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Case report: Circumscribed Juvenile Pityriasis Rubra Pilaris

Tahani S Magliah*, Mazin M Aljabri, Malak A Aldahasi, Ahmed N Alharbi

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Authors' Affiliation:
Department of dermatology, Heraa General Hospital, Makkah, Kingdom of Saudi Arabia

Contact Information
Tahani S. Magliah: dr.tahanimagliah@gmail.com
Mazin M. Aljabri: mazinjabri@hotmail.com
Malak A. Aldahasi: malakaldahasi@hotmail.com
Ahmed N. Alharbi: Ahmadnaif001@gmail.com

*Corresponding Author
Consultant dermatologist, Department of dermatology, Heraa General Hospital,
Makkah, Saudi Arabia
Email: dr.tahanimagliah@gmail.com

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ABSTRACT

We report juvenile Circumscribed Pytriasis Rubra Pilaris in 13 year Saudi Boy, came to dermatology clinic with well circumscribed scaly plaques over elbows, knees and ankles, not improving on topical steroids. Histopathology was in consistent to Pytriasis rubra Pilaris. There are six types of Pytriasis rubra pilaris, one of them is Juvenile circumscribed type which is uncommon to be seen.

Keywords: Skin Diseases, Pityriasis rubra pilaris (PRP), juvenile Circumscribed Pytriasis Rubra Pilaris

1. INTRODUCTION

Pityriasis rubra pilaris (PRP) is known as rare papulosquamous disorder of unknown etiology consisting of follicular keratotic papules, orange–red scaling plaques and palmoplantar keratoderma (Arnold & Buechner, 2004; Martin et al., 2014; Zeeshan et al., 2020). In 1835, Claudius Tarral was the first one who described Pityriasis rubra pilaris (PRP) as a variant of psoriasis, later on, in 1857 Devergie recognized it as a separate entity and named it "pityriasis pilaris". In 1889 Besnier was named it "pityriasis rubra pilaris" (Bolognia et al., 2018). PRP has no gender bias; both males and females are affected equally. The incidence has two peaks concentrating in the first and second decades and fifth to sixth decades, Most cases are acquired, But there are reports of familial forms, with inheritance patterns describe autosomal dominant mainly and less frequently autosomal recessive (Bolognia et al., 2018; Shahidullah & Aldridge, 1994; Yang et al., 2008).

Griffiths proposed a classification of PRP into five types based on their clinical features, age of onset, and duration (Bolognia et al., 2018; Arnold & Buechner, 2004). Whereas, a sixth type has more recently described in patients associated with HIV infection. The most common PRP Types are type I (classic adult) and type III (classic juvenile) they represent clinically the same disease but differ in the age at onset (Bolognia Jean et al., 2018; Arnold & Buechner, 2004). Both are typically self-limited, clearing within 3 years in majority of patients, there features are Perifollicular keratotic papules, Red–orange plaques with islands of sparing and Waxy palmoplantar keratoderma. Type II is also seen in adults and differs from type I by a palmoplantar keratoderma with scales which is lamellated and course in shape, a more ichthyosiform

scaling on the lower extremities, and occasional alopecia. Only minority of type II patients show majority clearance within 3 years (Bolognia et al., 2018).

PRP types III–V are seen in children and adolescents. Type IV also called (circumscribed) type , is the commonest type of the PRP in children, and its focal nature with well-defined involvement, frequently affecting the knees and elbows distinguish it from the other PRP forms. Type V, the atypical juvenile form, has features similar to adult type II disease with more hyperkeratosis scaling and a chronic course. Moreover, scleroderma-like changes of the fingers have been described. PRP type VI is associated with HIV infection and it often fails to respond to conventional therapy for PRP, it may improve with antiretroviral therapy (Bolognia et al., 2018). Herein, we report a case of Circumscribed Juvenile Pityriasis Rubra Pilaris in a young boy, which has been treated with topical on calciptriol (50 microgram/g) ointment successfully.

2. CASE REPORT

This is A 13 year old Saudi boy, presented to dermatology clinic, with 1 year history of skin lesions, started over both knees and ankles, progressing to elbows, sometimes itchy, progressing, with mild palmar hyperkeratosis and scaling, visited many clinics and given different topical treatments: Topical potent and intermediate corticosteroids, Topical antifungal and Topical Antibiotics but no improvement. The boy has unremarkable medical history, no joint pain or stiffness, no systemic complains no physical activity limitation. This skin rash is not preceded by fever, respiratory or gastrointestinal infections, not preceded by vaccination or any other new medication. No history of allergy to food or drug, positive family history of the same lesions (father) but disappeared while growing.

On examination: over extensor surfaces of the knees, ankles and elbows there are will defined slivery scaly plaques, surrounded by normal skin, no secondary changes. No mucous membrane involvement. No scalp and nail involvement. No diffuse palmoplantar keratoderma (Figure 1 and 2).

4mm skin punch biopsy (right knee) was sent to Histopathology. Microscopic findings showed: Epidermis shows regular acanthosis and Psoriasiform hyperplasia, Alternating orthokeratosis and parakeratosis in both vertical and horizontal directions. The hyperkeratosis tracks down the opening of follicular structures forming follicular plugs. The dermis contain sever mixed inflammatory cells infiltrate. So the picture of punch biopsy was in consistent to Pytriasis rubra Pilaris (Fig 3, 4 and 5). Patient was started on calciptriol (50 microgram/g) ointment once per day applying topically for 1 month, patient mentioned moderate improvement, and decrease scale thickening, lessen itching.

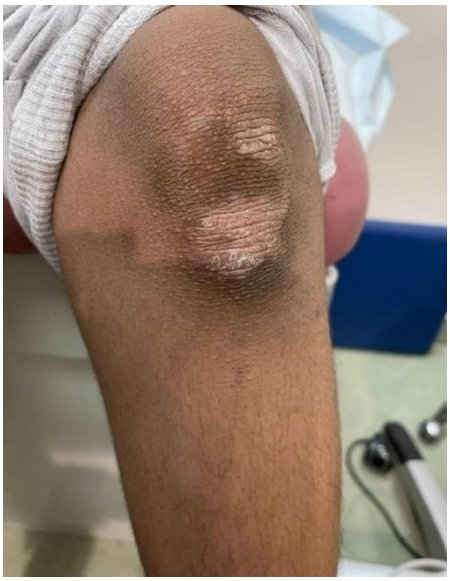

Figure 1 well defined erythematous scally plaque with some follicular prominence over right knee

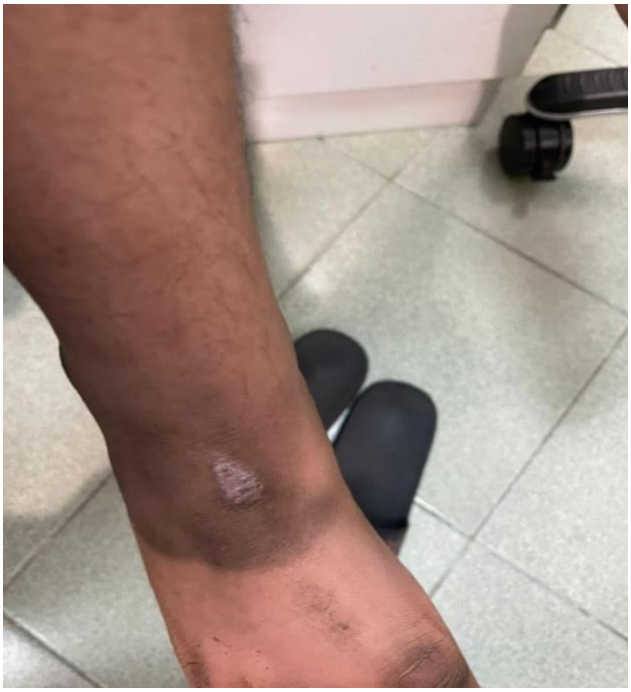

Figure 2 well defined erythrematous scaly plaque over left ankle

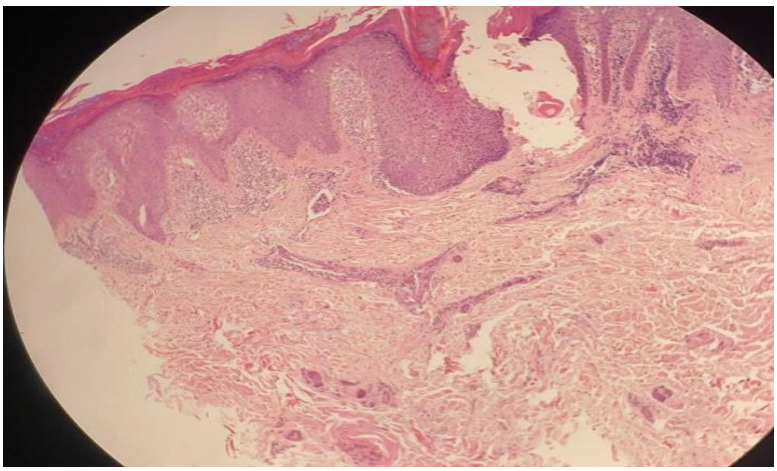

Figure 3 low power histopathological picture showed acanthosis and psoriasiform hyperplasia

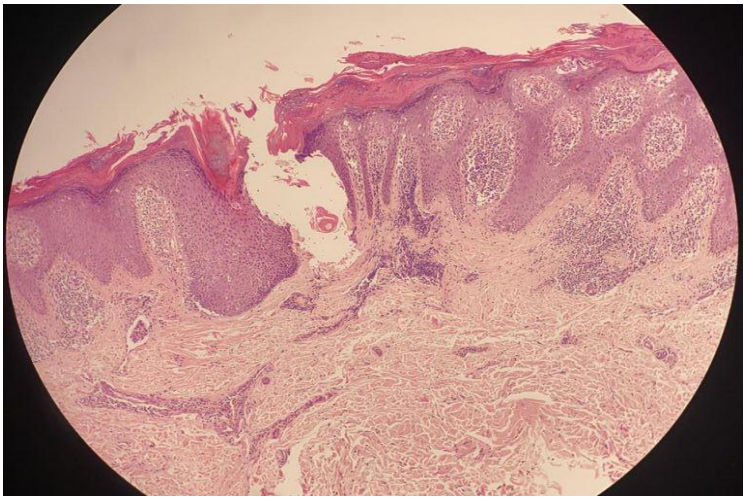

Figure 4 histopathological picture under high power showed follicular changes

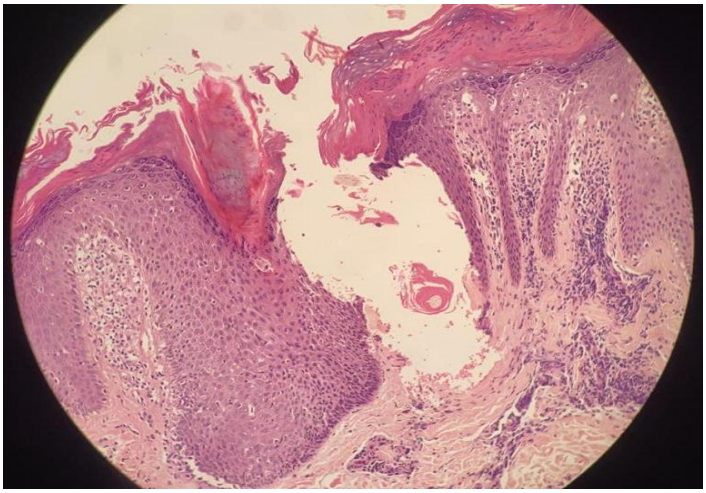

Figure 5 slide showing orthokeratosis, Parakeratosis and dermal mixed inflammatory infiltrate

3. DISCUSSION

Pityriasis rubra pilaris (PRP) is a rare papulosquamous disorder of unknown etiology, the key finding in classic PRP is follicular keratotic papules, especially on the dorsal aspect of the proximal fingers. These papules are also seen on the trunk, upper and lower limbs, and they form large orange red to salamon colored plaques with distinctive "islands of sparing", also palms and soles are commonly involved with keratoderma (palmoplantar keratoderma) (Arnold & Buechner, 2004; Martin et al., 2014; Zeeshan et al., 2020; Bolognia et al., 2018). Griffiths classified PRP into five distinct categories. Our case is diagnosed as Type IV circumscribed juvenile form which is the most common forms of juvenile PRP, with – as photos – presented as well defined involvement, and in this type usually affecting the knees and elbows, distinguishing it from the other PRP forms (Bolognia et al., 2018).

The Diagnosis of pityriasis rubra pilaris cannot be made only based on the histologic features, microscopic evaluation can be helpful to rule out other disorders with similar clinical characteristics, to rule out other differential diagnosis (Cohen & Prystowsky, 1989). Pityriasis rubra pilaris has characteristic features in classic presentation, the other clinical differential such as papulosquamous and erythrodermic disorders must be considered (Cohen & Prystowsky, 1989). Psoriasis has a major entity in the differential diagnosis of PRP. The distinctive orange–red palmoplantar keratoderma and the keratotic follicular papules, the classic islands of sparing of the trunk, a finer scale with negative family history of psoriasis may differentiate PRP from psoriasis (Bolognia et al., 2018).

In histological picture, there is acantholysis and focal acantholytic dyskeratosis, if present, are especially helpful in diagnosing PRP, hypergranulosis, short thicker rete pegs, limited vascular dilation in the dermal papillae, and the absence of both neutrophilic migration towards the epidermis and Munro micro abscesses further help distinguish PRP from psoriasis. Histopathological findings in the PRP group, included psorasiform hyperplasia, follicular plugging, and hypergranulosis, the latter two variables being of important value with respect to separating PRP from psoriasis, as Psoriasis has hypogranulosis (Bolognia et al., 2018; Magro & Crowson, 1997). The goals of treatment in PRP are to reduce morbidity of the disease course and to prevent possible complications. Because of unknown etiology, the rarity of the disease and the spontaneous resolution nature course of the disease, it was difficult to assess the outcomes treatment, and the current treatment is primarily empiric. Topical therapies are typically ineffective for most patients with PRP, unless there is limited disease. However it's can be used in adjuncts to systemic therapy, as high-potency corticosteroids, tar, calcipotriene (calcipotriol), keratolytics, and tretinoin.

Multiple Systemic therapies could be used according to patient status, including systemic retinoids, methotrexate (MTX), azathioprine, cyclosporine A (CsA), vitamin D anlogues, fumaric acid esters, stanozolol, and also phototherapy have all been used with variable success in cases of PRP after the initial reports of successful treatments for PRP involved oral vitamin A. However, because of the toxic mega doses of vitamin A and its liver toxicity, Systemic retinoids, such as acitretin and isotretinoin and are widely used as first-line therapy, they have good effects. Isotretinoin, in doses of 1–1.5 mg/kg per day, can induce significant clearing within 3–6 months of instituting therapy. Higher doses (up to 2 mg/kg per day) are occasionally required. Acitretin is also beneficial in PRP treatment. Methotrexate has also been reported to lead to significant clinical improvement of PRP. Weekly oral or subcutaneous doses of 10–25 mg are usually administered, and responses are seen within 3–6 months.

Hepatotoxicity and myelosuppression are well notable side effects. When more toxic, but not yet proven to be more effective, regimens are used (e.g. methotrexate 5 days per week), In severe cases of PRP, methotrexate and systemic retinoids has been tried to be given in combination to each other, but the evidence for efficacy is inconsistent. Both medications can be given at the beginning of therapy, or the second medication can be added if the response to the first is inadequate. But an increased risk for hepatotoxicity must be kept in mind. Biologics like TNF-α inhibitors (etanercept, infliximab, adalimumab), secukinumab and ustekinumab have been reported in few puplication for the treatment of resistance cases. Presently, the most recent successful infliximab use has been shown in a patient after ineffective pretreatment with cyclosporine and acitretin therapy. Other potential therapies include systemic immunosuppressives (e.g. azathioprine, prednisone, cyclosporine) and, in response to reports of decreased serum levels of retinol binding proteins in patients with PRP, anabolic steroids. However, these medications have met with variable results.

Although UV light therapy runs the risk of exacerbating the disease, there are some reports of success with narrow band UVB, UVA1, or PUVA in combination with an oral retinoid 43; even one patient with UVB-provoked PRP reportedly responded to bath PUVA. Lower doses are recommended for the classic juvenile form of PRP, given the high likelihood of spontaneous remission and the potential long-term effects of aggressive systemic therapies (Bolognia et al., 2018; Müller et al., 2008).

4. CONCLUSION

Juvenile Circumscribed Pytriasis rubra pilaris is uncommon dermatological disorder, seen in children, many lines of treatment might be considered, diagnosed clinically and histopathological, need to see more cases to know more about prognosis and effectiveness of the treatment.

Authors' Contributions

Dr. Tahani S. Magliah, Dermatology consultant, MD (Principle Investigator and corresponding author) - Abstract, introduction, case report, discussion and conclusion; Dr. Mazin M. Aljabri, Dermatology consultant, MD (Co-Author) - Abstract and discussion; Malak A. Aldahasi, MD (Co-Author) - Introduction, case report and discussion; Ahmed N. Alharbi, MD (Co-Author) - Abstract and conclusion

Informed consent

Informed consent was obtained from the patient.

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Conflicts of interest

The authors declare that there are no conflicts of interests.

Data and materials availability

All data associated with this study are present in the paper.

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