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Severe pellagra masked by concurrent plaque psoriasis: a case report of a hidden diagnosis

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Abstract

Despite characteristic features, psoriasis can mimic other dermatologic conditions, such as seborrheic dermatitis, lichen simplex chronicus, and certain nutritional deficiencies such as pellagra. We present a patient with a longstanding history of severe plaque psoriasis who presented with disfiguring scaly plaques involving greater than 80% body surface area. The patient's disease was minimally responsive to multiple therapies. Repeat punch biopsies demonstrated parakeratosis, psoriasiform hyperplasia, and dilated blood vessels consistent with psoriasis. Given atypical clinical features and overall poor treatment response additional work up was obtained. A serum nutritional panel was consistent with niacin deficiency and the patient later revealed extensive alcohol intake. A diagnosis of concurrent pellagra was made and the patient was started on niacin supplementation and instructed to reduce alcohol intake, while continuing adalimumab and high potency topical steroids. Within two weeks, his disease had markedly improved. Pellagra presents characteristically with a photosensitivity dermatitis that may appear clinically and histologically similar to psoriasis. It is important to maintain an index suspicion for a secondary pathology in treatment-resistant psoriasis.

Keywords: pellagra, psoriasis, psoriasiform, photosensitivity, nutritional deficiency

Introduction

Psoriasis is a chronic inflammatory skin condition that affects 4.7% of the US population [1]. It is classically characterized by well-demarcated, erythematous plaques with silvery scales. Key histological features include parakeratosis, spongiosis, and flattened rete ridges [2]. Despite these characteristic features, psoriasis can mimic other dermatologic conditions, such as seborrheic dermatitis, lichen simplex chronicus, and certain nutritional deficiencies such as pellagra [3].

Given typically classic features, psoriasis is usually a clinical diagnosis. However, atypical and/or treatment-resistant presentations merit further investigation for alternative or secondary conditions that may require a different therapeutic approach. In the case of pellagra, which also appears as scaly plaques, treatment would be targeted towards dietary supplementation instead. Although uncommon in developed countries, it can be seen in the setting of malnourishment and/or malabsorption [4, 5]. Clinical pellagra should be considered as a diagnosis in patients with chronic alcoholism, malnutrition, and amino acid imbalance.

Pellagra is classically described as a trio of symptoms: dementia, photosensitive dermatitis, and diarrhea [7]. Specifically, cutaneous findings appear as well-demarcated, hyperpigmented, symmetric, and scaly plaques in sun-exposed areas, such as the face, neck, and distal extremities [1,6]. Since its histologic features also include psoriasiform hyperplasia, pellagra can be difficult to differentiate from other histologically similar pathologies, making its diagnosis challenging.

Herein we report a case of a patient with clinical and histological characteristics of psoriasis, who did not improve with aggressive multi-regimen treatment until a second diagnosis of concurrent pellagra was diagnosed and treated.

Case Synopsis

Patient is a 33-year-old man, with a longstanding history of severe plaque psoriasis, who presented with disfiguring scaly plaques involving greater than 80% body surface area. Thick plate-like scaling and fissures were significantly worse in photodistributed areas such as the face and hands (**Figure 1**). The patient's disease was minimally responsive to multiple therapies, including etanercept, cyclosporine, and currently, adalimumab. He previously had required multiple hospitalizations for frequent superinfections and severe pruritus. In addition, the patient was adamant that recent flares were directly related to UV exposure. He also was suffering from anxiety and depression because of the severity of his disease and appearance.

Repeat punch biopsies demonstrated parakeratosis, psoriasiform hyperplasia, and dilated blood vessels consistent with psoriasis (**Figure 2**). There was also subtle pallor noted of the granular layer. Given atypical clinical features and overall poor treatment response

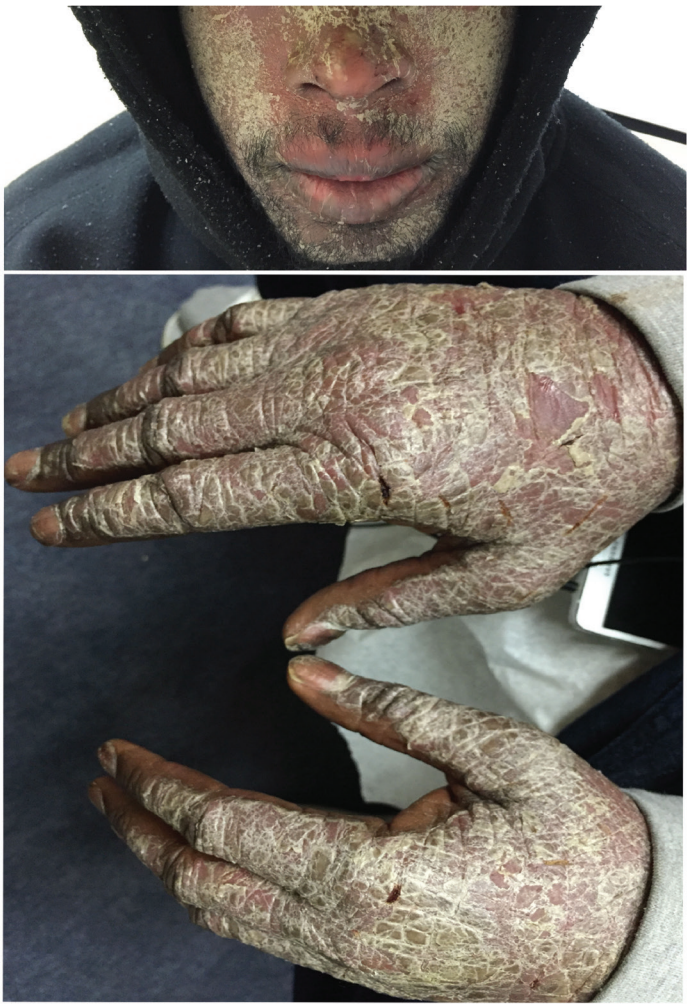

Figure 1. *Clinical photographs prior to treatment. Patient had thick plate-like scaling and fissures that were significantly worse in photodistributed areas such as the face and hands.*

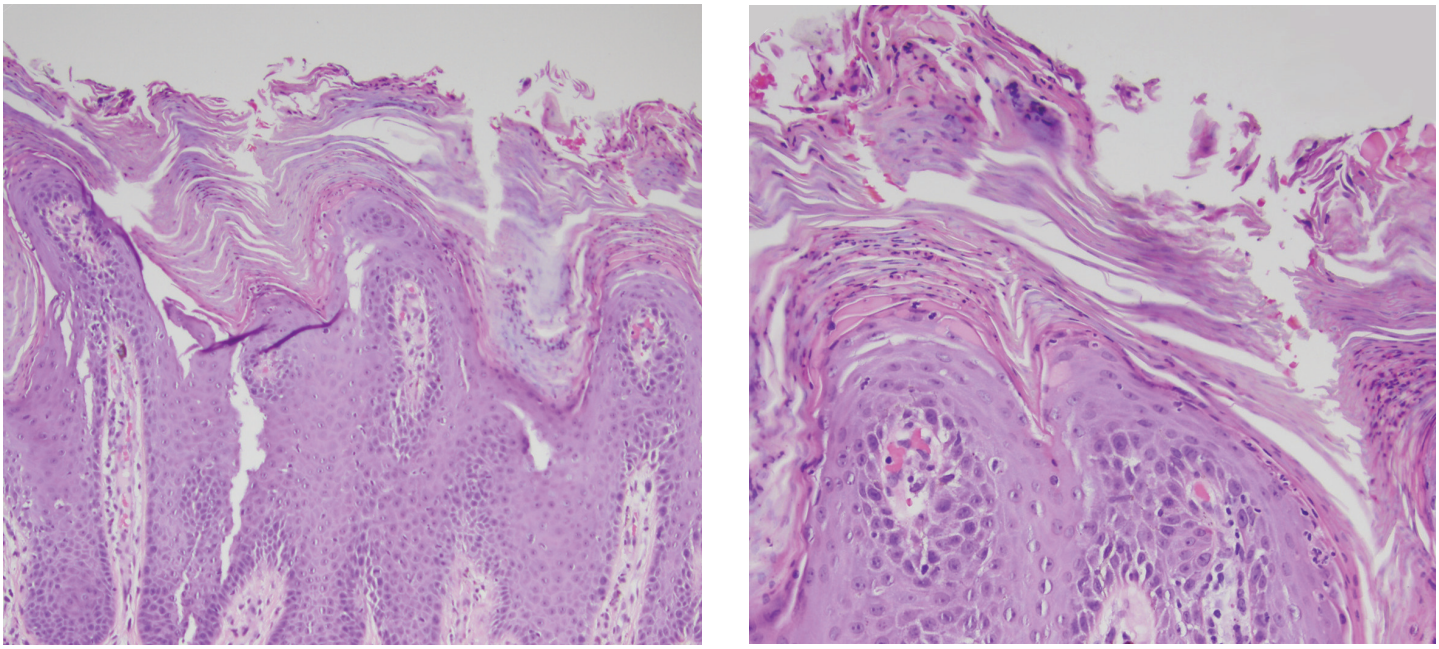

Figure 2. *Microscopic findingsprior to treatment. A) The medium power (10x) view demonstrates confluent parakeratosis and hypogranulosis overlying papillated epidermal hyperplasia. B) The high power view (20x) demonstrates aggregates of neutrophils within the cornified epidermis associated with subtle pallor of the granular layer. Within the papillary dermis are dilated vessels beneath a thinned epidermis. H&E.*

Figure 3. *Clinical photographs after treatment with oral niacin supplementation. Patient exhibited rapid improvement of scales and fissures after niacin supplementation.*

additional work up was obtained, including serology for autoimmune diseases. After a negative work-up, further investigation into other etiologies was pursued. Although the patient initially denied dietary restrictions and alcoholism, a serum nutritional panel was obtained. The serum nutritional panel was significant for undetectable levels of nicotinic acid and nicotinamide, normal cobolamin levels, and low thiamine levels. Upon further questioning, patient disclosed a significant drinking history. A diagnosis of concurrent pellagra was made and the patient was started on niacin and multivitamin supplementation and instructed to reduce alcohol intake, while continuing adalimumab and high potency topical steroids. Within two weeks his disease had markedly improved (**Figure 3**).

Case Discussion

Pellagra may present in the "classic triad", which includes photosensitive dermatitis, dementia, and diarrhea [7]. Few patients demonstrate the full triad, however. It has been reported that 33% of patients have dermatitis as a single finding [8]. There are a variety of biochemical etiologies that lead to photosensitivity in patients with pellagra, one of which is niacin deficiency. The deficiency in co-factors alters the process of skin cellular repair, either by impairing repair of UV damage or decreasing the energy transfer to rapidly turn over skin cells [4, 5].

Pellagra is more commonly seen in developing countries, but it can also be seen in developed areas in the setting of secondary causes of malnutrition, such as alcoholism [4, 5]. Alcoholism is known to interfere with nutrient absorption [4, 5], and chronic alcohol consumption can exacerbate existing nutritional deficits.

The diagnosis of pellagra is based on the clinical presentation, as well as rapid improvement following oral niacin supplementation. Biopsy findings can also support the diagnosis. However, as demonstrated by this case report, findings of psorisiform hyperplasia can be nonspecific and can closely resemble other conditions. Microscopic descriptions of pellagra include parakeratosis and epidermal pallor, the latter of which was subtlety present in this case [8]. Of note, when pallor is present, the lesions can histologically resemble acrodermatitis enteropathica and necrolytic migratory erythema.

Conclusion

In this case report, several clinical features were suggestive of an alternative diagnosis. The distribution of scaling and plaques in photodistributed areas is less commonly seen in the setting of psoriasis. Additionally, the diagnosis of pellagra also explains why the patient's dermatitis was refractory to multiple psoriasis treatment regimens and flared with phototherapy. His background psoriasis made it challenging to correctly diagnose, owing to clinical and histopathologic overlap with pellagra. Our case report highlights the overlap that psoriasis can have with pellagra at both the clinical and histologic levels and points out differentiating features. It is important to maintain an index of suspicion for a secondary process in treatment-resistant psoriasis.

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