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JUVENILE PITYRIASIS RUBRA PILARIS- THREE CASE SERIES

Dermatology

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ABSTRACT

Pityriasis rubra pilaris (PRP) belongs to a group of rare papulo squamous disorders that in common have circumscribed follicular keratosis, orange red erythema, branny scales and palmoplantar keratoderma. PRP has no gender bias. Incidence in India is 1 in 50000 visits. We report a three case series of juvenile PRP with varied presentations.

KEYWORDS

Pityriasis Rubra Pilaris, Juvenile

INTRODUCTION:

Pityriasis rubra pilaris (PRP) was first described by Claudius Tarral in 1835 as a variant of psoriasis[1]. In the year 1857, Devergie considered it as separate entity. Griffith described it into six types[2]. PRP has a bimodal age of onset. The etiology is unclear but some authors suggest the role of vitamin A deficiency and infections in the causation of the disease. Here we report a 3 case series of PRP with each case belonging to a different spectrum of the juvenile classification.

CASE REPORT 1

A 10-year-old boy born of a consanguineous marriage presented to the skin OPD with complaints of scaly lesions all over the body from four years of age indicating a chronic course. Scaly lesions started over the trunk at the age of 4 years associated with itching. It progressed to involve the whole body in a haphazard manner. Systemic examination was normal. Dermatological examination revealed scaly erythematous plaques all over the body involving the face, scalp, chest, back, bilateral hands and legs with islands of sparing (Figure 1). Few papules were noted. Scaling and thickening present over the palms suggestive of PRP sandals. Nails showed pitting. With the above said findings, we came to a conclusion of *Atypical Juvenile PRP.*

CASE REPORT 2

A 9 year old boy presented to the skin OPD with complaints of itchy, scaly lesions over the knees, elbows, palms and face for the past 1 year. He developed scaly lesions over the palms 1 year back, which over the next one year developed over other sites. Dermatological examination revealed well circumscribed papules over bilateral elbows and knees, some of which coalesced to form plaques. He also had a few skin-coloured to hypo pigmented papules over the face. His palms and soles showed thickening and scaling (Figure 2). We made a diagnosis of *Circumscribed juvenile PRP* based on the above mentioned findings.

CASE REPORT 3

A 12 year old male child presented to our skin OPD with complaints of generalized raised scaly lesions for the past 2 years. He was apparently normal 2 years back after which he developed raised scaly lesions on the face which then progressively spread in a cephalo-caudal direction to involve the entire body. The lesions were associated with itching. On dermatological examination, he had scaly, papules over the face, bilateral upper and lower limbs as well as on the trunk (Figure 3). Palms and soles showed thickening. A diagnosis of *Classical Juvenile PRP* was made.

A biopsy was taken from all 3 patients and the histopathology showed common findings of hyperkeratosis, alternate orthokeratosis and parakeratosis in a "checker-board" pattern, spongiosis, broad and short Rete ridges, focal hypergranulosis and perivascular lymphocytic infiltrate (Figure 4).

DISCUSSION:

Pityriasis rubra pilaris also known as Lichen Ruber pilaris, Devergie's disease and Lichen Ruber Acuminatus is a rare folliculocentric disorder of keratinization.

Clinically PRP presents with classical features like wide spread, minute, follicular acuminate, scaly plaques distributed symmetrically on the trunk and limbs[3]. Branny scales with reddish orange erythema may also be appreciated. Horny follicular papules on the dorsum of the proximal phalanges of the fingers may impart a nutmeg grater appearance. Sometimes erythroderma with islands of normal skin known as Nappes Claires can be seen, which is diagnostic for the disease. The palms and soles are thickened and known as "PRP sandals".

Griffiths classified PRP into 6 types: type 1-classic adult type; type 2-atypical adult type; type 3-classic juvenile type; type 4-circumscribed juvenile type; type 5-atypical juvenile type and type 6-PRP associated with human immunodeficiency virus[2].

Classical juvenile PRP presents with a cephalo-caudal progression of lesions with presence of PRP sandals and Nappes Claires[4]. It is usually self-limiting and resolves within 3 years.

Circumscribed juvenile type has well-demarcated plaques of follicular plugging with variable erythema commonly over the knees and elbows.

Atypical juvenile type is of familial and runs a chronic course. It is usually associated with ichthyosis and erythrokeratoderma.

PRP should be differentiated from psoriasis, erythrokeratodermas, follicular ichthyosis, lichen scrofulosorum and other follicular keratosis like keratosis pilaris, lichen nitidus, keratosis spinulosa, to name a few[5].

Histopathology will show hyperkeratosis, characteristic alternate orthokeratosis and parakeratosis in a "checker-board" pattern, spongiosis, broad and short Rete ridges, focal hypergranulosis and perivascular lymphocytic infiltrate.

Treatment involves topical emollients and topical keratolytics for mild cases and they help by reducing the exfoliation and restoring the skin barrier function. In moderate cases, topical retinoids can be applied. Vitamin A supplements may be advised as well. Systemic retinoids like Isotretinoin 0.5-1mg/kg/day can be given. Other modalities of treatment include topical vitamin D analogues, methotrexate, PUVA, NB-UVB as well as biologics like etanercept, infliximab and secukinumab have found to be beneficial in some cases.

CONCLUSION

We have reported a three cases series of juvenile pityriasis rubra pilaris of different types with a predilection for males.

LEGENDS TO FIGURES

Figure 1: Erythematous, scaly lesions with islands of sparing on the trunk with associated thickening of palms.
Figure 2: Well-circumscribed papules coalescing into plaques present on the face, knees and elbows with associated palmar keratoderma.

Figure 3: Scaly papules on the forearm
Figure 4: Histopathological picture in low power and high power showing checkerboard pattern of orthokeratosis and parakeratosis, focal hypergranulosis and spongiosis.

FIGURE 1

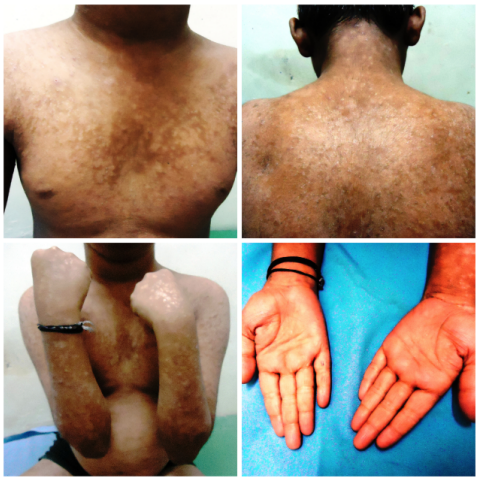


FIGURE 2

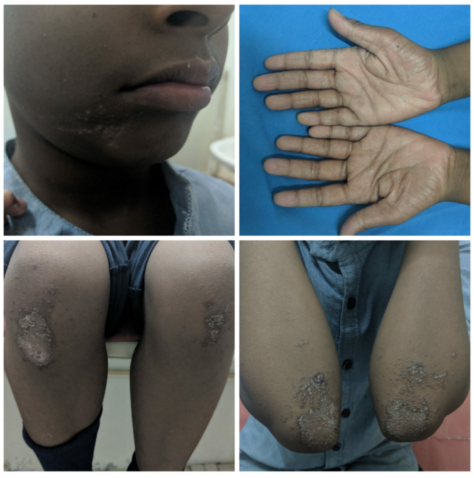


FIGURE 3

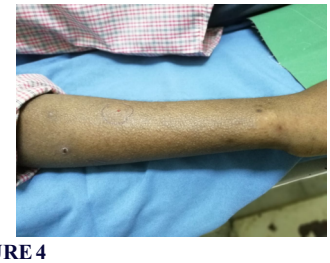
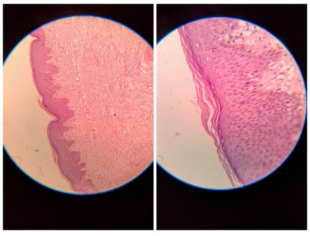


FIGURE 4



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