occurred after one month prompting the restart of doxycycline.

Figure 2 Few superficial ulcers on erythematous base on the oral mucosa

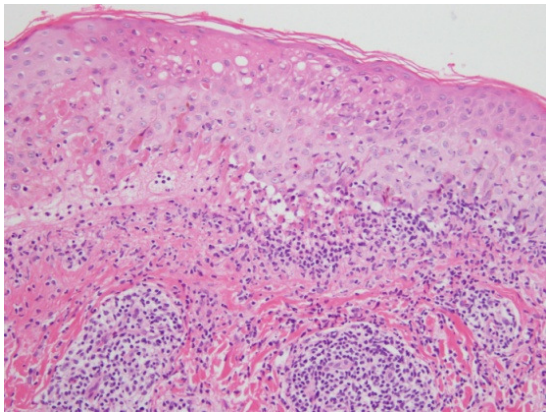

Figure 3 Histopathology of the vesicular lesion revealed subepidermal separation with numerous necrotic keratinocytes and dense band-like lymphocytic infiltrations (haematoxylin and eosin stain, x40)

Discussion:

Only a few cases of vesicular form of PP have been reported, mostly from Japan.[2-6] The pathogenesis of PP is unknown. The exogenous triggering factors are sunlight, sweating, friction, contact allergens while the endogenous factors include ketosis, fasting, dieting, diabetes mellitus, and pregnancy.[7-9] Diabetes mellitus and ketonuria were particularly associated with bullous PP.[3]

Histopathology of PP is rather non-specific. Böer et al. proposed that superficial perivascular and interstitial neutrophil infiltrations with epidermal spongiosis, ballooning and necrotic keratinocytes were found in early lesions. In fully developed lesions, eosinophils and lymphocytes subsequently predominated in the dermis. The late lesion showed hyperplastic epidermis with parakeratosis and hyperpigmentation along with melanophages in the upper dermis. Direct immunofluorescence was constantly negative.[7-9] In the vesicular form, intraepidermal or subepidermal separation with necrotic keratinocytes was observed. Dense, band-like lymphohistiocytic infiltration should also be seen along the dermo-epidermal junction.[5]

In our case, early lesions consisted of very itchy erythematous papules with vesicles. Our differential diagnoses included dermatitis herpetiformis, pemphigus herpetiformis and linear IgA bullous dermatosis. All of these diseases were ruled out by clinical and histopathological findings. Erythema multiforme (EM) was also included in the histopathological differential

diagnosis but there was no target lesion. Our histological sections were compatible with the severe form of PP. Laboratory investigations showed no other associated diseases. Interestingly, oral mucosal involvement has never been mentioned in any previous reports.

Generally, minocycline is the treatment of choice for PP with dramatic response.[10] Other systemic agents include dapsone, sulfamethoxazole, potassium iodide, and doxycycline.[11] Topical and systemic corticosteroids render modest benefit. Regarding the bullous form of PP, minocycline was effective.[3] One report showed a good response of vesicular PP to isotretinoin but the patient experienced clinical flare after discontinuation of the drug.[4]

Conclusions

We reported the first case of vesicular PP in Thailand with a good response to doxycycline. This case also had unusual oral involvement which has never been reported.

References

1. Böer A, Asgari M. Prurigo pigmentosa: an underdiagnosed disease? Indian J Dermatol Venereol Leprol 2006; 72: 405-9.
2. Kubota Y, Koga T, Nakayama J. Bullous prurigo pigmentosa and diabetes. Eur J Dermatol 1998; 8: 439-41.
3. Matsumoto C, Kinoshita M, Baba S, et al.: Vesicular prurigo pigmentosa cured by minocycline. J Eur Acad Dermatol Venereol 2001; 15: 354-6.
4. Requena Caballero C, Nagore E, Sanmartín O, et al. Vesiculous prurigo pigmentosa in a 13-year-old girl: good response to isotretinoin. J Eur Acad Dermatol Venereol 2005; 19: 474-6.
5. De Francesco V, Quinkenstein E, Mariuzzi L, et al. Bullous prurigo pigmentosa. Eur J Dermatol 2006; 16:184-6.
6. Kim SK, Kang HY, Lee ES. Bullous prurigo pigmentosa. Int J Dermatol 2007; 46: 888-90.
7. Böer A, Misago N, Wolter M, et al. Prurigo pigmentosa: a distinctive inflammatory disease of the skin. Am J Dermatopathol 2003; 25:117-29.
8. Lin SH, Ho JC, Cheng YW, et al. Prurigo pigmentosa: a Clinical and histopathologic study of 11 cases. Chang Gung Med J 2010; 33: 157-63.
9. Oh YJ, Lee MH. Prurigo pigmentosa: a clinicopathologic study of 16 cases. J Eur Acad Dermatol 2012; 26: 1149-53.
10. Aso M, Miyamoto T, Morimura T, et al. Prurigo pigmentosa successfully treated with minocycline. Br J Dermatol 1989; 120: 705-8.
11. Mok YJ, Lim KS, Tey HL. Doxycycline – an emerging therapy for prurigo pigmentosa. J Eur Acad Dermatol 2012; 26: 526-7.