

Case Report

Circumscribed juvenile type pityriasis rubra pilaris (type 4): are we missing them?

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Abstract Pityriasis rubra pilaris (PRP) is a rare disease. The lack of pathognomonic features and specific clinical or diagnostic features often make identification of PRP very difficult. PRP type 4 (circumscribed juvenile type) is characterized by scaly plaques on elbows, knees and palmoplantar keratoderma. Sometimes these lesions may become extremely difficult to differentiate from those of psoriasis. We report 3 cases of PRP type 4, noted in just 5 months. We feel PRP, specially the type 4, is not so rare in Indian subcontinent. A good clinical knowledge and high index of suspicion may help to identify more cases.

Key words

Pityriasis rubra pilaris, circumscribed juvenile type, palmoplantar keratoderma

Introduction

Pityriasis rubra pilaris (PRP) is a chronic papulosquamous disorder of unknown etiology characterized by keratotic follicular papules, reddish orange scaly plaques and palmoplantar keratoderma. It was first described by Tarral in 1828 and was named by Besnier in 1889. Though the pathogenesis of PRP is not clear, theories include a dysfunction in keratinization or vitamin A metabolism, a physical trigger such as trauma, an autoimmune phenomenon, and a superantigen-mediated process.¹ In adults both sexes are equally affected but in children males are more commonly affected (male to female ratio, 3:2).²

Griffiths divided PRP into 5 categories: classic adult type (type 1), atypical adult type (type 2), classic juvenile type (type 3), circumscribed

juvenile type (type 4), and atypical juvenile type (type 5).¹ More recently, an HIV-associated type (Type 6) has been added to this classification system.³ A few reports have also described PRP associated with underlying malignancy.⁴

We report three cases of PRP (type 4) diagnosed in a period of only 5 months.

Case report

The clinical presentations of these patients are summarized in **Table 1** and **Figures 1-3**. All of them presented to our out-patient Department for evaluation of a skin condition previously diagnosed as ichthyosis or psoriasis by other clinicians. All were males in the age range of 7-11 years. The lesions appeared at age of 1-3 years and were initially limited to the soles. Gradually they spread to involve the hands, elbows and knees. Palmoplantar keratoderma was characteristically noted in all of them with follicular papules extending beyond the lateral border to the dorsum of

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Table 1 Clinical presentation of the patients.

No.	Age (yrs)/ Sex	Clinical features	Involvement of palms and soles	Extent of skin involve- ment	Family history	Preceding events	Lab abnorm- alities	Treatment
1	10/M	Itchy scaly follicular papules on knee, elbows	Thickening of skin with prominent fissures	10%	No	No	Hb- 9gm/dl	Oral vitamin A- 50,000 IU, multivitamins, topical retinoids, emollients
2	7/M	Itchy scaly follicular papules, plaques on knee, elbows, shins, ankles	Erythemat- ous thickening	15%	No	Recurrent upper respiratory tract infections	Hb - 8gm/dl, TLC- 11,800/ mm ³ ESR - 33mm 1 st hour	Oral vitamin A- 50,000 IU, multivitamins, keratolytic, topical steroid, emollients.
3	11/M	Itchy scaly follicular papules on knee, elbows, shins and ankles	Thickening of skin	15-18%	Similar history in sister	No	Hb -9.5 gm/dl	Oral vitamin A- 50,000 IU, multivitamins, keratolytics, topical steroids, emollients.

fingers and hands. The lesions were pruritic with occasional development of painful fissures and had winter exaggeration. The face, scalp, trunk, and nails were not involved. The patients had no known medical problems except mild anemia and one of them gave history of recurrent upper respiratory tract infections that might be a predisposing factor. History of similar complaints was present in the sister of one of them. Their growth and developmental milestones had been normal. There was no family history of psoriasis, ichthyosis, palmoplantar keratoderma, or other skin diseases.

Skin biopsy was done in all the patients and the histopathology was consistent with the diagnosis of PRP. Biopsy sample of patient 1 showed slight irregular psoriasiform hyperplasia of the epidermis with sparse superficial perivascular lymphohistiocytic infiltrate. The papillary dermis showed edema. Mild focal spongiosis

was present throughout the epidermis. The granular layer was prominent. Stratum corneum showed alternating zones of parakeratosis and orthokeratosis without neutrophils (**Figure 4**).

All the patients were prescribed oral vitamin A and emollients. Two received keratolytics and steroids, and only one received topical retinoid.

Discussion

PRP is a rare disease. The reported incidences in South Africa (1.5 case per year),⁵ and Spain (1.6 case per year)⁶ are fairly less than the Griffith's series (4 cases per year).⁷ In India, the reported incidence is 1:50,000. Sehgal reported 4 cases in 2000.⁸ Sarkar *et al.*⁹ described seven cases, including 2 cases of circumscribed juvenile type (type4).

The three most common important histologic features include hyperkeratosis with alternating



Figure 1 Scaly plaques with follicular papules on elbows (Case 1).



Figure 3 Scaly circumscribed plaques with follicular papules on knees, anterior legs and ventral aspect of ankles.



Figure 2 Diffuse desquamative hyperkeratosis of palms and soles, follicular papules with keratotic plugs and circumscribed scaly plaques on bilateral elbows, knees and ankles (Case 2).



Figure 4 Epidermis shows acanthosis with broad and blunted rete ridges, spongiosis, prominent granular layer, and alternating areas of orthokeratosis with parakeratosis. The papillary dermis shows edema with perivascular lymphohistiocytic infiltrate (H&E, X 40).

orthokeratosis and parakeratosis (checkerboard pattern) in the stratum corneum, focal or confluent hypergranulosis and follicular plugging with perifollicular parakeratosis

forming a shoulder effect. Psoriasis is a very close clinical differential. The presence of more prominent granular layer, follicular plugging, thick suprapapillary plates, blunt rete ridges, occasional acantholysis (reported as an additional finding in PRP)¹⁰ and the absence of dilated capillaries, neutrophils and epidermal pustulation may aid in differentiation of pityriasis rubra pilaris from psoriasis.¹¹

The treatment options include topical glucocorticoids, topical vitamin D analogues, topical and systemic retinoids (systemic drug of choice), methotrexate, azathioprine, cyclosporine, etc. Ultraviolet (UV) light may exacerbate PRP, but there are recent reports of successful treatment with psoralen plus UVA (PUVA photochemotherapy),¹² UVA1 phototherapy,¹³ and narrow-band UVB phototherapy.¹⁴ Newer treatment modalities like calcineurin inhibitors (pimecrolimus),¹⁵ biologicals (etanercept, infliximab),^{16,17} etc. have also been tried.

Type IV PRP (circumscribed juvenile) accounts for about 25% of all cases of PRP. It presents in the prepubertal children with lesions localized to the elbows, knees, ankles, and dorsal aspects of the hands and feet. In these sites, perifollicular papules with central keratotic plugs coalesce to form demarcated, scaly, erythematous plaques. A waxy, orange-red, diffuse, palmoplantar keratoderma is also commonly observed. The prognosis is uncertain. It rarely progresses with some reports of improvement in the late teenage years.

All our patients were males. Palmoplantar keratoderma was seen in all. There was no preceding illness except history of mild recurrent upper respiratory tract infections, noted in one. These findings are in contrast to the findings of Sarkar *et al.*⁹ where both the

cases were females, preceding infections were noted in both, and palms and soles were spared.

The classification of PRP is very confusing and there are at least three types of classifications. The relationship between classical PRP (type 1, 3) and other types (types 2, 4, 5) is obscure. The atypical varieties (types 2, 5) may represent a different group of follicular ichthyosis in the future. However the type 4 stands out for its unique clinical distribution and behaviour. PRP type 4 (circumscribed juvenile type) is characterized by scaly plaques on elbows, knees and palmoplantar keratoderma, often very identical to the lesions in psoriasis. Till now there are no standardized clinical and pathological diagnostic criteria for PRP. This makes identification and study of PRP more difficult.

Paucity of reports of PRP type 4, especially in the Indian literature, prompted us to report these cases here. Interestingly these three cases were diagnosed in only 5 months during the tenure of one of the authors (SK) in two different colleges (first two were seen in J.N.M.C. Sawangi and the last one in B. S. Medical College, Bankura). All these cases were seen earlier by different pediatricians, dermatologists but no diagnosis could be reached.

Though this high incidence may be purely coincidental, we feel that incidence of PRP, specially the type 4 (circumscribed juvenile type), is underestimated. A sound clinical knowledge and high index of suspicion may help in the better diagnosis of this subset of patients.

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