

Prurigo Pigmentosa: An Itchy, Urticarial Eruption Confused for Food Allergy



Peter Capucilli, MD^{a,b}, Joy Wan, MD, MSCE^{b,c}, James Corry, MD^{a,b}, Jennifer H. Chuang, MD, MS^{b,d}, Rebecca L. Linn, MD^{b,e}, and Marissa J. Perman, MD^{b,c} Philadelphia, Pa

A 17-year-old female of Chinese descent presented to our allergy clinic with concern for food allergy reaction. Chief complaint was “hives” starting 30 minutes after ingesting mixed nuts. She described acute onset of pruritus and development of pink-brown eruptions symmetrically across the upper chest, back, and shoulders, progressive over several hours. No other immediate allergic symptoms were noted. She underwent evaluation at urgent care and was treated with antihistamines and systemic and topical steroids for presumed urticaria. Despite these interventions, her rash persisted over 2 weeks without improvement, prompting referral to our clinic.

On assessment, her history was deemed inconsistent with food allergy. She had tolerated all tree nuts previously without reaction. Furthermore, persistence of a fixed eruption over weeks despite antihistamine and steroid treatment was inconsistent with an IgE-mediated process. Thus, skin prick and serum-specific IgE testing were not performed.

Physical examination was significant for a very thin female with reticulated, pink-brown papules coalescing into plaques distributed symmetrically over the shoulders, chest, and central back (Figure 1, A). Review of the patient’s growth chart revealed a profound drop in weight and body mass index from the 25th percentile to below the 3rd percentile over several months. With acknowledgement of this, she admitted to limiting food groups such as dairy and wheat for acne prevention. Vital signs were likewise concerning for bradycardia of 48 bpm, which prompted referral for emergency evaluation and hospitalization for severe malnutrition.



A



B

FIGURE 1. A, PP initial evaluation. Erythematous papules and plaques in a reticulated pattern on the back. B, Mature PP. Reticulated hyperpigmented patches on back.

^aDivision of Allergy and Immunology, The Children’s Hospital of Philadelphia, Philadelphia, Pa

^bPerelman School of Medicine at the University of Pennsylvania, Philadelphia, Pa

^cSection of Dermatology, Division of General Pediatrics, The Children’s Hospital of Philadelphia, Philadelphia, Pa

^dCraig-Dalsimer Division of Adolescent Medicine, The Children’s Hospital of Philadelphia, Philadelphia, Pa

^eDepartment of Pathology and Laboratory Medicine, The Children’s Hospital of Philadelphia, Philadelphia, Pa

Patient and parental consent was obtained for reproduction of clinical history and patient photos for academic publications. The original case report was reviewed and approved by The Children Hospital of Philadelphia Institutional Review Board per protocol number 18-014833.

Conflicts of interest: The authors declare that they have no relevant conflicts of interest.

Received for publication February 18, 2018; revised manuscript received and accepted for publication February 27, 2018.

Available online March 23, 2018.

Corresponding author: Peter Capucilli, MD, Division of Allergy and Immunology, The Children’s Hospital of Philadelphia, 3401 Civic Center Blvd, Philadelphia, PA 19104. E-mail: capucilli@email.chop.edu.

J Allergy Clin Immunol Pract 2018;6:1381-2.
2213-2198

© 2018 American Academy of Allergy, Asthma & Immunology
<https://doi.org/10.1016/j.jaip.2018.02.033>

Laboratory testing included an elevated beta hydroxybutyrate level of 0.60 mmol/L (reference range, 0.10-0.30 mmol/L), consistent with trace ketosis, but was otherwise unremarkable. A punch biopsy was performed by Dermatology, with pathology showing spongiosis with superficial perivascular infiltrate of lymphocytes and eosinophils with a hyperkeratotic epidermis, overall consistent with the diagnosis of prurigo pigmentosa (PP).

During her 9-day hospitalization, the patient was ultimately diagnosed with anorexia nervosa, and with nutritional restoration, had steady improvement of rash with fading of the erythema to light brown reticular patches (Figure 1, B). Follow-up at 1 month showed improved hyperpigmentation without recurrence of erythema.

First described in Japan in 1971,¹ PP is a relatively unknown dermatosis, characterized by a rapidly developing and intensely pruritic truncal rash, resembling the symptoms of acute urticaria. It is most frequently diagnosed in women of Asian descent, and although the exact etiology is unknown, an association with ketosis is suspected, with cases described in patients with diabetic ketoacidosis,² anorexia nervosa,³ and adherence to ketogenic diets.⁴

Classically, PP presents in stages. Early lesions include reticulated and urticarial papules, with evolution over days to a late stage characterized by a faded brown net-like hyperpigmentation, specifically involving the trunk and without mucosal involvement.^{5,6} Most importantly, PP is unresponsive to antihistamines and topical steroids, with oral antibiotics being the mainstay of treatment.^{5,6}

In this case, evaluation of an atypical urticarial rash led to the recognition and diagnosis of severe malnutrition in a patient with anorexia nervosa. We highlight that close review of the growth chart was integral to the diagnosis, and should be intrinsic to the allergist's examination of children and adolescents. We encourage clinicians to consider PP in the differential diagnosis of urticarial eruptions and to consider possible states of ketosis as an underlying etiology.

Acknowledgements

We acknowledge Dr John Seykora for assistance with clinicopathologic correlation.

REFERENCES

1. Nagashima M, Ohshiro A, Shimizu N. A peculiar pruriginous dermatosis with gross reticular pigmentation. *Jpn J Dermatol* 1971;81:38-9.
2. Ohnishi T, Kisa H, Ogata E, Watanabe S. Prurigo pigmentosa associated with diabetic ketoacidosis. *Acta Derm Venereol* 2000;80:447-8.
3. Nakada T, Sueki H, Iijima M. Prurigo pigmentosa (Nagashima) associated with anorexia nervosa. *Clin Exp Dermatol* 1998;23:25-7.
4. Michaels JD, Hoss E, DiCaudo DJ, Price H. Prurigo pigmentosa after a strict ketogenic diet. *Pediatr Dermatol* 2015;32:248-51.
5. Boer A, Misago N, Wolter M, Kiryu H, Wang XD, Ackerman AB. Prurigo pigmentosa: a distinctive inflammatory disease of the skin. *Am J Dermatopathol* 2003;25:117-29.
6. Satter E, Rozelle C, Sperling L. Prurigo pigmentosa: an under-recognized inflammatory dermatosis characterized by an evolution of distinctive clinicopathological features. *J Cutan Pathol* 2016;43:809-14.

Reproduced with permission of copyright owner. Further reproduction prohibited without permission.