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Case Report

Pityriasis Rubra Pilaris in a 2-Year-Old Arabic Male; Case Report

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Abstract

Pityriasis rubra pilaris (PRP) is a skin disorder presenting with palmoplantar keratoderma and follicular hyperkeratotic papules that coalesce into large, scaly, erythematous plaques which may progress to exfoliative erythroderma. The exact incidence and pathogenesis are unknown. The diagnosis of PRP is based on the clinical signs and histopathological features mainly. Here we report the 23rd case of Pityriasis Rubra Pilaris type III reported in the medical literature.

Keywords: *Pityriasis Rubra Pilaris; Erythroderma; Psoriasis; Keratosis Pilaris*

Abbreviation

PRP: Pityriasis Rubra Pilaris

Introduction

Pityriasis rubra pilaris (PRP) is a skin disorder presenting with palmoplantar keratoderma and follicular hyperkeratotic papules that coalesce into large, scaly, erythematous plaques which may progress to exfoliative erythroderma.

Case Report

A 2-year-old Saudi male presented to our dermatology clinic at King Fahd Hospital of the University with a 10 days history of slightly pruritic skin rash. His past medical history was unremarkable. Family history, allergic history and drug history were negative. On examination the patient was active, afebrile, and his vital signs were normal. Upon examination of the skin there was well demarcated red-orange waxy scaly plaques with islands of sparing affecting elbows, knees, Trunk, the palms and soles symmetrically and bilaterally (Figure 1 and 2). Systemic examination was normal. Laboratory investigations including CBC, LFT, RFT and IgE level were normal. Two punch biopsies were performed at the back and left arm, it showed compact stratum corneum with focal parakeratosis, prominent granular layer, widening of rete ridges, mild spongiosis with follicular plugging confirming the diagnosis of Pityriasis Rubra Pilaris. The patient was started on topical Daivobet® ointment (Calcipotriol 50 micrograms and Betamethasone 0.5 mg) twice daily for 6 weeks with complete resolution of the condition after 12 weeks.

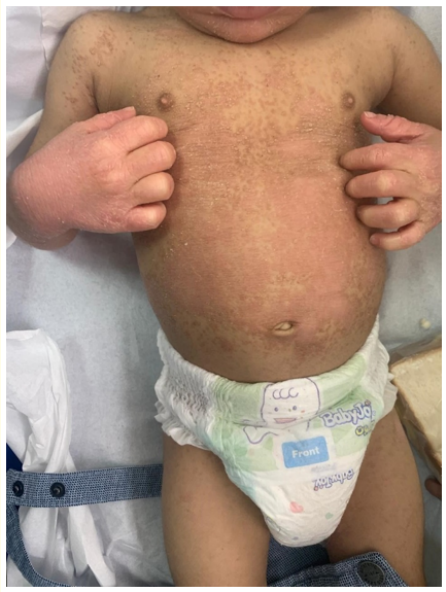

Figure 1a

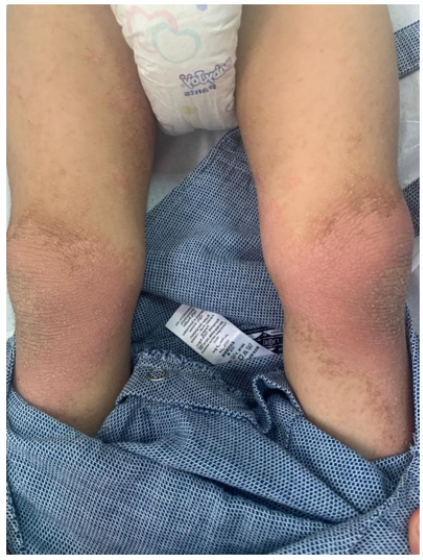

Figure 1b

Figure1a and 1b: *Well demarcated red-orange waxy scaly plaques with islands of sparing affecting the trunk and knees.*

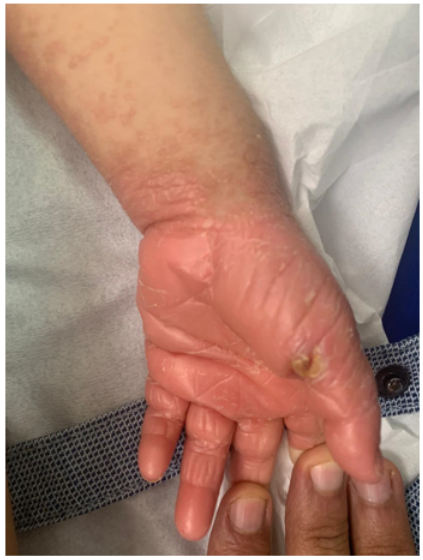

Figure 1a

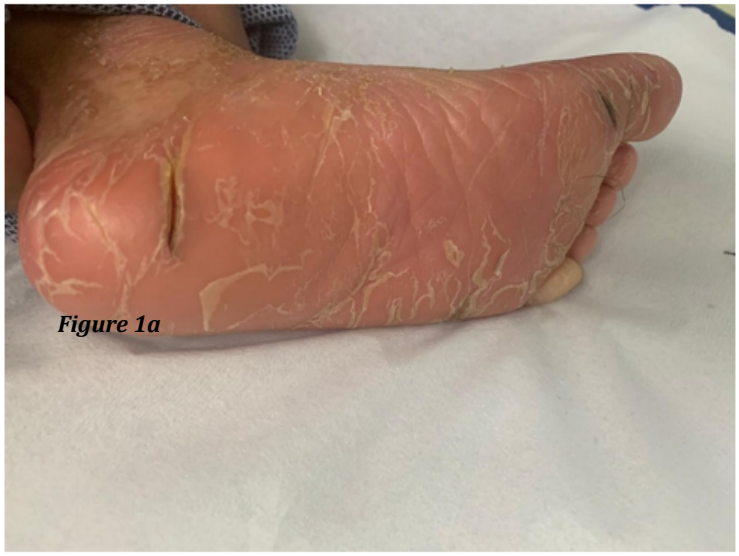

Figure 9

Discussion

Pityriasis rubra pilaris (PRP) is a rare inflammatory skin disorder presenting with palmoplantar keratoderma and follicular hyperkeratotic papules that coalesce into large, scaly, erythematous plaques which may progress to exfoliative erythroderma.

The exact incidence is unknown. In the pediatric group, an incidence of 1 in 500 new patients presenting with dermatologic disease has been reported [1]. PRP affects both sexes equally and has a bimodal distribution, peaking in the first and fifth decades of life [2]. In 1980, Griffiths classified PRP into five subtypes [2] and Miralles., *et al.* [3] added a type VI subtype.

Type I: Classical adult

The most common form accounting for up to 55% of all cases [2]. It is acute and initially affects the upper half of the body then the lesions spread caudally, affecting the trunk, arms, and legs. Lesions are red-orange keratotic follicular papules coalescing into plaques with islands of sparing. Erythroderma and a waxy palmoplantar keratoderma may occur. Nail changes include rough, thickened nails, yellow–brown discoloration, subungual hyperkeratosis and longitudinal ridging [2,4]. Other findings include lymphadenopathy and associated arthropathy [4,5]. This type has the best prognosis, with up to 80% spontaneous remission within 3 years [4].

Type II: Atypical adult

Accounts for 5% of all PRP cases [3]. Clinically, there is ichthyosiform dermatitis with a predilection for the lower extremities, coarse palmoplantar hyperkeratosis with lamellated scales and alopecia. It is usually chronic with less than 20% experiencing clinical resolution within 3 years [2,6].

Type III: Classic juvenile

Accounts for 10% of all cases [2]. It is similar to type I, with the exception of onset within first 2 years of life. The prognosis is good spontaneously resolving within 1 year [4]. There has been 22 cases of classic juvenile PRP reported in the medical literature with 3 cases has been reported previously in Saudi Arabia [7-10]. Our case is the 23rd case reported which might indicate a poorly reported condition or an uncommon condition.

Type IV: Circumscribed juvenile

Accounts for about 25% of cases [2]. It affects prepubertal children and presents with sharply demarcated areas of follicular hyperkeratosis and erythema over the elbows and knees. Prognosis is less favorable than classical juvenile PRP, with a third of patients achieving remission within 3 years.

Type V: Atypical juvenile

Accounts for 5% of cases [2]. It has an early onset, patients present with follicular hyperkeratosis and ichthyosiform features.

Type VI: HIV-associated

It occurs in HIV patients and have additional unique features such as follicular occlusion, acne conglobata, hidradenitis suppurativa, and lichen spinulosus-like lesions [3,11-13]. Most patients start developing erythematous desquamating follicular papules and prominent follicular plugging with formation of spicules. Erythroderma with islands of sparing is a frequent complication [4].

The pathogenesis of PRP unknown but several possible mechanisms have been postulated:

- **Genetics:** Familial forms have been commonly seen in type V PRP with autosomal dominant inheritance and variable penetrance, but autosomal recessive fashion has also been observed [14-16]. Autosomal dominant PRP has been linked to gain-of-function mutations in the caspase recruitment domain family, member 14 (CARD14) gene on chromosome 17q25 [15], related to the activity of genes involved in immune and inflammatory reactions.

- **Vitamin A metabolism abnormalities:** Similarities between the cutaneous manifestations of phrynoderma and PRP amongst Chinese patients and Ugandan prisoners led to early theories of vitamin A deficiency causing PRP [17-19]. However, administration of high doses of vitamin A has not always led to remission of PRP [20].

- **Infections:** PRP skin lesions may be the initial manifestation of underlying HIV disease [3,13]. Infections due to Staphylococcus aureus and Streptococcus pyogenes have been found in patients with juvenile PRP, and resolution of skin lesions after proper antibiotic treatment has been reported [21,22], which in turn has given rise to the theory of bacterial superantigens triggering PRP. Other infectious triggers that have been reported include HIV, cytomegalovirus, Epstein-Barr virus, hepatitis A virus, and varicella zoster virus [23-26].

- **Other factors:** Trauma in children [27,28], Photoaggravation [29,30], drugs such as kinase inhibitors [31-33], modern antivirals for hepatitis C [34,35] and topical imiquimod has been reported [36-38]. PRP has been associated with autoimmune diseases such as myasthenia gravis [35], autoimmune thyroiditis [40-42], celiac disease [43], vitiligo [44] and solid-organ malignancies [45-55].

The diagnosis of PRP is based on the clinical signs and histopathological features which include alternating orthokeratosis and parakeratosis (checkerboard' pattern), focal hypergranulosis, irregular acanthosis, thick supra-papillary plates, Sparse perivascular lymphohistiocytic infiltrate in the dermis and Follicular plugging with parakeratosis at the edges of follicular orifice (shoulder parakeratosis).

Depending on the type of PRP: Types I and III PRP may be confused with psoriasis. Types II and type V PRP may present with ichthyosiform changes. Type IV PRP, may be misdiagnosed as lichen spinulosus, keratosis pilaris, Darier's disease, pemphigus foliaceus, and epidermal nevus [52,53].

Evaluating treatment efficacy in PRP is confounded by its rarity, as well as its natural tendency towards spontaneous resolution. Favorable responses are observed when topical corticosteroids are used in pediatric patients or those with limited disease (e.g. type IV PRP). Other topical treatments that have been tried include topical retinoids, calcipotriol and calcineurin inhibitors. Oral retinoids such as acitretin, isotretinoin, and etretinate, are usually considered a first-line systemic treatment but is restricted in pediatric age group. Methotrexate, phototherapy and biologics including tumor necrosis factor-a inhibitors (TNFi) (infliximab, etanercept), ustekinumab and more recently, secukinumab have all been used in PRP. Other Therapies include: azathioprine, cyclosporine, apremilast, mycophenolate, intravenous immunoglobulin, penicillins, fumarates, and extracorporeal photopheresis [2].

Conclusion

PRP is a rare inflammatory dermatosis affecting both children and adults. Clinicopathologic correlation is essential in confirming the diagnosis, Treatment of PRP is challenging and spontaneous disease resolution is possible. Topical treatments should be considered as a preferred choice in patients with PRP, especially in pediatric age group, as in our case which is the 23rd case of Pityriasis Rubra Pilaris type III reported in the medical literature and who showed complete resolution of the condition with topical calcipotriene and betamethasone ointment for 12 weeks.

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