



INFLAMMATORY SKIN DISEASES (OTHER THAN ATOPIC DERMATITIS & PSORIASIS)

PITYRIASIS RUBRA PILARIS: A RARE CHRONIC DISEASE

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Background: Pityriasis rubra pilaris (PRP) is a rare chronic papulosquamous disease of unknown etiology. It affects all races and there is no predilection for gender, with an incidence of 1 in 50000 in India. Clinically, PRP is characterized by reddish orange scaly plaques with islands of sparing, palmoplantar keratoderma and keratotic follicular papules. Lesions are symmetrical and usually spread caudally with possible development of erythroderma.

Observation: A 19-year-old boy presented with a nine-year history of a pruritic and scaly eruption on his scalp, torso, upper and lower limbs. Physical examination found sharply demarcated erythema of the trunk and extremities with follicular prominence of scale and distinct islands of sparing. The palms, plants and nails were not involved.

Skin biopsy reveals an alternating orthokeratosis and parakeratosis in both vertical and horizontal directions forming the checkerboard pattern, acanthosis and a superficial dermal lymphocytic infiltrate, consistent with PRP.

He was treated with oral isotretinoin 20mg daily for one year with partial response. The patient developed major depressive disorder and the drug was discontinued. It was started psoralen and ultraviolet A therapy that was also disappointing due to increased skin lesions severity. Methotrexate (12,5 mg weekly) was introduced with great improvement, but no complete remission was observed.

Key message: PRP is a clinically uncommon heterogeneous skin disease with a bimodal age distribution, being most common in the first and fifth decades of life and has been divided into six distinct subtypes. Despite of having type III pattern, that usually remits within one year, our patient presented no remission in ten years of follow up. Although systemic retinoids are considered first-line therapy, in our patient it had to be discontinued due to a side effect such as depression. Methotrexate is considered a second-line agent, and has been reported to be effective for difficult cases.

