CASE REPORT

A case of prurigo pigmentosa aggravated by ketogenic diet and treated by carbohydrate rich diet

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ABSTRACT
Prurigo pigmentosa is a rare inflammatory skin disease more common in Asians. Usually presenting with itchy erythematous papules and plaques on the trunk and back, the rash takes a reticulated and interrupted shape which later on usually followed by postinflammatory reticulated hyperpigmentation.
Although the cause might be obscure in some cases yet, nickel, detergents, hypersensitivity to helicobacter pylori and metabolic ketotic states including diabetes and pregnancy had been found to be offending agents in reported cases.
Inflammation in prurigo pigmentosa was attributed to increase proliferation and hyperactivity of neutrophils and lymphocytes, the use of drugs such as tetracyclines and dapsone as a treatment leads to reduction of phospholipase A2 enzyme on the cell membrane and consequently reduces the cytokine activity and inflammation. Topical steroids proved to be ineffective in such cases but removal of the offending agent was considered to be the most practical as it was shown in our case.

INTRODUCTION
A young male patient presented for the first time in his life with reticulated patches of itchy maculopapular eruption mainly on the lateral aspects of the trunk especially on the right side. No history of exposure to nickel, detergents or chromium. No history of diabetes but the patient confirmed that he was on a carbohydrate free diet two weeks before the rash and still on diet. Relying on such a fact, patient was treated solely by adding carbohydrates to his diet and to our surprise the patient dramatically improved within five days for both the rash, erythema and the itching without any other treatment either topical or systemic. Clinical features, histopathological features and treatment of prurigo pigmentosa will be also discussed.

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he started a carbohydrate free diet two weeks back before the visit which meant and according to the history that the eruption started one week after the diet. A 5mm punch biopsy from each side of the trunk were taken and the diagnosis of prurigo pigmentosa was reconfirmed when histologic examination revealed marked spongiosis, exocytosis, superficial inflammatory infiltrate, and dermal haemorrhage while medium power revealed subcorneal pustules and interstitial infiltrate composed mainly of lymphocytes with the presence of neutrophils, additionally, scattered dermal melanophages in different areas were seen which denoted that the intensity of inflammation led to injury of dermo-epidermal junction and the presence of itching could be an aggravating factor for such an injury. Absence of acanthosis, hypergranulosis confirmed the early eruption and acuity of the lesions in spite of absence of necrotic keratinocytes. Parakeratosis was seen in some areas which confirmed the inflammatory process and explained the associated itching as well. Surprisingly, for the patient and for us as well, there was a dramatic improvement of the symptoms and rash within few days after discontinuation of the ketogenic diet solely and without topical or systemic treatment. In conclusion, the clinical signs and symptoms as well as the dermatopathological picture and the dramatic improvement after removing the precipitating factor were all suggestive of an early stage of prurigo pigmentosa due to carbohydrate free diet.

Fig. 1 Erythematous rash on the right side of the trunk

Fig. 2 The patches are reticulated, Erythematous and papulomacular.

Fig. 3 Low power x10, showing superficial inflammatory infiltrate, spongiosis, exocytosis and dermal haemorrhage.

Fig. 4 Medium power x20, subcorneal neutrophilic collection, spongiosis and exocytosis.
Prurigo pigmentosa is a rare inflammatory skin disease more common in adults, it was described for the first time by Nagashima in year 1971 and the disease was called after his name for 7 years till year 1978 when the term prurigo pigmentosa was used for the first time.\(^1\)

The disease has a unique distribution involving the upper back, clavicular areas, chest and scapular region as well,\(^2\) but in some cases it was seen intergluteal,\(^3\) lumbosacral, antecubital fossae and abdominal.\(^4\) Usually prurigo pigmentosa is common in spring and summer,\(^5,6,7\) could be due to contact allergy e.g. with chromium,\(^5,8\) humidity, detergents,\(^2\) Helicobacter pylori\(^12\) and strict dieting were also suggested as predisposing factors leading to occurrence and even recurrence of the disease.\(^2,10,11,13\)

Histopathologically, early lesions in prurigo pigmentosa usually show spongiosis, balloonning, necrotic keratinocytes & neutrophilic infiltrate perivascular and in upper dermis.\(^5,14\) In more mature lesions, lymphocytic infiltrate predominates and distribute in patchy lichenoid pattern, additionally, some neutrophils and eosinophils can be seen and there would be more liquefaction degeneration of basal layer with pigmentary incontinence.\(^5,14,15\) In late lesions hyperplasia and parakeratosis could be seen.\(^14,15,4\)

In our case the clinical picture and the history go with the early stage of the disease which could be proved dermatopathologically by the marked spongiosis, subcorneal pustules, neutrophils in the epidermis and dermis although necrotic keratinocytes could not be seen. Lack of epidermal hyperplasia, the insignificant parakeratosis and the absence of epidermal pigmentation also confirm acuity and absence of recurrent attacks. Although the clinical picture and symptoms were suggestive of ketosis as a causative agent yet, we checked the helicobacter pylori antigen in stool and it proved to be negative.

Clinical picture, history of carbohydrate free diet, dermatopathology and dramatic relief of symptoms after discontinuing the diet were all suggestive of an acute stage of prurigo pigmenta-
tosa.

Our recommendation if prurigo pigmentosa was confirmed clinically, we have to find out the offending agent before waiting for the results of skin biopsy to avoid exposure of the patient to more side effects or aggravation of inflammation and post inflammatory hyperpigmentation as well.

We also recommend checking the serum and urine ketone bodies if the patient is diabetic, fasting or on carbohydrate free diet and use mucin stain in dermatopathology to exclude reticulate erythematous mucinosis as a differential diagnosis. We also recommend leading further researches to study the effect of ketone bodies on the skin and whether it is a direct injurious effect or just a hypersensitivity reaction.

REFERENCES