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Case Report

Atypical adult onset pityriasis rubra pilaris: a rare chronic form of pityriasis rubra pilaris

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ABSTRACT

Pityriasis rubra pilaris (PRP) are a group of clinically similar papulo squamous dermatoses which present with erythematous, hyperkeratotic perifollicular papules which tend to coalesce to form plaques and may progress to erythroderma. We report a case of 46-year-old female patient who came with chief complaint of itching and burning sensation and scaly patches on extremities since one year. On dermatological examination well demarcated hyperkeratotic scaly plaques with follicular plugging and peripheral rim of erythema were seen over extensor and flexor aspects of elbows, knees, thighs, legs, dorsum of foot and gluteal region. Diffuse palmoplantar keratoderma, diffuse scaling and follicular plugging of the scalp, thickened brownish black nails were seen. On histopathology alternating layers of orthokeratosis and parakeratosis, follicular plugging with perifollicular parakeratosis, acanthosis with broad rete ridge, perivascular lymphocytic infiltration in upper dermis was seen, suggestive of PRP. Patient responded well to oral acitretin 25 mg once daily combined with oral methotrexate 10 mg once weekly along with topical emollients and corticosteroids after 6 weeks of treatment.

Keywords: Atypical adult onset PRP, Palmoplantar keratoderma, Unusual clinical presentation

INTRODUCTION

Pityriasis rubra pilaris (PRP) is a papulo squamous dermatosis of unknown cause characterized by reddish orange erythema, plaques with branny scales, palmo plantar keratoderma, and keratotic follicular papules. The pathogenesis of PRP is unknown. It has been divided into five clinical types by Griffiths and a sixth type related to human immune-deficiency virus (HIV) infection has been described.[1,2] Classical adult-onset PRP (type I) is the most common type of PRP and most common variant of PRP seen in adults. Atypical adult PRP (type II), classical juvenile PRP (type III), circumscribed juvenile PRP (type IV), atypical juvenile PRP (type V), HIV infection associated PRP (type VI) are the other clinical clinical types of PRP among which circumscribed juvenile PRP (type IV) is the most common type of PRP seen in children. We report a rare case of atypical adult onset PRP which is uncommon type, not commonly seen in clinical practice.

CASE REPORT

A 46 years female patient came with chief complaint of itching and burning sensation and peeling of skin over extremities since one year. Patient was apparently normal one year back when she developed itchy, raised lesions initially on front and back sides of forearms, elbows and knees. Later, she developed similar skin changes over the on dorsum of bilateral feet, legs, buttocks region and scalp. Current distribution of lesions was present since 3 months. No history of similar complaints in the past. Family history and drug history was not significant. Patient was well built and nourished, taking mixed diet, normal sleep, appetite,

bowel and bladder habits. Patient was not suffering from other chronic diseases.

On general physical examination no evidence of pallor, icterus, cyanosis, clubbing, lymphadenopathy and pedal edema. Examination of respiratory, cardiovascular, central nervous system and gastrointestinal system were normal. On dermatological examination, well demarcated hyperkeratotic scaly plaques with follicular plugging and peripheral erythema were seen over extensor and flexor aspects of elbows, knees, forearms, legs thighs, dorsum of bilateral feet and hands (Figures 1 and 2). Grouped follicular papules with scaling was seen over bilateral gluteal regions and shoulder regions. Diffuse palmoplantar keratoderma with thick scaling, desquamation, fissuring and erythema seen over bilateral palms and soles (Figure 3 and 4). Diffuse scaling and follicular plugging of scalp and thickened brownish black nails were seen. Mucosal surfaces were normal.

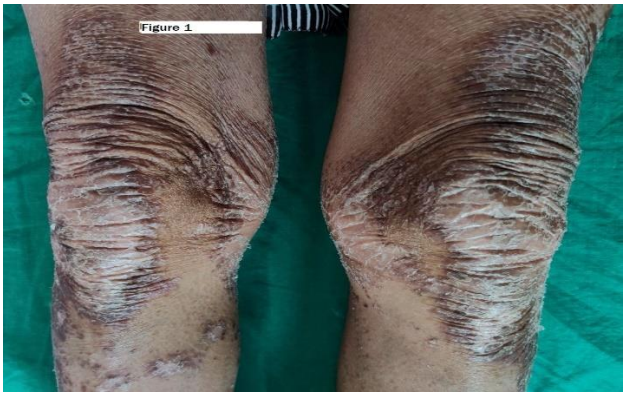

Figure 1: Well demarcated hyperkeratotic scaly plaques with follicular plugging and peripheral erythema over extensor flexor aspects of bilateral knee regions.

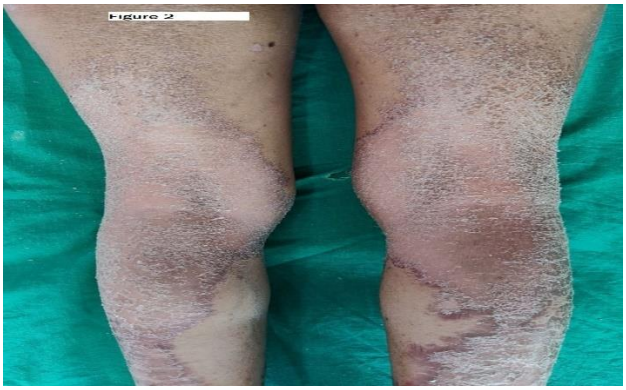

Figure 2: Hyperkeratotic scaly plaques with follicular plugging and peripheral rim of erythema over posterior aspects bilateral thighs and legs.

Based on history and clinical findings a differential diagnosis of pityriasis rubra pilaris, erythrokeratoderma variabilis, psoriasis, chronic dermatophytic infection and adult-onset atopic eczema were considered. Patient was subjected to blood investigation such as complete blood picture, random blood sugar (RBS), liver function test (LFT) and renal function test (RFT). Results of complete hemogram, serum biochemistry and viral screening for HIV, HCV, and HbsAg were normal. Imaging studies such as chest-X ray and ultra sound abdomen did not show any abnormalities. Four millimeters punch biopsy sample was taken from left shin region for histopathological examination which showed, epidermis with hyperkeratosis, alternating orthokeratosis and parakeratosis (Figure 5), follicular plugging with perifollicular parakeratosis, acanthosis with blunting of rete ridges and focal basal cell vacuolization. Upper dermis showed, dilated capillaries and perivascular infiltration in the upper dermis.

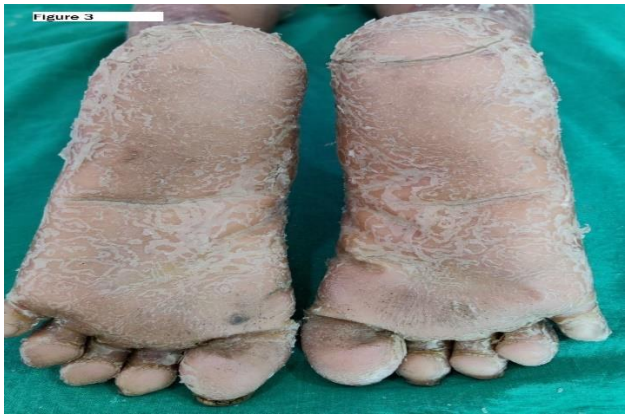

Figure 3: Erythema, thick scaling, desquamation and fissuring of bilateral soles.

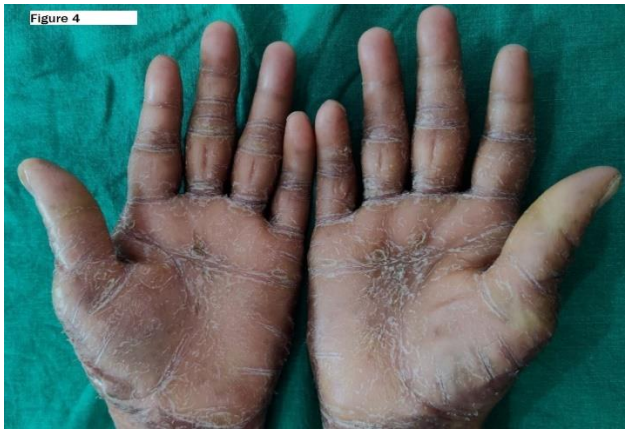

Figure 4: Erythema, lichenification, desquamation, scaling and fissuring of bilateral palms.

Based on clinical and histological findings, a diagnosis of pityriasis rubra pilaris was confirmed. Absence of generalized erythema, rapid spread, cephalo caudal progression and slow onset of localized lesions predominantly over extremities favoured a diagnosis of atypical adult onset pityriasis rubra pilaris (type II). The patient was treated with oral acitretin 25 mg once a day at bed time. She was also administered oral methotrexate 10 mg once weekly. Topical application of emollients on

lesions at morning time after bath and topical clobetasol propionate on lesions at night time was advised. Marked clinical improvement was observed after 6 weeks of treatment (Figures 6 and 7).

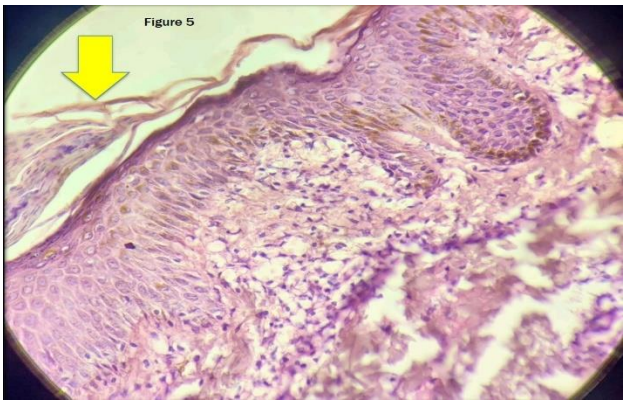

Figure 5: Hyperkeratosis, alternating layers of orthokeratosis and parakeratosis.

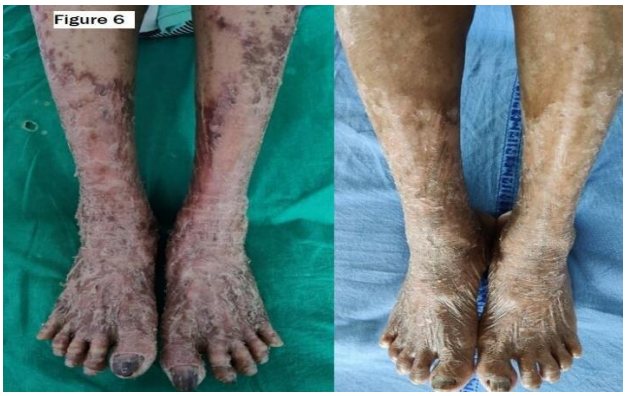

Figure 6: Hyper keratotic scaly plaques with follicular plugging, desquamation and peripheral rim of erythema over bilateral legs and dorsum of feet at the time of presentation and improvement of lesions with decreased erythema and scaling over bilateral legs and dorsum of feet after 6 weeks of treatment.

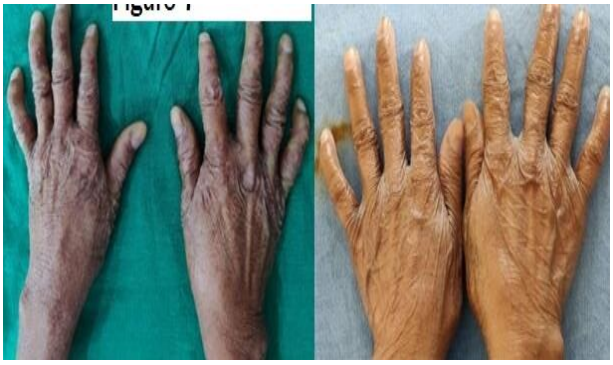

Figure 7: Erythematous, hyperkeratotic scaly plaques over bilateral dorsum of hands at the time of presentation and improvement of lesions with decreased scaling and erythema over bilateral dorsum of hands after 6 weeks of treatment.

DISCUSSION

Pityriasis rubra pilaris is also called as Devergie disease, lichem ruber acuminatus, pityriasis pilaris, keratosis universalis multiformis and keratosis circumscripta. Nearly all cases are acquired with occasional reports of familial forms.[3] The acquired forms of PRP have a bimodal age distribution with peaks in the first and fifth decades with equal sex incidence. The rare familial form of PRP starts in early childhood. Familial occurrence has been recorded in upto 6.5% of patients, with autosomal dominant inheritance, onset in such cases is usually in childhood. Autosomal recessive pattern has also been described in few cases. Streptococcal superantigens have been implicated in children with sporadic forms of PRP.[4,5]

It presents as well-defined salmon-red or orange-red dry scaly plaques, which may coalesce and become wide spread. Typically islands of normal skin are present, so-called as "nappesclaires".[6] The disease often starts on the scalp before spreading down over the rest of the body. Some patients may become erythrodermic. Pruritus may be present in the early stages of the disease. On the elbows, wrists and the backs of the fingers, characteristic follicular hyperkeratosis may be present – 'nutmeg grater' papules. Koebner phenomenon may be present.[7] Palms and soles may become thickened and fissured with an orange discoloration known as "PRP sandal". The nails are thickened, brittle, dull, and rough with splinter hemorrhage, subungual accumulation of keratinous debris can be seen.

Classical histopathological findings are alternating orthokeratosis and parakeratosis in both vertical and horizontal directions (checkerboard pattern)[8]; focal or confluent hypergranulosis; follicular plugging with perifollicular parakeratosis forming a shoulder effect. Thick suprapapillary plates, broad rete ridges, narrow dermal papillae, and sparse superficial perivascular infiltration, mostly of lymphocytes.[9] Histopathology of skin biopsy sample in our case study showed, epidermis with hyperkeratosis, alternating orthokeratosis and parakeratosis (Figure 5), follicular plugging with perifollicular parakeratosis, acanthosis with blunting of rete ridges, focal basal cell vacuolization were seen. Upper dermis showed, dilated capillaries and perivascular infiltration in the upper dermis.

Atypical adult onset PRP (type II) is a much more chronic form which constitutes only 5% of cases of PRP.[10] The scaling is rather more variable than in type I PRP: although peri follicular scale and palmoplantar keratoderma are both features, many patients show eczematous features and the keratoderma is coarser than in other types. The rapid progression of inflammation from the head down towards the feet as occurs in type I PRP does not occur and erythroderma is less common. In our current case study, on dermatological examination well demarcated hyperkeratotic scaly plaques with follicular plugging and peripheral erythema were seen over extensor and flexor

aspects of elbows, knees, forearms, legs, thighs, dorsum of bilateral foot and hands and gluteal region (Figure 1 and 2). Diffuse scaling and follicular plugging of scalp, erythema, thickening, scaling, desqamation and fissuring of bilateral palms and soles were seen (Figures 3 and 4). Thickened brownish black nails were seen. Mucosal surfaces were normal.

Common treatment modalities for PRP includes emollients, topical corticosteroids and oral acitretin up to 50 mg/day as 1st line agents. Topical calcipotriol, topical tazarotene and oral methotrexate up to 20 mg once weekly as 2nd line agents. TNF alpha antagonists, narrowband UVB phototherapy and extracorporeal photochemo-therapy as 3rd line agents.

The present case was treated with oral acitretin 25 mg once daily and oral methotrexate 10 mg once weekly along with topical emollients and topical corticosteroids which showed marked clinical improvement after 6 weeks. Patient was kept under regular follow up for 1 year in view of the chronic recurrent nature of the disease.

CONCLUSION

The clinical picture of keratoderma over palms and soles with coarse scaling over extremities and follicular papules over extremities with large diffuse, well marginated scaly plaques along with areas of follicular papules favoured a diagnosis of atypical adult onset of PRP.

The purpose of the current case report is to highlight the unusual clinical features of atypical adult onset PRP (type II) which is a rare form of PRP.

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