Bullous prurigo pigmentosa following a ketogenic diet

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To the Editor: Prurigo pigmentosa is a rare inflammatory dermatosis first reported in Japan in 1971. Typical lesions of prurigo pigmentosa are erythematous papules with a reticular arrangement that quickly resolve leaving pigmentation. As a rare manifestation, bullous prurigo pigmentosa has been reported in cases[1,2] and case series.[3] We herein describe a case of prurigo pigmentosa with vesiculobullous formation following a ketogenic diet.

A 21-year-old woman presented to our department with itchy eruptions that had been evolving for 2 weeks, which showed no improvement after the use of topical corticosteroid. Her past medical history and family history were unremarkable. On physical examination, multiple scaly erythematous papules and brownish macules coalescing into plaques and patches arranged in a reticulate pattern on the chest, back, sacrum, and mons pubis. Furthermore, tense vesicles were scattered on the chest, back, and sacrum [Figure 1A–1E]. Direct microscopy examination of scrapings from the scaly papules on the chest showed negative for fungi. Serum biomarkers of systemic lupus erythematosus were all within the normal ranges. Two skin biopsies were taken, including a vesicle for regular hematoxylin and eosin staining and a perilesional normal-appearing site for direct immunofluorescence.

In the absence of a clear diagnosis during the first visit, the patient was not given any treatment. When she came back 1 week later for biopsy results, the eruptions had resolved spontaneously, leaving reticular pigmentation. She reported that she had been following a ketogenic diet for 2 weeks to lose weight, before the emergence of eruptions. When first presented in the department, she had already returned to a normal diet with carbohydrates. The biopsy demonstrated focal epidermal necrosis, intra-epidermal bulla with a few lymphocytes, and neutrophils. Lymphocytic infiltrate in the papillary dermis was also observed [Figure 1F]. Direct immunofluorescence was negative for IgM, IgG, IgA, and C3. Prurigo pigmentosa was diagnosed. Urinary examination revealed normal ketone levels on the second visit. After a 20-month follow-up, there was no relapse.

Prurigo pigmentosa is characterized by typical pruritic lesions arranged in a reticulate configuration which is resolved with hyperpigmentation, with a predominant incidence in young females. The histopathological finding of prurigo pigmentosa is stage-specific, with an initial superficial perivascular neutrophil infiltrate followed by a scattering of neutrophils in the dermal papillae. The main differential diagnosis includes confluent reticulated papillomatosis, acute lupus erythematosus, and dermatitis herpetiformis.

Although the etiology of prurigo pigmentosa remains to be elucidated, some conditions including ketosis, diet, diabetes mellitus, and pregnancy have been associated with the disease or even speculated to have a causal link with it. Prurigo pigmentosa after a ketogenic diet, which consists of reducing the carbohydrate content in the diet (usually to <50 g/d) while increasing the fraction of fat and protein intake, has been reported in young women.[4]

Bullous prurigo pigmentosa is rarely reported. Clinically, it may be induced by intense rubbing. Histopathologically, extensive inflammation with intercellular and intracelluar edema, basal liquefaction, and papillary dermal edema may result in vesiculobullous lesions.[1] This is a rare case of bullous prurigo pigmentosa following a ketogenic diet. Furthermore, the most commonly affected sites of prurigo pigmentosa are the back, chest, and neck. In our case, the mons pubis is also involved, which is quite rare according to a literature review performed in 2017.[5]

In most of the cases, the reintroduction of carbohydrates into the diet is sufficient to cure prurigo pigmentosa; if insufficient, oral cycling antibiotics are usually the first-line therapy.
Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for his images and other clinical information to be reported in the article. The patient understands that her name and initials will not be published and due efforts will be made to conceal the identity of the patient, although anonymity cannot be guaranteed.

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Conflicts of interest

None.

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