CASE REPORT

# Recurrent prurigo pigmentosa after prolonged fasting in Ramadan and with recurrent ketogenic diet

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## ABSTRACT

Prurigo Pigmentosa is an uncommon disease. Usually presenting with recurrent itchy erythematous papules and plaques over trunk and back followed by reticulated hyperpigmentation, sometimes in rare occasions it might present with vesicular or bullous lesions. Topical and systemic steroids are ineffective, and treatment is usually with drugs which interfere with the function of neutrophils, reduces lymphocytic proliferation, and reduces phospopholipase A2 enzyme on the cell membranes of inflammatory cells thus reducing cytokine activity and consequently reduces inflammation, such drugs include tetracycline, dapsone, and macrolides.

## INTRODUCTION

A young female with a history of recurrent patches of erythematous maculo-papular eruption followed by reticulated hyper pigmentation after prolonged fasting in Ramadan and recurrent ketogenic dieting. The diagnosis of prurigo pigmentosa was suspected and dermatopathology was done but after the eruption of the third attack. Treatment with oral tetracycline was very effective although discontinuation of the ketogenic diet also resulted in dramatic improvement. Clinical features, histopatological features and treatment of prurigo pigmentosa will be discussed.

## CASE REPORT

A 22 years old Jordanian female visited emergency room in Sharjah University hospital with a heroically itchy papular and erythematous rash all over neck, chest and mid-back after a strict ketogenic diet; she was diagnosed as an acute irritant eczema to which she received topical steroids, systemic IV hydrocortisone and antihistamine but without improvement. After one week she visited skin clinic in Sharjah University hospital with an acneiform eruption which was attributed to the topical corticosteroids and to which she received doxycycline 100mg daily with dramatic improvement of the whole rash including the acneiform eruption. After two months in June 2014 she had a prolonged Ramadan fasting (20hours) when she was in Germany (sunset in Germany in June is 9:50 pm) and again she developed the same rash which dramatically healed after the end of Ramadan fasting. Four months later and after another low carbohydrate diet she started to have another attack and then she came again to Sharjah university skin clinic where a correlation between the rash and dieting was confirmed by histopathology. She had a 5mm punch skin biopsy from the chest area and the diagnosis of prurigo pigmentosa was

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confirmed when a low power view showing acanthosis, parakeratosis, hypergranulosis and perivascular inflammatory infiltrate composed mainly of lymphocytes, a picture suggestive of a fully developed stage of prurigo pigmentosa as perivascular lymphocytic infiltration was more prominent than neutrophilic infiltration also keratinocyte necrosis was not prominent as it used to be in early and acute stage, and in addition to that, the epidermal hyperplasia was conforming with the history of recurrent attacks.

According to the histopathological, clinical findings and dramatic improvement under short term tetracycline, a diagnosis of prurigo pigmentosa was suggested although a differential diagnosis of reticular erythematous mucinosis (REM) and acute lichen planus (LP) was put into consideration clinically. Histopathologically there was negative mucin stain which excluded REM, no band lymphocytic infiltrate, no melanophages, or basal vacuolar degeneration thus excluding LP; however, there was hypergranulosis and acanthosis. The dramatic alleviation of symptoms and rash after receiving a normal carbohydrate diet and/ or doxycycline were all suggestive of prurigo pigmentosa.

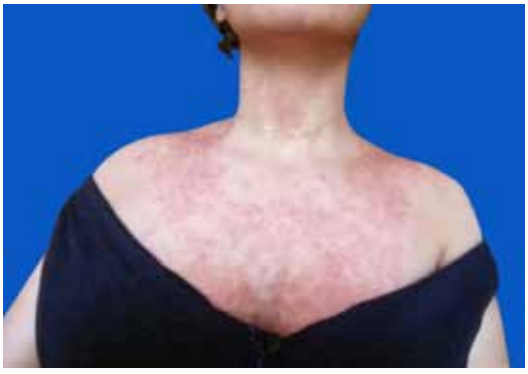


**Fig. 1** Ill defined erythematous and papular patches over the neck and chest.

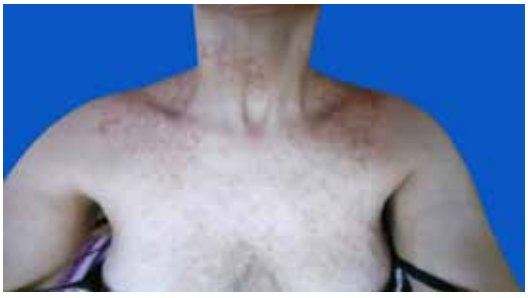
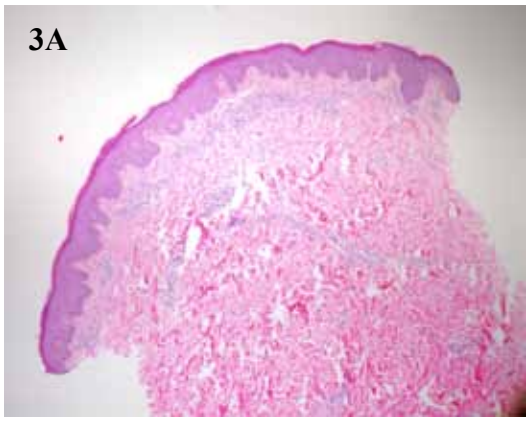


**Fig. 2** Post inflammatory hyperpigmentation after one week from stopping diet.



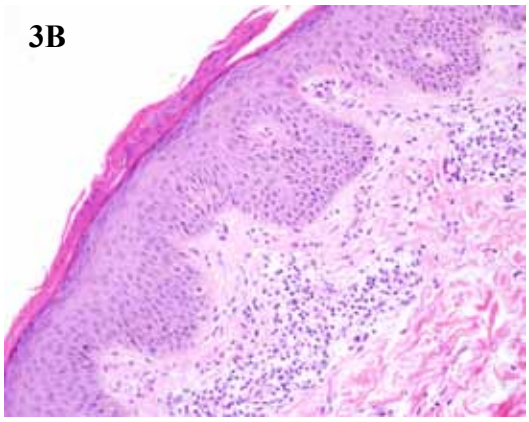




**Fig. 3.A.** Acanthosis, hypergranulosis, parakeratosis, perivascular inflammation. **B.** high power

## DISCUSSION

Prurigo Pigmentosa is a rare disease although more commonly prevalent in adult Japanese women where it was first described by Nagashima and was named after his name in 1971, was termed later in 1978 as prurigo pigmentosa.[1] The resolution of the primary lesions occurs within days leaving reticulate hyperpigmentation.[2,12] The distribution of the lesion in the back, including upper back, and scapular regions, clavicular re-

gions, and chest.[9] Sometimes, lesions may occur on abdomen, lumbosacral, forehead and antecubital fossae,[8] intergluteal area involvement was also reported.[11]

The total duration may range from 1 month to 11 years with a mean duration of 2.9 years[2] and in spite of such a chronicity, lichenification of lesions is very rare.[12] The female:male ratio is 2:1.[4,12] Usually prurigo pimentosa is mostly seen in spring and summer,[2, 5, 12] could be due to contact allergy e.g with chromium.[2, 3] Dietary change was suspected as a cause of prurigo pigmentosa.[9, 13] Strict diet as well as known associated factors like humidity were suggested as predisposing factors to the occurrence of prurigo pigmentosa.[9, 10, 13] The lesions in our case was basically related either to prolonged fasting or to ketogenic diet.

The diagnosis of prurigo pigmentosa in general is dependent upon clinical history and physical examination because histopathological findings are often non-specific, not only this but also varies according to the stage of the disease.[6] Early lesions show spongiosis, ballooning and necrotic keratinocytes, neutrophilic infiltrate perivascular and in upper epidermis.[2,6] In more mature lesions, lymphocytic infiltrate predominates and distributes in patchy lichenoid pattern. Some neutrophils and eosinophils can be seen, there would be more liquefactive degeneration of basal layer with pigmentary incontinence.[2, 6, 7] In late lesions hyperplasia and parakeratosis could be seen.[6, 7, 8]

In our case the lesions are mature with perivascular lymphocytic infiltrate predominance and rare neutrophils, in addition to prominent acanthosis, parakeratosis and hypergranulosis which are suggestive of recurrent attacks. Rare spongiosis and absence of necrotic keratinocytes are consistent with maturity of the lesions; furthermore, the pigmentary incontinence as well as the epidermal hyperpigmentation was not seen in spite of the recurrent attacks.

Our recommendation if REM could be suspected as a differential diagnosis, we should do mucin stain to confirm. If prurigo pigmentosa was confirmed clinically we have to put into consideration that staging of the disease is very important and it should be reconfirmed dermatopathologically whether it is in early, fully developed or late stage and if it was a first or a recurrent attack, we also recommend lab tests for serum and urine ketone bodies if prurigo pigmentosa was suspected especially, if the patient is diabetic, fasting or on strict ketogenic diet. Finally, if confluent reticulate papillomatoses could be also suspected clinically, a potassium hydroxide test should be needed to rule out Malassezia furfur.

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