Erythematous papules evolving into reticulated hyperpigmentation on the trunk: A case of prurigo pigmentosa

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INTRODUCTION

Prurigo pigmentosa is a rare cutaneous disorder consisting of a network of erythematous, pruritic papules evolving into reticulated hyperpigmentation with a specific predilection for the trunk. It was first described in Japan in 1971, where the condition still predominates. Although recognition is increasing outside of Japan, the disease is still not well known in the West. Herein, we present the case of a 19-year-old white woman with prurigo pigmentosa diagnosed based on the correlation of clinical and histopathologic findings. Although prurigo pigmentosa is well known in Japan, the low incidence of this disease entity in the United States is likely secondary to underdiagnosis. Thus, the purpose of this case is to increase recognition of this unusual pruritic, reticulated rash.

CASE REPORT

A young white woman in her late teens presented with a 3-year history of pruritic, sometimes painful, erythematous papules and plaques that resolved leaving behind reticulated hyperpigmented patches over her posterior neck, midback, chest, and upper abdomen. She was otherwise healthy with a normal review of systems and denied dieting. Additionally, she denied any provoking or exacerbating factors. Physical examination found a symmetric netlike pattern of erythematous papules coalescing into reticulated plaques over the interscapular back (Figs 1 and 2). Underlying this were hyperpigmented patches of a similar netlike configuration. A reticulated pattern of hyperpigmentation was also observed on the patient’s inter- and inframammary chest, upper abdomen, and posterior neck. The neck lesions were accompanied by excoriations. The nails and oral mucosa showed no abnormalities. Laboratory results were within normal limits including complete blood count, aspartate aminotransferase, alanine transaminase, thyroid-stimulating hormone, and herpes simplex viruses 1 and 2. Antinuclear antibody was weakly positive at 1:80. Previous treatments with calcipotriene, topical tacrolimus, and oral and topical steroids were unsuccessful. Two 4-mm punch biopsies were performed from the chest and left neck for histopathologic examination (Figs 3-5). Hematoxylin-eosin staining showed vacuolar changes and many necrotic keratinocytes along the epidermal-dermal junction, with lymphocytes present along the junctional zone and in the papillary dermis. Scattered eosinophils and neutrophils were also observed. The overlying cornified layer showed mostly parakeratosis. Relying on clinical and histopathologic correlation, a diagnosis of prurigo pigmentosa was made. Doxycycline, 100 mg 2 times daily, was initiated with resolution of the inflammatory component, although the pigmentary alternation remains.

DISCUSSION

Prurigo pigmentosa, first described in the Japanese literature in 1971 by Nagashima et al,1 is a rare cutaneous disorder consisting of a network of erythematous, pruritic papules eventually evolving into reticulated hyperpigmentation with a specific predilection for the trunk. Although the disease continues to be found predominately in Japan, it has become increasingly recognized worldwide. The first case of prurigo pigmentosa in a white
person was reported in 1981. The initial reports mention a possible proclivity within the Japanese population. However, in more recent literature, investigators argue that because prurigo pigmentosa is not as well known outside of Japan, it is often misdiagnosed or underdiagnosed.

Prurigo pigmentosa predominates in young females with a ratio of 4 to 6:1 with an average age of onset in the mid-20s. It has never been seen in prepubescent children or the elderly. No family history of similar cutaneous disorders has been reported in affected patients. The lesions first appear as symmetrically distributed pruritic erythematous, urticarial papules or papulovesicles coalescing into plaques with a predilection for the upper back, posterior neck, clavicular area, and chest. On the upper back, the lesions typically form a wedge shape with the apex pointing inferiorly. Less commonly, the abdomen and shoulders are involved. Often, secondary changes of excoriation, scaling, or crusting are observed. Although the individual lesions resolve quickly over 1 week, residual netlike hyperpigmentation remains. The disease course is punctuated by frequent recurrences followed by remissions lasting weeks to years. Abnormalities of the hair, nails, and mucous membranes have not been observed.

Fig 1. Multiple erythematous papules and papulovesicles coalescing into reticulated plaques accompanied by hyperpigmentation on the back.

Fig 2. A closer view of the erythematous, edematous papules with adjacent hyperpigmentation on the chest.

Fig 3. Histopathologic findings show parakeratosis, vacuolar changes, and lymphocytes present along the junctional zone and in the papillary dermis along with scattered eosinophils and neutrophils (Hematoxylin-eosin stain; original magnification: ×4).

Fig 4. Histopathologic specimen shows the vacuolar changes and necrotic keratinocytes found in the epidermis. This is accompanied by dermal changes including the primarily lymphocytic infiltrate and sparse eosinophils observed here (Hematoxylin-eosin stain; original magnification: ×100).
Histologically, the early disease course shows a superficial perivascular and interstitial infiltrate of neutrophils with collections also in the epidermis and dermal papillae. Spongiosis and necrotic keratinocytes are also noted as the process further evolves. Fully developed lesions show predominantly lymphocytes in a lichenoid pattern along with eosinophils and variable neutrophils. Necrotic keratinocytes and spongiosis are also accompanied by epidermal ballooning, which can lead to vesiculation or vacuolar changes of the dermal-epidermal junction. Finally, in the late stage, the epidermis shows hyperplasia and parakeratosis, and melanophages are seen in the upper dermis with a sparse infiltrate of lymphocytes. Immunofluorescence studies have been negative or nonspecific when performed. Biopsy findings alone are not specific enough to make the diagnosis of prurigo pigmentosa, thus, the importance of the clinical and histopathologic correlation.

No clear etiology for the development of prurigo pigmentosa has been determined. Suspected factors include friction from clothing, allergic contact, diabetes, ketosis, or dieting. Although many associations have been proposed, none have been consistently proven throughout the literature. No underlying systemic symptoms have been associated. When performed, routine laboratory test results have been within normal limits in patients with prurigo pigmentosa.

The differential diagnosis of prurigo pigmentosa includes pigmented contact dermatitis, confluent reticulated papillomatosis, and Dowling-Degos disease. Although a pigmented contact dermatitis may have preceding erythematous papules resolving with a postinflammatory hyperpigmentation similar to prurigo pigmentosa, this dermatitis would be expected to respond to topical or oral steroids. The hyperpigmentation seen with prurigo pigmentosa could appear similarly to confluent reticulated papillomatosis; however, the latter is not preceded by erythematous papules. Dowling-Degos disease classically has hyperpigmentation in flexural sites; however, one would expect to potentially see hyperpigmentation of axillae or groin, which are not involved in prurigo pigmentosa. The characteristic findings found on biopsy of Dowling-Degos disease also help to differentiate it from prurigo pigmentosa.

Therapeutic options used most commonly for prurigo pigmentosa include the tetracycline class of antibiotics and dapsone. Both treatments are thought to be effective by inhibiting neutrophil chemotaxis. Both minocycline and doxycycline at doses of 100 mg 1 to 2 times daily have been used successfully. Dapsone at doses of 25 mg to 100 mg daily has also been used with great efficacy. Topical and oral steroids have shown no to limited improvement. Although the medications are quite effective in stopping the inflammatory component, they do not affect the residual hyperpigmentation. Once treatment is discontinued, recurrence is possible.

Prurigo pigmentosa is a rare dermatosis that is often misdiagnosed in the West because of a lack of awareness of the condition. Correlation of the clinical and histopathologic findings is essential to making the diagnosis.

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