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A unique case of prurigo pigmentosa related to ketogenic diet

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Abstract
Prurigo pigmentosa is a rare inflammatory dermatosis that primarily affects Japanese patients. Various triggers have been identified, including the ketogenic diet. Given the current popularity of the ketogenic diet, the incidence of prurigo pigmentosa may be on the rise. Herein, to the best of our knowledge, we present the first case of prurigo pigmentosa reported in a Hispanic patient in the United States.

Keywords: prurigo pigmentosa, ketogenic diet, inflammatory dermatoses, diet and dermatology

Introduction
Prurigo pigmentosa is a rare inflammatory skin disorder, which is characterized both clinically and histologically by two distinct phases; an initial acute inflammatory phase followed by a more chronic, hyperpigmented phase. Although the cause of this eruption is unknown, various triggers have been suggested including physical triggers, hormonal triggers, and metabolic triggers. We present a case of ketosis-induced prurigo pigmentosa in a Hispanic woman.

Case Synopsis
A healthy 22-year-old Hispanic woman presented to the dermatology clinic with a pruritic rash two weeks after starting a ketogenic diet for weight loss. She did not take any medications and denied systemic symptoms, travel, or new topical products. Physical examination revealed pink papulovesicles and vesicles coalescing into reticulated plaques on the neck, chest, arms, and back (Figure 1). A punch biopsy from the chest showed spongiosis, intraepidermal vesicles, and microabscesses containing neutrophils and eosinophils (Figure 2). A diagnosis of prurigo pigmentosa was made. She was started on oral minocycline and counseled to resume a non-ketogenic diet. After two weeks the eruption improved, leaving residual hyperpigmented reticulated patches (Figure 3).

Case Discussion
Prurigo pigmentosa is a rare inflammatory dermatosis originally described by Nagashima in the Japanese population [1]. Patients present with pruritic papules and papulovesicles on the neck, chest, and back, which then progress to crusted papules and finally to hyperpigmented macules in a reticular pattern. The vast majority of cases are seen in the Japanese population, with women twice as likely to be affected as men [2, 3]. Although the pathogenesis is incompletely understood, it is postulated that neutrophil-mediated inflammation in the skin is induced by exogenous stressors such as...

Figure 1. Acute eruption of prurigo pigmentosa. Initial presentation with pruritic pink papulovesicles and vesicles coalescing into reticulated plaques on the A) neck, chest, arms, and B) back, two weeks after starting a ketogenic diet.
ketosis [4]. Elevated levels of pro-inflammatory cytokines including interleukin-6 and tumor necrosis factor have been demonstrated in ketotic cows and show a positive correlation with elevated blood concentrations of non-esterified fatty acids [5]. It is conceivable that a similar mechanism may account for the inflammation seen in human skin. The differential diagnosis of prurigo pigmentosa is broad and varies depending on the timing of presentation, but may include urticaria, acute contact dermatitis, pityriasis lichenoides et varioliformis acuta, and confluent and reticulated papillomatosis. Histologic features also vary depending on the time of biopsy, with earlier lesions showing spongiosis, papillary dermal edema, and neutrophils and later lesions showing hyperpigmentation of the epidermis, dermal melanophages, and some degree of acanthosis [6]. In cases of ketosis-induced prurigo pigmentosa, treatment consists of cessation of the causative diet as well as administration of antibiotics that affect neutrophil chemotaxis, such as minocycline and doxycycline [3]. The rash is not responsive to topical corticosteroids.

**Conclusion**

Based upon our literature review, we did not find any previously published cases of prurigo pigmentosa in Hispanic patients in the United States. Given the current trend toward high fat, low carbohydrate diets including the ketogenic diet, the incidence of prurigo pigmentosa in the United States may be on the rise. Though less than twenty cases have been described in the United States, public awareness of this entity is certainly growing; several websites and online forums for paleo and ketogenic diet enthusiasts contain references to “the keto rash.” It is therefore more important than ever for dermatologists to be aware of this entity in order to ensure prompt diagnosis and appropriate management.

**Potential conflicts of interest**

The authors declare no conflicts of interests.
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