Prurigo pigmentosa in a patient with newly diagnosed type one diabetes with diabetic ketoacidosis

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Abstract

Prurigo pigmentosa is a rare inflammatory cutaneous disorder associated with ketosis that resolves with reticulate hyperpigmentation. A 19-year-old man was admitted to the hospital with diabetic ketoacidosis. He also had developed reticulate papules coalescing into plaques over the posterior neck, back, and abdomen. Histopathological findings consistent with the clinical findings suggested the diagnosis of prurigo pigmentosa. After treatment with topical triamcinolone and oral doxycycline along with resolution of his ketosis after insulin administration, the patient's rash healed with reticulate hyperpigmentation.

Keywords: hyperpigmentation, ketosis, prurigo pigmentosa, type one diabetes

Introduction

Prurigo pigmentosa is a cutaneous inflammatory disorder identified by the sudden onset of pruritic and erythematous macules, urticarial papules, and plaques that coalesce to form a reticular pattern and heal with reticular hyperpigmentation [1]. An atypical presentation can occur as a vesiculobullous eruption [2]. Ketoacidosis has been associated with the development of this rash, as it is believed that ketone bodies accumulate around the blood vessels, leading to neutrophilic inflammation [2]. Prurigo pigmentosa has been reported in patients with diabetes mellitus, anorexia nervosa, and in patients fasting and dieting. Specifically, the ketogenic diet, a popular low-carbohydrate and high-fat diet, is associated with prurigo pigmentosa [1,3].

Histopathological findings for prurigo pigmentosa are unfortunately nonspecific. The early stage is characterized by superficial and perivascular dermal neutrophilic infiltrate and slight spongiosis. Later stages show lymphocytic and eosinophilic dominance in the upper dermis, epidermal parakeratosis, and marked spongiosis [1]. In the final resolution stage, there is a sparse lymphocytic dermal infiltrate along with upper dermal melanophages associated with post-inflammatory hyperpigmentation [3,4].

Case Synopsis

A 19-year-old man was diagnosed with type one diabetes after three weeks of polydipsia, polyphagia, and a five-pound weight loss. He was prescribed an insulin regimen including insulin glargine and insulin lispro. However, before he could begin his insulin regimen, he was found to have a blood glucose level of 516mg/dl, an anion gap of 20mEq/L, and was admitted to the hospital for diabetic ketoacidosis. He additionally developed a new pruritic cutaneous eruption that began one day before his type one diabetes diagnosis on the neck and spread to the back and abdomen.

Physical examination revealed pink minimally scaly reticulate papules coalescing into plaques over the

posterior neck, back, and abdomen. A few excoriations over the back were appreciated (**Figure 1**). Histologic analysis revealed spongiosis, epidermal acanthosis and parakeratosis, and dermal perivascular lymphocytic inflammatory cell infiltrate along with numerous eosinophils (**Figure 2**). The overall clinical and histologic findings were consistent with prurigo pigmentosa.

He was started on treatment with oral doxycycline 100mg twice a day and topical triamcinolone 0.1% ointment twice a day. His ketosis resolved after two days of insulin administration. After treatment with doxycycline and triamcinolone, the patient's cutaneous eruption resolved with residual reticulate hyperpigmentation (**Figure 3**).

Case Discussion

Prurigo pigmentosa is a rare inflammatory cutaneous disease that usually involves the upper back, abdomen, and chest and spares the mucous membranes, hair, and nails; it rarely involves the face or limbs. Prurigo pigmentosa tends to primarily affect young Asian women [5].

Figure 1. *Active prurigo pigmentosa with pink minimally scaly reticular papules coalescing into plaques over the back.*

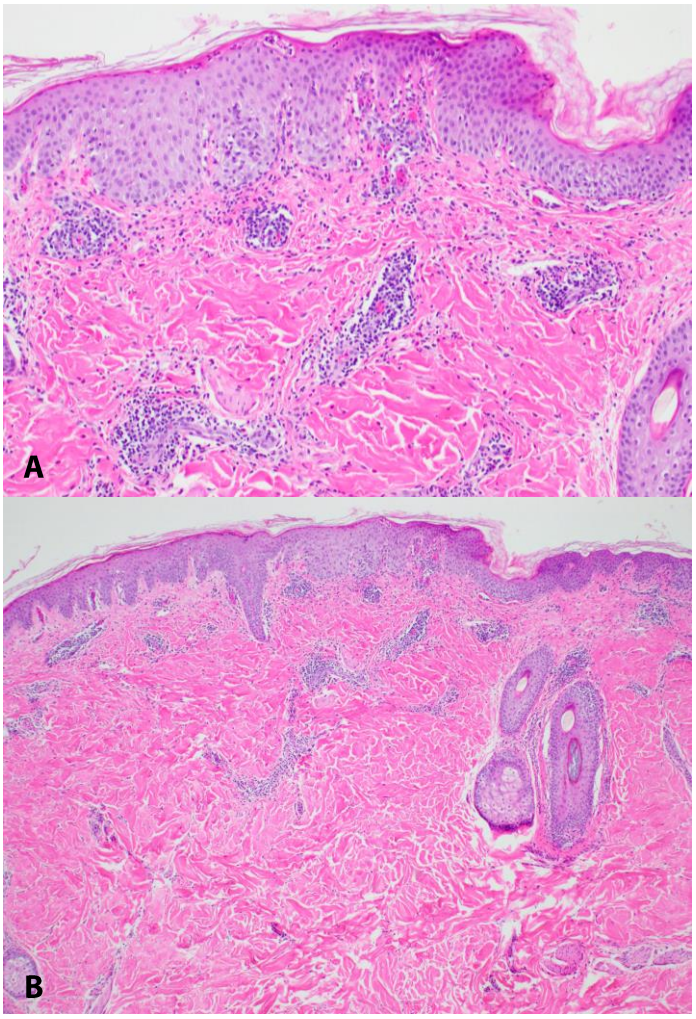

Figure 2. *Pathology slides of prurigo pigmentosa revealing spongiosis, epidermal acanthosis, parakeratosis, and dermal perivascular lymphocytic inflammatory cell infiltrate and numerous eosinophils.* ***A).*** *H&E, 4×.* ***B).*** *H&E, 10×.*

Tetracyclines are the preferred treatment owing to their effect on neutrophil migration and function and cytokine expression [1]. Notably, corticosteroids have not been shown to be helpful for prurigo pigmentosa but can help treat pruritus as in this case [5]. Control of blood ketone levels or cessation of a ketogenic diet have been beneficial if ketosis is the cause.

Several patients with prurigo pigmentosa have had associated diabetes (**Table 1**). A previous case report of a 49-year-old man with type two diabetes who was found unconscious with Kussmaul breathing with a rash was diagnosed as having prurigo pigmentosa. The rash resolved after improvement of acidosis and closing of the anion gap [3]. Another

Figure 3. *Resolution of prurigo pigmentosa with reticular post inflammatory hyperpigmentation.*

case report described spontaneous resolution of prurigo pigmentosa after five days of insulin administration in a 15-year-old boy with newly diagnosed type one diabetes mellitus with ketosis. The eruption resolved along with the ketosis, leaving some reticulate pigmentation [6]. Furthermore, another report described a 17-year-old boy who experienced resolution of prurigo pigmentosa with residual reticulate hyperpigmentation after 10 days of 100mg doxycycline daily [7]. Additionally, other physiologic states associated with increased ketonemia can be associated with prurigo pigmentosa. A 30-year-old pregnant woman who was hospitalized with severe hyperemesis developed prurigo pigmentosa which resolved after fluid therapy, weight stabilization, and bed rest [8].

One common dermatosis physicians should remember in the differential diagnosis of prurigo pigmentosa is Gougerot-Carteaud syndrome, a rare dermatosis which involves the development of scaly or verrucous papules that coalesce to form confluent plaques centrally with a reticulate pattern peripherally. What distinguishes this from prurigo pigmentosa is the lack of pruritus as well as histopathological features of compact hyperkeratosis, acanthosis, and papillomatosis [9].

Table 1. Prurigo pigmentosa in patients with diabetes mellitus.

Study [Reference]	Patient characteristics	Resolution of prurigo pigmentosa
Kobayashi et al. 1996 [10]	6-year-old girl with diabetes mellitus	Resolved after stabilization of blood glucose levels
Yamada et al. 2018 [3]	49-year-old man with type 2 diabetes	Improvement of ketoacidosis and closing of anion gap
Yokozeki et al. 2003 [6]	15-year-old man with likely type 1 diabetes	Insulin and improvement of ketosis
Kubuta et al. 1998 [2]	32-year-old man with diabetes mellitus	Resolved with minocycline and control of urine glucose and ketone levels with glibenclamide
Levran et al. 2020 [11]	22-year-old woman with type 1 diabetes	Resolved after increased carbohydrate intake to 75 grams per day
Ohnishi et al. 2000 [12]	21-year-old woman with insulin dependent diabetes	Resolved after insulin administration and dapsone

Conclusion

This present case demonstrates a case of prurigo pigmentosa in the setting of diabetic ketoacidosis. Endocrinologists, dermatologists, and hospitalists should consider prurigo pigmentosa when evaluating patients with symptoms of ketosis and a pruritic reticulate rash.

Potential conflicts of interest

Authors declare no conflicts of interest.

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