Original Research Paper

Volume - 12 | Issue - 11 | November - 2022 | PRINT ISSN No. 2249 - 555X | DOI : 10.36106/ijar

Dermatology

A RARE CASE REPORT OF PITYRIASIS RUBRA PILARIS

Dr. T. Ajay Pradeep* Final year postgraduate, Sri Venkateswara Medical College, DR.NTR University of Health Sciences. *Corresponding Author

Dr. Chandni Nair Final year postgraduate, Sri Venkateswara Medical College, DR.NTR University of Health Sciences.

ABSTRACT PITYRIASIS RUBRA PILARIS is a rare papulosquamous disorder of unidentified etiology. Shows bimodal age distribution. It is mostly acquired , occasionally familial with autosomal dominant inheritance. Hereby we report a rare case of Pityriasis Rubra Pilaris in a 20 year old male patient.

KEYWORDS : pityriasis rubra pilaris, papulosquamous disorder," nutmeg grater" papules, PRP sandal, horny keratotic papules

INTRODUCTION

Pityriasis rubra pilaris (PRP) is a rare chronic papulosquamous disorder of unknown etiology. It is characterized by well-defined orange red erythematous plaques with branny scales which coalesce and become widespread with typical islands of normal skin. It is rare condition, with incidence of 1:50000 patients in INDIA.

CASE REPORT

A 20 years old male patient, who came to our hospital with complaints of itchy , reddish raised scaly skin lesions over scalp, both upper and lower limbs , trunk, buttocks, palms and soles since 2 months. Initially, the lesions started over scalp and then it progressed to involve torso and limbs. He went initially to a local practitioner and was treated as seborrheic dermatitis and used ketoconazole lotion for 2 weeks and not improved.

On cutaneous examination, multiple, symmetrical, well to ill - defined erythematous, dry scaly plaques with surrounding horny follicular papules present over trunk, forearms, arms, legs, thighs and buttocks. Scalp – showing diffuse erythema and scaling. Palms and soles – showing hyperkeratosis with exaggerated skin markings. Hair, nail and mucosa– normal. Patient was advised for biopsy but not willing. It was diagnosed as Pityriasis Rubra pilaris (type 1) based on clinical history and examination. Routine investigations (including liver function and serum fasting lipid profile, HIV- non reactive) were within normal limits. The patient was treated with oral isotretinoin, topical betamethasone lotion, oral vitamin supplements and emollients for months and improvement in lesions seen.

Figure 1: photograph showing small follicular pink yellow scaly papules which coalesce to form plaques with islands of normal skin.

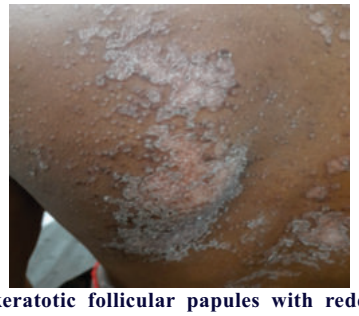

Figure 2: keratotic follicular papules with reddish orange erythema.

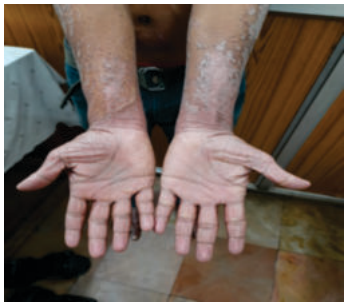

Figure 3: hyperkeratosis of palms

DISCUSSION

Pityriasis rubra pilaris (PRP) is a rare inflammatory papulosquamous disorder of unknown etiology. There are 6 distinct subtypes which occur in both children and adult. Fundamental elements noted across all subtypes include distinct, well-demarcated plaques of various sizes with characteristic reddish-orange hue, may have varying degrees of scaling. More generalized subtypes often demonstrate intervening areas of unaffected skin, known as "islands of sparing," which is a signature characteristic of PRP.

There is a broad spectrum of presentations from mild disease isolated to extremities to severe disease at times evolving into diffuse erythroderma. (1)

Exact etiology of PRP remains elusive. PRP occurs worldwide with no predilection for gender or ethnicity. There is a lack of information in the literature regarding the true incidence or prevalence of PRP. Indian study suggests 1 in 50,000 visits. There appears to be a bimodal age distribution with peaks during the first to second and fifth to sixth decades of life. Most cases appear to be sporadic, though familial cases have been reported in up to 6.5% of cases. Familial inheritance pattern appears to be autosomal dominant with variable penetrance in most cases. (3)

Pathogenesis of PRP remains unclear though several prominent hypotheses exist including dysfunction in keratinization or vitamin A metabolism, autoimmune mechanism, abnormal immunologic triggers, such as infection or ultraviolet (UV) exposure. Genetics seem to play a role in the development of at least some cases of PRP, most prominently noted in the type-V PRP variant. A clear association with a gain-of-function mutation in the CARD14, also known as (PSOR2), a gene that encodes for the member 14 protein. Human immunodeficiency virus (HIV) has been associated with the development of type-VI PRP.

Histologically PRP presents with irregular, psoriasiform acanthosis of the epidermis along with a very characteristic overlying "checkerboard" pattern of parakeratosis (alternating vertical and horizontal ortho-and parakeratosis). Often predominant follicular plugging with shoulder parakeratosis is noted. Thick, rather than thin, suprapapillary plates and lack of neutrophils in the stratum corneum may help distinguish PRP from psoriasis. Variable mild superficial

perivascular lymphohistiocytic infiltrate, occasional epidermal spongiosis and focal acantholytic dyskeratosis have been reported but are not consistently found.

The spectrum of PRP has been divided 5 distinct subtypes using a widely accepted classification scheme initially proposed by Griffiths in 1980 publication. The original 5 subtypes were based upon the age of onset, lesion distribution, and prognosis. Subsequently, a sixth subtype was added due to a recent association with HIV which presents as a distinct entity.

- Type I: classical adult onset
- Type II: atypical adult onset
- Type III: classical juvenile onset
- Type IV: circumscribed juvenile onset
- Type V: atypical juvenile onset
- Type VI: HIV-associated

The cardinal features that appear across subtypes in variable degrees include red-orange papules and plaques, hyperkeratotic follicular papules, and palmoplantar hyperkeratosis. Lesions are generally asymptomatic though a small portion of patients endorses mild pruritus. Type I PRP is the classic adult variant which affects 55% of patients and is the most common in adults. The most distinguishing features are classic red-orange papules and plaques with islands of sparing, perifollicular keratotic papules, and waxy palmoplantar keratoderma. This variant is typically self-limited. Prognosis is good since eighty percent of these patients resolve within 3 years. The classic sequence of signs in type I PRP begins with a diffuse fine scale seen on the scalp, which may be mistaken for seborrheic dermatitis. Scalp findings might initially be mild, however, often rapidly progresses to diffuse scaly erythema of the scalp. Similarly, patients may provide a history of solitary red-orange to salmon color macule, or small group of follicular papules noted on either dorsal fingers or head and neck. Within weeks to months, more macules and papules erupt and often coalescing into patches and plaques that progress in a cephalocaudal manner appear.

Characteristic hyperkeratotic follicular papules are prominent both within the patches of erythema and within adjoining areas of uninvolved skin. Various terms including “nutmeg grater” or “exaggerated gooseflesh” have been used to describe these follicular-based papules. Involved skin is often sharply demarcated from adjacent uninvolved skin producing a very characteristic phenomenon coined “islands of sparing” or “skip areas”. In soles, unique red-orange waxy palmoplantar keratoderma (PPK) so-called “sandal-like” PPK . At times, the disease may even evolve in to diffuse erythroderma. Complications of erythroderma including subsequent ectropion have been reported.

PRP is diagnosed based on classic clinical and histopathological features. The treatment of PRP can be challenging, and no universal approach exists. Topical agents which have shown promise especially in mild disease include emollients, keratolytic agents, such as urea, salicylic acid, or alpha-hydroxy acid containing preparations, topical corticosteroids, tazarotene, and topical calcineurin inhibitors.

Generally accepted first-line systemic agent for both adults and children is oral retinoids. Oral isotretinoin, 1 to 1.5 mg/kg per day, has been shown to induce clearance in as little as 3 to 6 months. Methotrexate alone or in combination with oral retinoids has also been used frequently. However, combination therapy increased the risk for systemic toxicity. Recently, some case reports and small case series have shown a possible role for biologic immunosuppressive agents typically used in the treatment of psoriasis such as TNF-alpha inhibitors and secukinumab and ustekinumab. Although the risk of photo-triggered and/or photoactivated disease UV-light therapy has been implemented in certain cases, including narrowband UVB, UVA1, or PUVA in combination with an oral retinoid with some success. (2)

CONCLUSION

PRP is a very rare skin disorder that may be encountered by the primary care provider and nurse practitioner. Because there is no laboratory test to make a diagnosis, the patient should be referred to the dermatologist for further workup. Once diagnosed, the treatment can be challenging. In many cases, PRP is self-limited and asymptomatic therefore does not necessarily require treatment. There are no treatments approved by the US Food and Drug Administration. Most practitioners recommend combination therapy with topical for symptomatic management and systemic therapy aimed at reducing inflammation.

Conflict Of Interest: Nil

REFERENCES

1. Eastham AB. Pityriasis Rubra Pilaris. JAMA Dermatol. 2019 Mar 01;155(3):404..
2. Kromer C, Sabat R, Celis D, Mössner R. Systemic therapies of pityriasis rubra pilaris: a systematic review. J Dtsch Dermatol Ges. 2019 Mar;17(3):243-259.
3. Gál B, Göblös A, Danis J, Farkas K, Sulák A, Varga E, Nagy N, Széll M, Kemény L, Bata-Csörgő Z. The management and genetic background of pityriasis rubra pilaris: a single-centre experience. J Eur Acad Dermatol Venereol. 2019 May;33(5):944-949.