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A RARE CASE OF PITYRIASIS RUBRA PILARIS

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ABSTRACT Pityriasis rubra pilaris (PRP) is a rare inflammatory papulosquamous skin disease that has six distinct types. The typical types (type 1 in adults and type 3 in children) are characterised by disseminated yellowish-pink scaly plaques surrounding islands of normal skin that show a cephalocaudal spread, and palmoplantar orange waxy keratoderma, whereas the atypical types (type 2 in adults and type 5 in children) are characterised by ichthyosiform scaling. Here we report a case of 1 year old girl child presenting with four-month history of red, scaly patches that began on the face and subsequently spread to the trunk and extremities, with thickening of the skin on the palms and soles and generalized pruritus. Examination revealed diffuse well defined eroded, hypo pigmented patches. Involved skin is sharply demarcated from adjacent uninvolved skin, producing characteristic islands of sparing. Scaling on palms and soles. Diagnosis of Pityriasis rubra pilaris was confirmed by Skin biopsy. Child was treated with topical corticosteroids, topical emollients and antihistamines. Child recovered well and is on follow up. PRP in paediatric patients poses unique challenges due to the rarity of its occurrence and the limited therapeutic options suitable for young children.. Despite its rarity, awareness about the presentation of PRP in young children can help clinicians make an early diagnosis leading to timely intervention and improved outcomes.

KEYWORDS : Skin, Papulosquamous, Hypo Pigmented Patches, Plaques, Paediatric, Inflammatory

CASE REPORT

Here we report a case of second born 1 year and 4 months old girl child born out of NCM, presenting with four-month history of generalised, well defined eroded, scaly erythematous patches that began on the face and subsequently spread to the trunk and extremities. It initially started as follicular papules progressing to scaly erythematous patches. The parents also noted thickening of the skin on the palms and soles, as well as generalized pruritus, which caused significant discomfort and irritability.

Not associated with fever/discharge.

No other complaints. Child was otherwise thriving well.

Child was exclusively breast fed upto 6 months of age. After which was started on complementary feeds. Child now on Direct breast feeds and complementary feeds. No H/O Bottle feeding. No H/O Bad child rearing practices. Developmentally normal for age child. Immunised upto age according to NIS. No H/O similar lesions, skin disease in family.

Examination revealed generalised, well defined eroded, hypo pigmented patches. Involved skin is sharply demarcated from adjacent uninvolved skin, producing characteristic islands of sparing.Scaling on palms and soles present.

Dermatologist opinion obtained and advised skin biopsy. Skin biopsy revealed hyperkeratosis with checkerboard pattern of orthokeratosis and parakeratosis, Granular layer was normal.

Child was treated with topical corticosteroid, topical immuno-suppressive agents, topical emollients and antihistamines, and oral Vit A for 2 weeks.

Child recovered well and is on follow up.

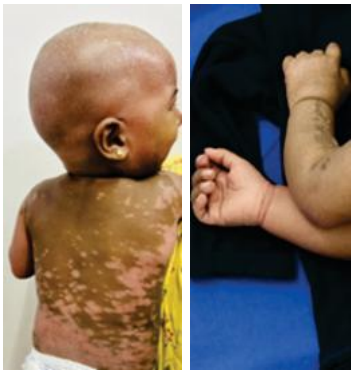

Fig-1: Before Treatment

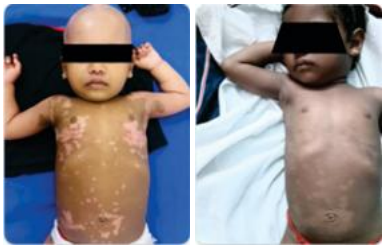

Fig-2: At Follow Up, After 6 Months

DISCUSSION

Pityriasis rubra pilaris (PRP) is a rare, chronic inflammatory skin disorder with a spectrum of clinical presentations. Most cases are acquired, but there are familial forms. Both autosomal dominant and less frequently autosomal recessive inheritance patterns have been described. PRP's pathogenesis is not well understood. [1] Still, there is evidence of increased expression of the interleukin IL-12/IL-23 and IL-17 in the affected skin. PRP comprises six types. The skin lesions in all types are generalized except in type 4, the most common type of PRP in children, where the lesions are localized to palms,elbows and knees. Overall, the most common type is type 1 [2]. It is classified into six types based on age of onset and specific clinical features. Types 1 and 3, seen in adults and children respectively, are characterized by widespread orange-red scaly plaques and palmoplantar keratoderma. In contrast, types 2 and 5, the atypical variants, present with ichthyosiform scaling and a more chronic course. Many cases cannot easily fit into any of these classifications. Patients may exhibit characteristics of one type that then evolve into another type. Most familial cases of PRP are type 5 which typically manifests as an autosomal dominant with a gain of function mutation on chromosome 17q25 in the caspase recruitment domain family 14 (CARD14) gene [3]. This gene is involved also in familial psoriasis vulgaris [4]. Interestingly, CARD14 was reported in familial as well as sporadic type 5 PRP [5].Characteristic features include the presence of well-demarcated plaques with characteristic islands of sparing and the thickening of skin on the palms and soles. Accurate diagnosis often requires a skin biopsy, which reveals hyperkeratosis, acanthosis, and focal parakeratosis.

Due to rarity of the disease, there are no randomized clinical trials on the treatment of PRP. Therefore, only retrospective case series and case reports - levels four and five of evidence - were the sources of recommendations.Management of PRP can be complex, particularly in pediatric cases where treatment options may be limited. Systemic retinoids, such as acitretin, are often the mainstay of treatment, alongside topical corticosteroids and emollients to manage symptoms in older children. Immunomodulatory therapies, like methotrexate and biologics, may be considered in refractory cases. PRP appears to be a self-limited disease, resolving within 3 to 5 years, except for non classic forms.

Oral retinoids (isotretinoin, acitretin, and alitretinoin) are considered the first-line agents for PRP. Adequate therapeutic trials of retinoids require at least four to six months. Phototherapy is one of the treatment modalities in PRP. Other systemic agents include methotrexate, biologics, TNF-α inhibitors, and Th17/IL-23 inhibitors, especially ustekinumab, cyclosporine, azathioprine, and apremilast [6,7]. Topical agents which include high-potency corticosteroids, tar, calcipotriene (calcipotriol), keratolytics, and tretinoin can be used as adjuncts to systemic therapy [8].

Pediatric PRP requires a tailored approach to minimize adverse effects and ensure effective management. Multidisciplinary care involving pediatricians,dermatologists and other specialists is crucial.

CONCLUSION

PRP is a challenging disease, whether in its pathology, diagnosis, or treatment. The rarity of PRP, coupled with its similarity to other dermatological conditions like psoriasis and eczema, makes diagnosis challenging. Raising awareness of PRP's clinical presentation among clinicians can lead to earlier diagnosis, timely intervention, and improved patient outcomes.

This case underscores the importance of heightened awareness and clinical vigilance among healthcare providers to recognize PRP's unique features. In pediatric cases, the limited therapeutic options necessitate a careful, tailored approach to minimize side effects and maximize efficacy.

Continued research into the pathogenesis and treatment of PRP will further enhance our understanding and ability to manage this rare condition, ultimately leading to better patient care and quality of life.

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