Pruritic Rash on the Neck and Back

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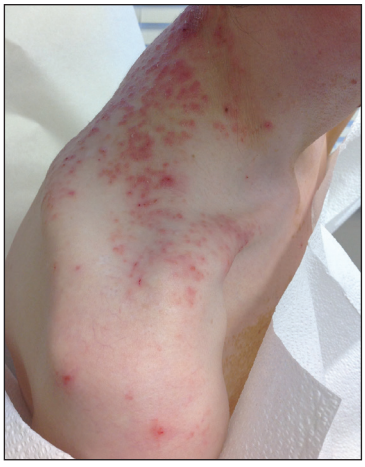

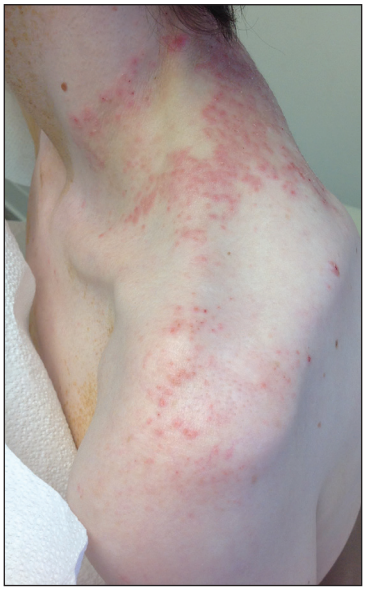

A 43-year-old woman presented with a pruritic rash across the neck and back of 6 months' duration that progressively worsened. She had a medical history of anorexia nervosa, herpes zoster with a recent flare, and peripheral neuropathy. Physical examination showed numerous red scaly papules across the upper back and shoulders that coalesced in a reticular pattern. No similar papules were seen elsewhere on the body.

WHAT'S YOUR **DIAGNOSIS?**

a. allergic contact dermatitis
b. bullous pemphigoid
c. *Pityrosporum* folliculitis
d. prurigo pigmentosa
e. urticaria

PLEASE TURN TO PAGE E39 FOR THE DIAGNOSIS

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THE **DIAGNOSIS:** Prurigo Pigmentosa

A comprehensive metabolic panel collected from our patient 1 month earlier did not reveal any abnormalities. Serum methylmalonic acid and homocysteine were both elevated at 417 nmol/L (reference range [for those aged 2–59 years], 55–335 nmol/L) and 23 µmol/L (reference range, 5–15 µmol/L), respectively. Serum folate and 25-hydroxyvitamin D were low at 3.1 ng/mL (reference range, >4.8 ng/mL) and 5 ng/mL (reference range, 30–80 ng/mL), respectively. Vitamin B_{12} was within reference range. Two 4-mm punch biopsies collected from the upper back showed spongiotic dermatitis.

Our patient's histopathology results along with the rash distribution and medical history of anorexia increased suspicion for prurigo pigmentosa. A trial of oral doxycycline 100 mg twice daily for 2 weeks was prescribed. At 2-week follow-up, the patient's mother revealed a history of ketosis in her daughter, solidifying the diagnosis. The patient was counseled on maintaining a healthy diet to prevent future breakouts. The patient's rash resolved with diet modification and doxycycline; however, it recurred upon relapse of anorexia 4 months later.

Prurigo pigmentosa, originally identified in Japan by Nagashima et al,[1] is an uncommon recurrent inflammatory disorder predominantly observed in young adults of Asian descent. Subsequently, it was reported to occur among individuals from different ethnic backgrounds, indicating potential underdiagnosis or misdiagnosis in Western countries.[2] Although a direct pathogenic cause for prurigo pigmentosa has not been identified, a strong association has been linked to diet, specifically when ketosis is induced, such as in ketogenic diets and anorexia nervosa.[3-5] Other possible causes include sunlight exposure, clothing friction, and sweating.[1,5] The disease course is characterized by intermittent flares and spontaneous resolution, with recurrence in most cases. During the active phase, intensely pruritic, papulovesicular or urticarial papules are predominant and most often are localized to the upper body and torso, including the back, shoulders, neck, and chest.[5] These flares can persist for several days but eventually subside, leaving behind a characteristic reticular pigmentation that can persist for months.[5] First-line treatment often involves the use of tetracycline antibiotics, such as minocycline or doxycycline.[2,4,5] Dapsone often is used with successful resolution.[6] Dietary modifications also have been found to be effective in treating prurigo pigmentosa, particularly in patients presenting with dietary insufficiency.[6,7] Increased carbohydrate intake has been shown to promote resolution.[6] Topical corticosteroids demonstrate limited efficacy in controlling flares.[6,8]

Histopathology has been variably described, with initial findings reported as nonspecific.[1] However, it was later described as a distinct inflammatory disease of the skin with histologically distinct stages.[2,9] Early stages reveal scattered dermal, dermal papillary, and perivascular neutrophilic infiltration.[9] The lesions then progress and become fully developed, at which point neutrophilic infiltration becomes more prominent, accompanied by the presence of intraepidermal neutrophils and spongiosis. As the lesions resolve, the infiltration transitions to lymphocytic, and lichenoid changes can sometimes be appreciated along with epidermal hyperplasia, hyperpigmentation, and dermal melanophages.[9] Although these findings aid in the diagnosis of prurigo pigmentosa, a clinicopathologic correlation is necessary to establish a definitive diagnosis.

Because prurigo pigmentosa is rare, it often is misdiagnosed as another condition with a similar presentation and nonspecific biopsy findings.[6] Allergic contact dermatitis is a common type IV delayed hypersensitivity reaction that manifests similar to prurigo pigmentosa with pruritus and a well-demarcated distribution[10] that is related to the pattern of allergen exposure; in the case of allergic contact dermatitis related to textiles, a well-demarcated rash will appear in the distribution area of the associated clothing (eg, shirt, pants, shorts).[11] Development of allergy involves exposure and sensitization to an allergen, followed by subsequent re-exposure that results in cutaneous T-cell activation and inflammation.[10] Histopathology shows nonspecific spongiotic inflammation, and the gold standard for diagnosis is patch testing to identify the causative substance(s). Definitive treatment includes avoidance of identified allergies; however, if patients are unable to avoid the allergen or the cause is unknown, then corticosteroids, antihistamines, and/or calcineurin inhibitors are beneficial in controlling symptoms and flares.[10]

Pityrosporum folliculitis (also known as *Malassezia* folliculitis) is a fungal acneform condition that arises from overgrowth of normal skin flora *Malassezia* yeast,[12] which may be due to occlusion of follicles or disruption of the normal flora composition. Clinically, the manifestation may resemble prurigo pigmentosa in distribution and presence of intense pruritus. However, pustular lesions and involvement of the face can aid in differentiating *Pityrosporum* from prurigo pigmentosa, which can be confirmed via periodic acid–Schiff staining with numerous round yeasts within affected follicles. Oral antifungal therapy typically yields rapid improvement and resolution of symptoms.[12]

Urticaria and prurigo pigmentosa share similar clinical characteristics, with symptoms of intense pruritus and urticarial lesions on the trunk.[2,13] Urticaria is an IgE-mediated type I hypersensitivity reaction characterized

by wheals (ie, edematous red or pink lesions of variable size and shape that typically resolve spontaneously within 24–48 hours).[13] Notably, urticaria will improve and in some cases completely resolve with antihistamines or anti-IgE antibody treatment, which may aid in distinguishing it from prurigo pigmentosa, as the latter typically exhibits limited response to such treatment.[2] Histopathology also can assist in the diagnosis by ruling out other causes of similar rash; however, biopsies are not routinely done unless other inflammatory conditions are of high suspicion.[13]

Bullous pemphigoid is an autoimmune, subepidermal, blistering dermatosis that is most common among the elderly.[14] It is characterized by the presence of IgG antibodies that target BP180 and BP230, which initiate inflammatory cascades that lead to tissue damage and blister formation. It typically manifests as pruritic blistering eruptions, primarily on the limbs and trunk, but may involve the head, neck, or palmoplantar regions.[14] Although blistering eruptions are the prodrome of the disease, some cases may present with nonspecific urticarial or eczematous lesions[14,15] that may resemble prurigo pigmentosa. The diagnosis is confirmed through direct immunofluorescence microscopy of biopsied lesions, which reveals IgG and/or C3 deposits along the dermoepidermal junction.[14] Management of bullous pemphigoid involves timely initiation of dapsone or systemic corticosteroids, which have demonstrated high efficacy in controlling the disease and its associated symptoms.[15]

Our patient achieved a favorable response to diet modification and doxycycline therapy consistent with the diagnosis of prurigo pigmentosa. Unfortunately, the condition recurred following a relapse of anorexia. Management of prurigo pigmentosa necessitates not only accurate diagnosis but also addressing any underlying factors that may contribute to disease exacerbation. We anticipate the eating disorder will pose a major challenge in achieving long-term control of prurigo pigmentosa.

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