

Case 21

A 19 year-old Thai female from Chanthaburi

Chief complaint: Recurrent multiple pruritic eruptions on the back and neck for 6 years



Fig. 21.1

Present illness:

The patient presented with a 6-year history of recurrent pruritic erythematous papules and plaques that resolved leaving behind reticulated hyperpigmented patches on her back and neck. Additionally, the lesions tend to erupt on the same site and be exacerbated by the summer season. Prior treatment with antihistamine and topical steroid were unsuccessful.

Underlying disease: Allergic rhinitis

Family history: No family history of similar cutaneous lesions

Dermatologic examination: (Fig. 21.1)

Multiple erythematous excoriated papules, interspersed among brownish reticulated hyperpigmented patches on the back and nape of the neck

Physical examination:

Other systemic examinations revealed no abnormality.

Investigations:

- **CBC**
 - WBC 11,610/mm³ (N 56, L 32, M 6, Eo 6, B 0%)
 - Hb 11.9 g/dL Hct 36.5 %, MCV 80 fL
 - Plt 421,000/mm³
- **Liver function test:** AST/ALT 15/7 U/L, ALP/GGT 73/12 U/L, TP/Alb 74/40 g/L, TB/DB 0.2/0.1 mg/dL
- **Renal function:** BUN/Cr 11/0.6 mg/dL
- **G6PD level:** normal

Histopathology: (S18-022114A, skin, back) (Fig. 21.2)

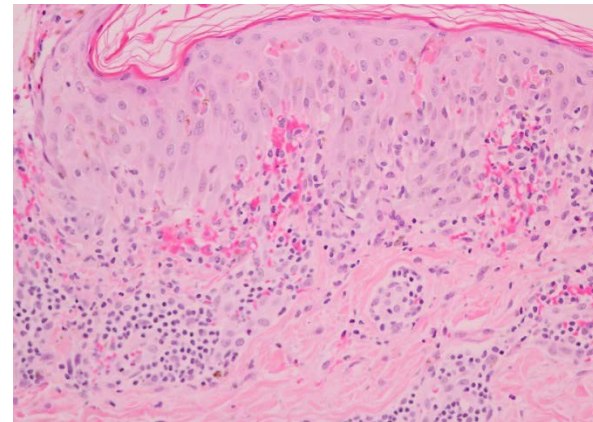


Fig. 21.2

- Intraepidermal vesiculopustules and numerous necrotic keratinocytes in the epidermis
- Lichenoid inflammatory cells infiltrate of lymphocytes, numerous extravasated erythrocytes, few eosinophils and melanophages in the superficial dermis

Diagnosis: Prurigo pigmentosa

Treatment

- Doxycycline (100) 1 tab twice daily for 2 months

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Discussion:

Prurigo pigmentosa (PP) also referred to as Nagashima’s disease is an uncommon, acquired inflammatory disorder with a predilection for the young Japanese woman.¹ Multiple cases of PP have been reported since its initial description in 1971 by Nagashima et al.² However, the condition has also been reported in men.³ and in individuals from other ethnic backgrounds.⁴ Although the pathogenesis of PP remains elusive, association with systemic conditions including adult-onset Still’s disease⁵, atopy⁶, H. pylori infection⁷, and Sjogren’s syndrome⁸ have been reported.

PP can be divided into three stages-early, fully developed, and late- each of which is distinguished by unique clinical and histologic features. Early-stage prurigo pigmentosa is characterized by pruritic urticarial papules or plaques that show a superficial perivascular neutrophilic infiltrate on pathologic examination. Patients with fully developed lesions present with crusted erythematous papules,

papulovesicles, and vesicles; spongiosis and numerous necrotic keratinocytes are the histologic hallmarks of this stage. Lastly, late-stage prurigo pigmentosa is characterized by the appearance of smooth-surfaced pigmented macules or netlike pattern. Histologic features of late-stage lesions include a predominantly lymphocytic infiltrate and melanophages in the papillary dermis.^{1, 9, 10} The preferred sites of involvement are the back, chest, and neck.² Clinical course of PP usually recurs over a period of months to years which upon resolution, leave behind a pattern of reticulated or mottled hyperpigmentation.

This condition typically resolves following with antibiotics. Oral minocycline is usually the first line therapy for prurigo pigmentosa.¹ Excellent results have also been achieved with doxycycline¹¹, macrolide antibiotics¹², and dapsone (diaminodiphenyl sulfone).¹ In our patient, oral doxycycline 100 mg twice daily had been being administered for 2 months, the erythematous papules and pruritus mostly resolved but its residual hyperpigmentation was persistent.

Reference:

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