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Case Report

A young gentleman with reticulated hyperpigmentation: a case of prurigo pigmentosa

一名患有網狀色素沈澱的年輕男性：色素性癢疹一例

EKY Yau 丘潔怡 and KC Yau 游高照

A 21-year-old Chinese man presented with four years' history of recurrent itchy erythematous papules and reticulated hyperpigmentation over the upper back and chest wall. The diagnosis of prurigo pigmentosa was made. Treatment with oral doxycycline resulted in reduction of pruritus and no emergence of new lesions. Clinical features, histological features and treatment of prurigo pigmentosa will be discussed.

一名二十一歲華裔男子，因爲在過去四年內，其上背及胸口不斷重覆出現發癢的紅色丘疹和網狀色素沈澱而求診。其診斷確立爲色素性癢疹；施以口服多西環素治療後，患者皮膚瘙癢減少及沒有新的病變發生。本文將討論色素性癢疹的臨床表現、組織學特徵及治療等。

Keywords: Prurigo pigmentosa, reticulated hyperpigmentation

關鍵詞：色素性癢疹，網狀色素沈澱

Introduction

Prurigo pigmentosa is an uncommon disease. The clinical presentation is recurrent itchy erythematous papules and plaques over the trunk followed by reticulated hyperpigmentation. The prognosis is favourable with good treatment response to minocycline, doxycycline or dapsone.

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Case report

A 21-year-old Chinese student was referred to the Social Hygiene Service, Centre for Health Protection, HKSAR for itchy rash over the chest wall and back. The rash resolved spontaneously after one to two weeks, leaving post-inflammatory hyperpigmentation. He had frequent similar attacks in the last four years. The condition did not respond to topical steroids prescribed by general practitioner. There was no systemic symptom or preceding injury. He enjoyed good past health. His family history was unremarkable. Examination revealed reticulated

hyperpigmentation over the upper back, chest wall and nape of neck. There were also ill-defined erythematous papules and erythema with excoriations over the upper back (Figures 1-3). The complete blood picture, renal function tests and liver function tests were normal. The titre of anti-nuclear antibody was 1:40 while anti-double-stranded DNA was negative. Differential diagnoses included lichen planus pigmentosus, confluent and reticulated papillomatosis of Gougerot and Carteaud, prurigo pigmentosa, reticular erythematous mucinosis, erythema ab igne and Riehl's melanosis.

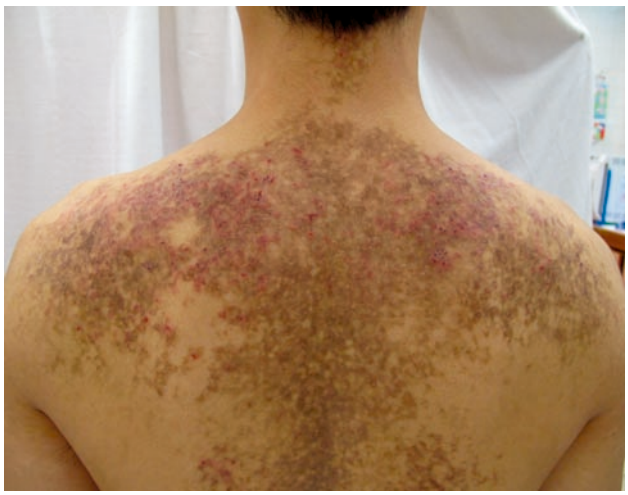

Figure 1. Ill-defined erythematous papules and erythema with reticulated hyperpigmentation over the back and nape of neck.

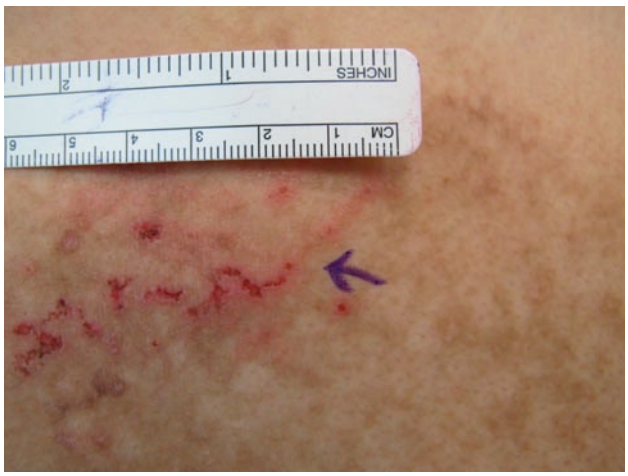

Figure 2. A close-up view of ill-defined erythematous papules with excoriations in a background of netlike hyperpigmentation over the back.

An incisional skin biopsy over the chest wall showed interface dermatitis with mild acanthosis, focal exocytosis, spongiosis, vacuolar alteration of the basal layer and necrotic keratinocytes (Figures 4 & 5). There was a superficial perivascular inflammatory cell infiltrate composed of lymphocytes, a few neutrophils and eosinophils. Pigmentary incontinence was present in the upper dermis. There was an increase in mucin. The direct immunofluorescence study was negative.

The clinical and histological findings were compatible with the diagnosis of prurigo pigmentosa. The patient was treated with oral doxycycline 100 mg daily. Despite the persistent post-inflammatory hyperpigmentation, he had significant clinical improvement with alleviation of pruritus and no new lesions were observed after four weeks of treatment.

Discussion

The condition was first described as Nagashima's disease by Nagashima et al in 1971 and was later named as prurigo pigmentosa in 1978.[1] It is an uncommon disease and the prevalence is as yet unknown. It is more commonly found in Japan, especially among adult Japanese women. Cases

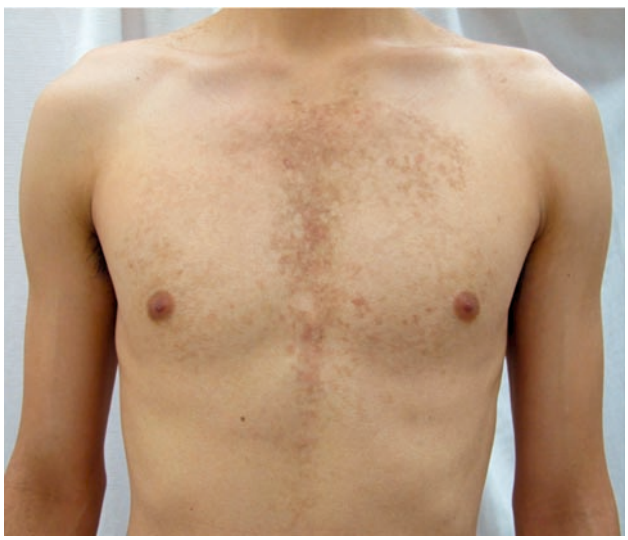

Figure 3. Reticulated hyperpigmentation over the chest wall.

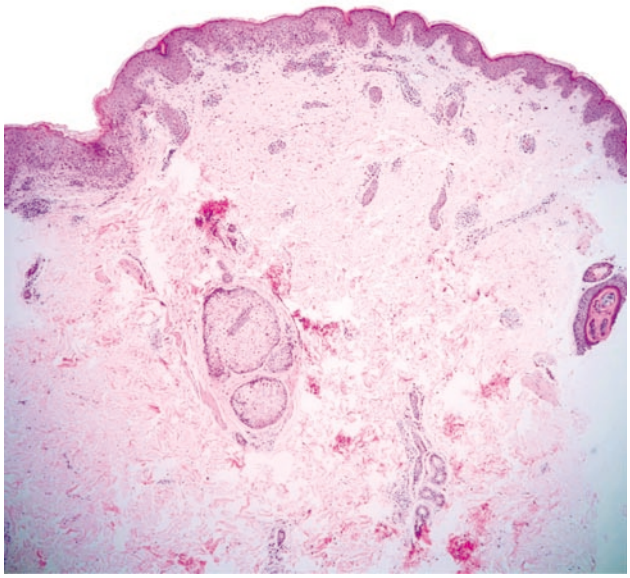

Figure 4. Low power view of the lesion with main pathology at the dermal-epidermal junction and superficial dermis.

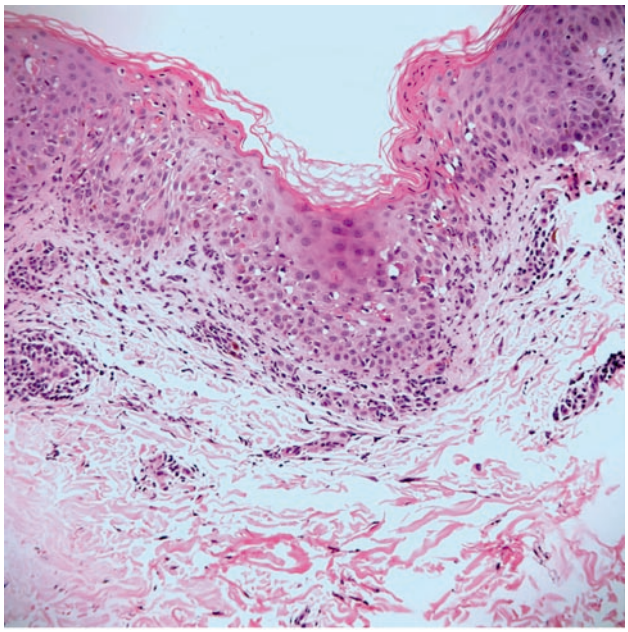

Figure 5. Medium power view showing epidermal acanthosis with associated spongiosis, lymphocytic exocytosis, apoptotic keratinocytes and vacuolar interface dermatitis. The superficial dermis shows a perivascular lymphocytic infiltrate with scattered melanophages.

have been reported in Korea, Singapore, Australia and Europe. Female to male ratio is about 2:1. Most patients develop symptoms between the ages of 10 and 30 years. The mean age of diagnosis is 22 years. Patients typically present with itchy erythematous papules or plaques, leaving behind characteristic net-like hyperpigmentation. The back is the most commonly affected area, following by the chest, shoulders, neck and arms. Sweating, exercise or wearing wet clothing may precipitate new lesions. Apart from the above physical factors, some cases were reported to be associated with ketosis, dieting, sudden weight loss, pregnancy and alcohol intake. The disease duration can range from one month to 11 years, with a mean duration of 2.9 years. About half of the patients experience recurrence of erythematous lesions, which occurs more frequently during spring and summer.[2] Our patient had onset of symptoms at the age of 17 years. He had recurrent typical erythematous papules with excoriations over the back and chest wall. The papules resolved into reticulated hyperpigmentation. No triggering or associating factor was found in his case. He did not have any history of diabetes mellitus, recent dieting or significant weight loss.

The pathogenesis of prurigo pigmentosa has not been elucidated yet. Some authors attributed the disease to exogenous factors like friction from clothing, which could be a mechanical stimulus during exercise or hot weather. Allergic reactions to chemical agents including chromium in acupuncture needle, nickel, chrome in detergents have been considered as causative factors.[3-5] Ketosis has also been described as a potential aetiology. Urine ketones were found in some cases of prurigo pigmentosa and treatment response of skin lesions correlated with the resolution of ketosis.[6,7] Moreover, prurigo pigmentosa was observed to be associated with diabetes mellitus or dieting in which ketosis could exist. The above findings supported the role of ketosis in the pathogenesis of prurigo pigmentosa. Other factors such as atopic diathesis and pregnancy

have also been suggested, yet the causative relationship has not been established.

The diagnosis of prurigo pigmentosa relies mainly on clinical findings from the history and physical examination because histopathological features of prurigo pigmentosa are often non-specific. Microscopic findings vary with different stages of lesions. Early lesions show superficial perivascular infiltrate mainly composed of neutrophils, together with oedematous dermal papillae, spongiosis, ballooning and scanty necrotic basal keratinocytes. Neutrophilic infiltrate may be seen in upper epidermis. In more mature lesions, lymphocytic infiltrate predominates and distributes in patchy lichenoid pattern. Some eosinophils and neutrophils can be seen. There would be more profound liquefactive degeneration of basal layer with pigmentary incontinence. Vesiculation within and below epidermis may be found. Epidermis becomes hyperplastic with parakeratosis in late lesions.[8,9]

There have been no randomised controlled trials in the treatment of prurigo pigmentosa. Minocycline (100-200 mg per day) and dapsone (50-100 mg per day) are the mainstays of treatment. Both have anti-inflammatory properties and can inhibit the migration of neutrophils which are the main components of the inflammatory infiltrate in early lesions. Significant improvement with rapid resolution of itchiness and rash has been observed in patients treated with minocycline or dapsone.[2] Other treatments such as doxycycline,[9] and macrolide antibiotics (clarithromycin and roxithromycin) have been reported to be effective.[10] Isotretinoin, and sulphamethoxazole, have also been tried but efficacy remains unclear.[11,12] Topical corticosteroids and oral antihistamines are ineffective in treating prurigo pigmentosa.

In conclusion, prurigo pigmentosa is characterised by reticulated hyperpigmentation following recurrent erythematous papules and plaques on the trunk. The histopathology of this condition can be non-specific and the diagnosis is based on clinico-pathological correlation. The prognosis is good with a favourable response to treatment.

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