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An itchy rash in a young Caucasian woman

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Case report

We report of a 23-year-old woman who presented with a six-month history of a recurring, pruritic rash located on her chest, upper abdomen and most recently on the lower back. Until then the patient had been healthy. She did not take any medications, was not aware of any allergies or a family history of skin diseases.

The rash had occurred initially six months ago and was marked by an acute onset and a chronic recurrent course. The patient noted a timely association between the onset of the skin lesions and the start of a vegetarian diet. Furthermore physical activity and sweating worsened the symptoms.

Clinically the woman presented with erythematous patches and urticarial papules as well as crusted vesicles and pustules with an erythematous base that were located on the chest and the inframammary folds and distributed in a symmetrical fashion (Figure 1A, B). Under the provisional diagnosis of an autoimmune bullous disease, the woman had recently received systemic and topical corticosteroids. Lesions progressed under this therapy. Additionally, new pink, slightly urticarial papules and plaques developed on the back. Intense pruritus was the main complaint of the patient. This symptom did not respond to antihistamine or steroid therapy.

Routine laboratory tests including blood count, blood chemistry, and serum protein levels were normal. No elevated antinuclear antibody titers were found. Direct and indirect immunofluorescence assays and an ELISA to rule out an autoimmune bullous disease were performed and were negative.

A skin biopsy taken from one of the most recent lesions on the back showed a sparse perivascular lymphocytic cell infiltrate with occasional neutrophils and a discrete edema of the papillary dermis (Figure 2). No epidermal changes were present. In contrast to these subtle changes, a skin biopsy taken from a fully developed inframammary lesion displayed dramatic epidermal changes consisting of ballooning of keratinocytes, necrosis en masse of keratinocytes with intraepidermal vesiculation and exocytosis of neutrophils and eosinophils. The epidermal changes were accompanied by a dense superficial interstitial and perivascular dermal inflammatory cell infiltrate consisting of lymphocytes mainly, but also neutrophils and eosinophils (Figures 3, 4).

The clinical and histopathologic findings summoned up to the diagnosis of prurigo pigmentosa. Alternative diagno-

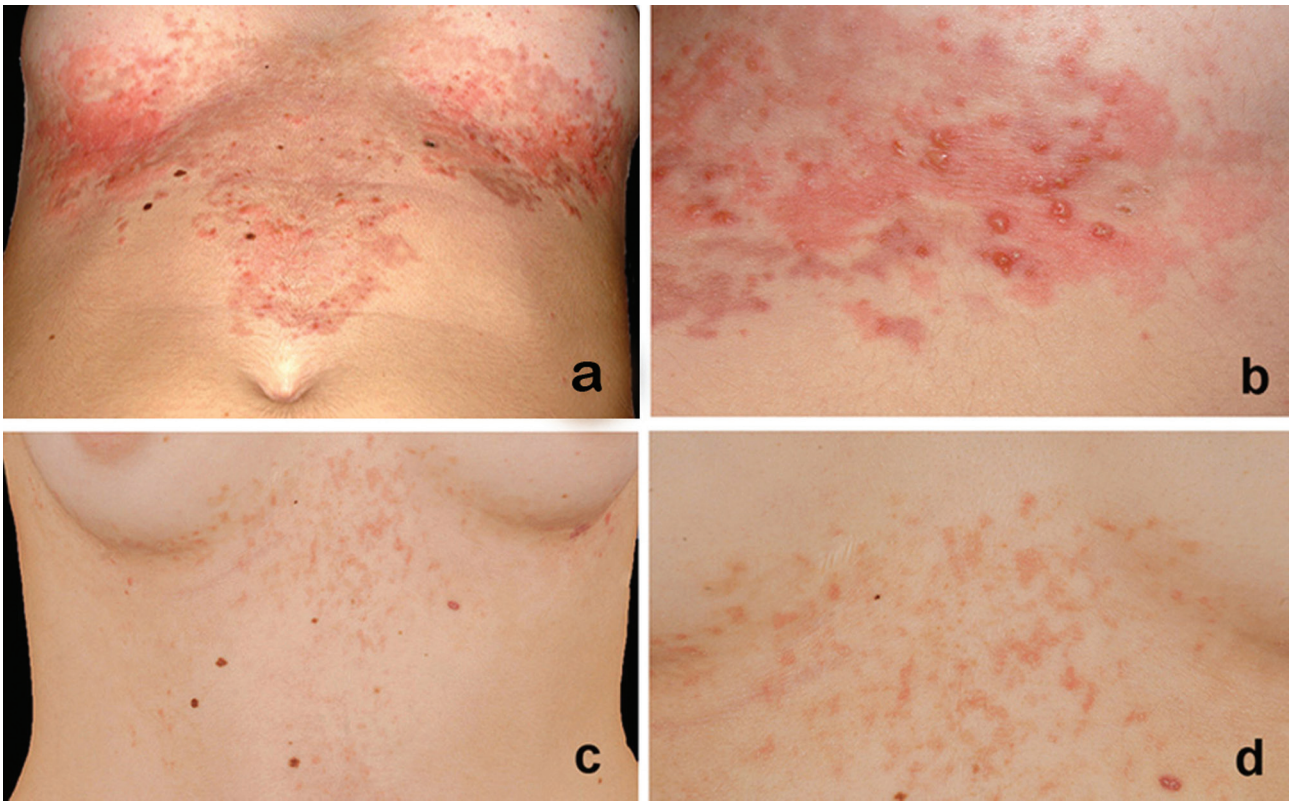

Figure 1. Clinical presentation. (A), (B) Fully developed lesions presenting as erythematous papules, plaques, pustules and vesicles. (C), (D) After a two-week course of doxycycline, skin-colored papules and hyperpigmented macules remain. [Copyright: ©2012 Jonak et al.]

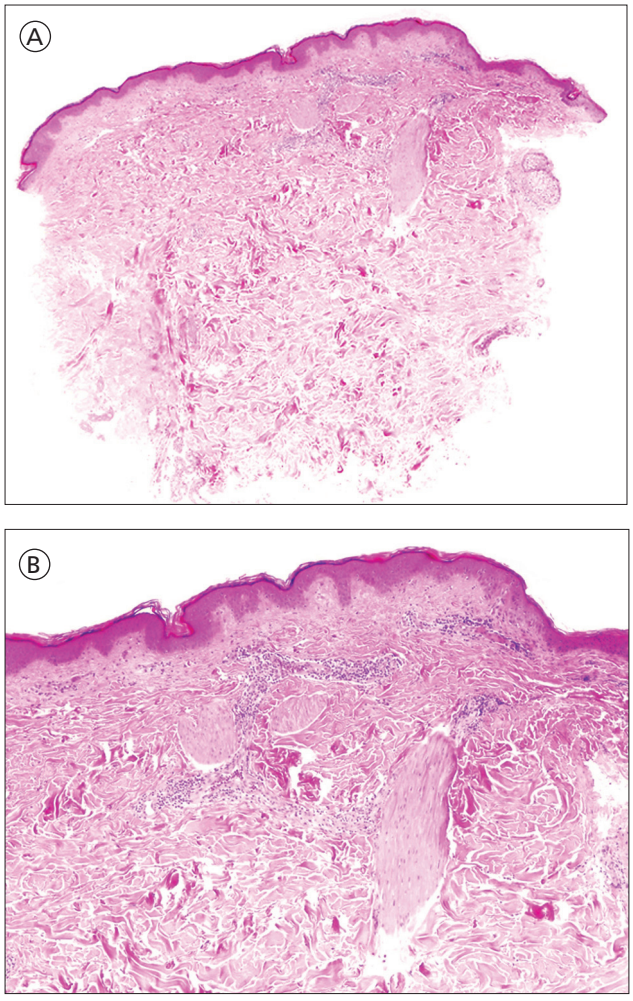

Figure 2. Photomicrographs of an early lesion taken from the lower back displays under an intact epidermis a sparse perivascular lymphocytic cell infiltrate with occasional neutrophils. Hematoxylin and eosin [HE] stain; magnification, (A) x4, (B) x20. [Copyright: ©2012 Jonak et al.]

ses were dismissed on the basis of the clinical, histopathologic and laboratory findings.

Treatment with doxycycline 200 mg daily was initiated and the lesions improved rapidly over the course of one week (Figure 1C, D), therefore, the daily doxycycline dose was reduced to 100 mg. The patient was advised to continue this therapeutic regimen for up to three weeks after alleviation of all symptoms. Our patient has not experienced a relapse within the last six months.

Conclusions

Prurigo pigmentosa is a rare inflammatory skin disorder that was first described by Nagashima et al in 1971 [1]. Most reports are of Japanese patients, while the disease is rarely diagnosed outside Japan. It has been subject to discussion if this reflects a higher incidence in the Japanese population or if the disease is underdiagnosed in other countries [2].

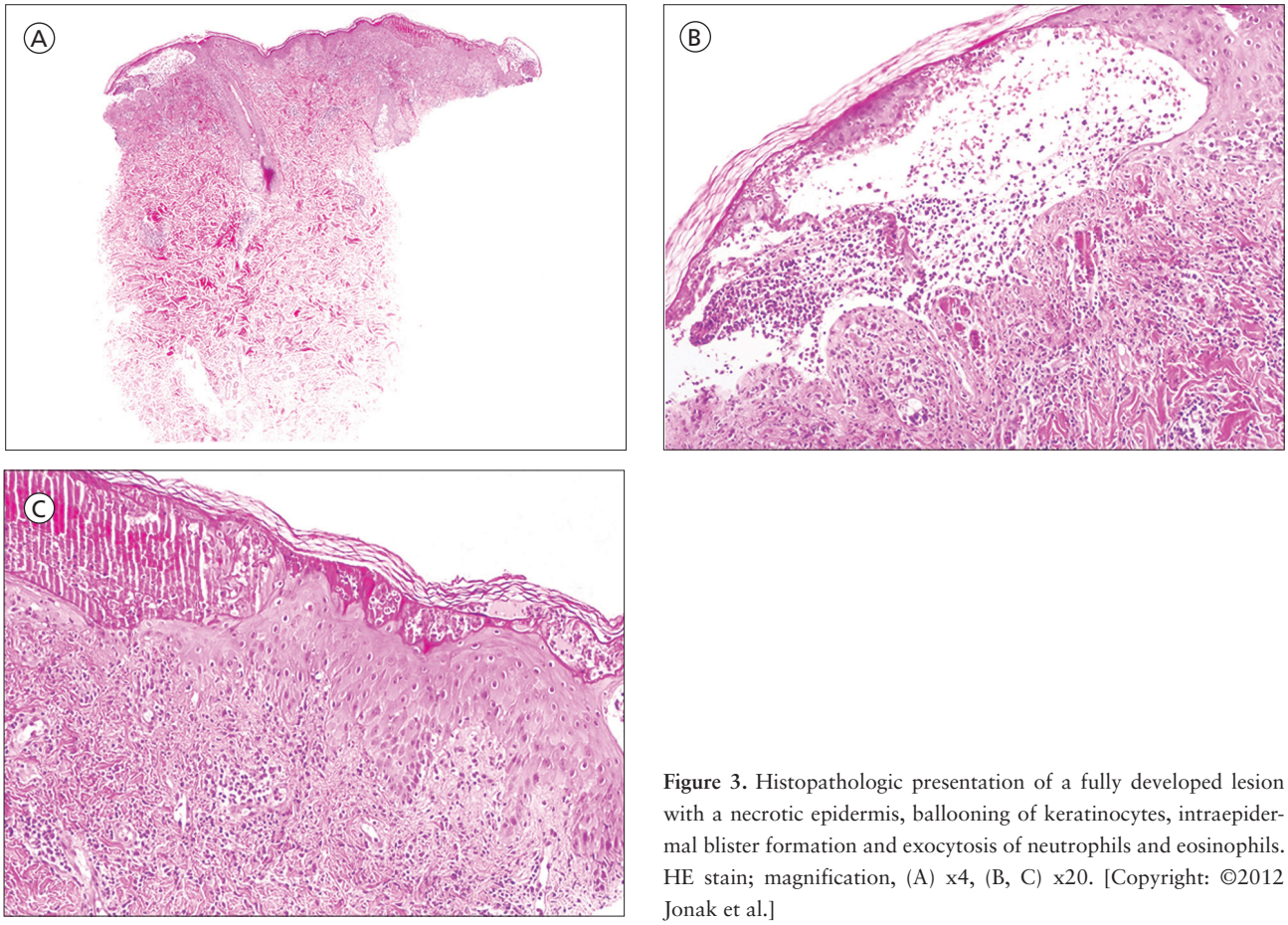

Figure 3. Histopathologic presentation of a fully developed lesion with a necrotic epidermis, ballooning of keratinocytes, intraepidermal blister formation and exocytosis of neutrophils and eosinophils. HE stain; magnification, (A) x4, (B, C) x20. [Copyright: ©2012 Jonak et al.]

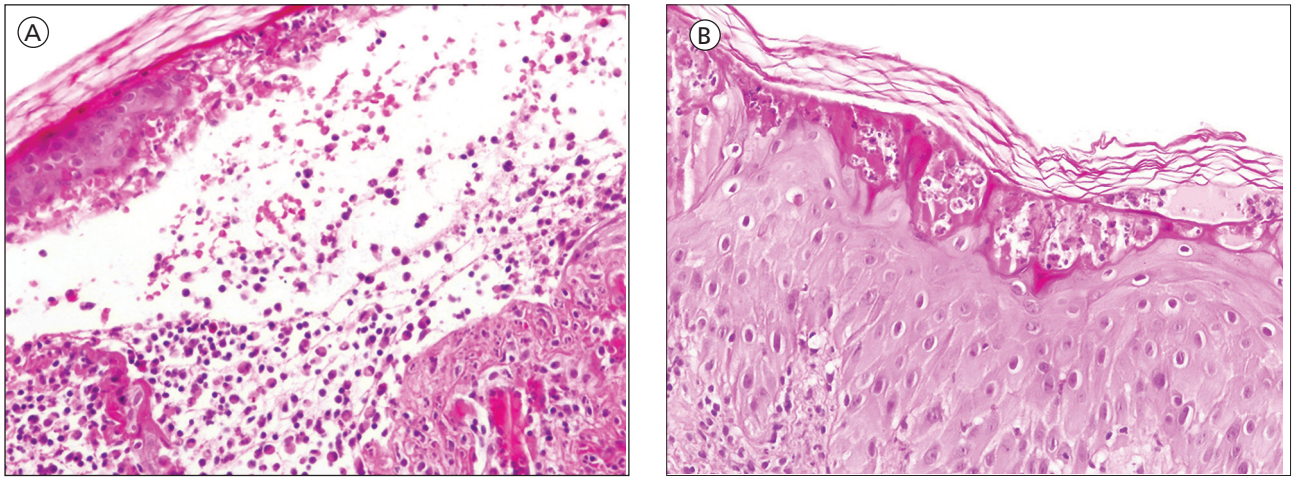

Figure 4. At higher magnification, numerous neutrophils and eosinophils can be identified within an intraepidermal blister (A), and subcorneal pustules are present beneath a basket-woven stratum corneum (B). HE stain; magnification, x40. [Copyright: ©2012 Jonak et al.]

The clinical presentation of prurigo pigmentosa varies according to the stage of the disease: it ranges from urticarial papules and plaques to blisters, pustules and finally to hyperpigmented reticulated macules. Accordingly, the differential diagnosis includes such different diseases like acute lupus erythematosus, dermatitis herpetiformis, linear IgA disease or confluent and reticulated papillomatosis of Gougerot and Carteaud [3].

Clinicopathologic correlation is the key to diagnosis. Böer and Ackerman, in 2003, defined criteria for histopathologic diagnosis of prurigo pigmentosa [4,5]. Still, the etiology of the disease remains unknown. In the majority of cases the condition affects otherwise healthy adolescents and young adults. Females are three to four times more frequently affected than males [6]. Metabolic disorders associated with ketosis, such as diabetes mellitus, dieting and fasting have been implied as causative factors [7]. Recently, two retrospective clinical studies reported ketosis in eight out of 10 and in six out of 16 patients, respectively [6,8]. This is in line with the observation of a timely association between dieting

and the disease onset in our patient. However, future studies will need to clarify if ketosis plays a causative role or if it merely represents an event that is unrelated to the pathogenesis of prurigo pigmentosa.

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