



Research Article

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Childhood pityriasis rubra pilaris-A case report

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ABSTRACT

Pityriasis rubra pilaris, a follicular hyperkeratotic disease affecting the skin can manifest in the young as well as elderly. The etiology is unknown and has a self remitting course in most of the cases. Recently an HIV associated type has been added to the classification because of its unique clinical features. Reporting of childhood pityriasis rubra pilaris is important to highlight the clinical features to the general physician.

Key words: Pityriasis rubra pilaris, papulosquamous dermatosis, follicular hyperkeratosis

INTRODUCTION

Pityriasis rubra pilaris (PRP) is an idiopathic papulosquamous keratotic dermatosis. It affects men and women equally with two peaks of age of onset in first and fifth to sixth decades. [1, 2] Six types of PRP have been described. Griffiths [3] divided PRP into 5 categories: type I - classic adult type, type II - atypical adult type, type III - classic juvenile type, type IV - circumscribed juvenile type, and type V - atypical juvenile type. More recently, an HIV-associated type [4] (type VI) has been added to this classification system which has same manifestation as type I along with acne conglobata, hidradenitis suppurativa and lichen spinulosus. Because of the rare reporting of the disease in childhood, we are reporting this classical case of childhood onset PRP.

EXPERIMENTAL SECTION

A fourteen years old girl presented with complaints of scaly papular rash over whole body since two months. The disease started as thickening and scaling over palms and soles 1 year back. Gradually she developed horny papular lesions over elbows and knees extending to trunk and extremities. On examination she had diffuse involvement of the skin (Figure 1 and 2) with follicular keratotic papules, coalescing to form scaly plaques over elbows, knees and back and areas of sparing in between. Bilateral palms (Figure 3) and soles were thickened and rough to touch.



Figure 1. Diffuse follicular keratotic papules with scaly plaques on elbows and upper and lower back

Figure 2. Scaly plaques on bilateral knees



Figure 3. Diffuse thickening and rough palms

There was diffuse scaling from the scalp. All mucosa were normal and there were no systemic complaints.

RESULTS AND DISCUSSION

Pityriasis rubra pilaris is an uncommon papulo-squamous disorder of unknown etiology. Classical juvenile onset PRP based on Griffiths classification type III presents as generalised coalescent hyperkeratotic follicular papules and plaques with islands of spared skin and frequently progressing to exfoliative erythroderma. Palms and soles are almost always involved and there is a distinctive orange hue of the affected skin. Pruritus occurs in approximately 20% of the patients. [5] Common differential diagnosis [6,7] includes psoriasis, seborrhoeic dermatitis, follicular eczema, follicular ichthyosis and lichen planopilaris.

Diagnosis is mainly clinical with supportive histopathological features of irregular acanthosis, alternating orthokeratosis and parakeratosis, hypergranulosis and sparse to moderate lymphohistiocytic infiltrate. Hair follicles are dilated and keratotic plug is present.

An universal standard treatment for PRP is lacking. Therapeutic options [8,9] include vitamins, retinoids, antimetabolites, immunosuppressive agents, phototherapy and biologic agents. Systemic retinoids yield excellent therapeutic results. [10] Because of a relatively favourable course in most patients with juvenile PRP, aggressive systemic treatment is usually unnecessary except in erythrodermic and disabling disease.

CONCLUSION

Because of the classical clinical features except in few cases, the diagnosis lies in the hands of the clinicians, mostly dermatologists and paediatricians. Also, in spite of the fiery look of the disease, usually it is a self remitting disease in children. So, a knowledge of the clinical picture of the disease would reduce the stress in part of the clinician as well as the patient.

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