ORIGINAL RESEARCH PAPER

Volume-7 | Issue-8 | August-2018 | PRINT ISSN No 2277 - 8179

INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

PITYRIASIS RUBRA PILARIS WITH FACIAL DYSMORPHISM – A RARE CASE REPORT

Dermatology

Dr. Paavai.s Junior residents, Department of Dermatology, Sree Balaji Medical College, and Hospital, Chrompet, Chennai.

Dr. Sivaramakrishnan. S* Junior residents, Department of Dermatology, Sree Balaji Medical College, and Hospital, Chrompet, Chennai.*Corresponding Author

Dr. Jayakar Thomas Professor and HOD, Department of Dermatology, Sree Balaji Medical College, and Hospital, Chrompet, Chennai.

KEYWORDS

INTRODUCTION

Pityriasis rubra pilaris is a rare chronic papulosquamous disorder of unknown etiology. It is characterised by reddish orange erythema, plaques with branny scales, pamoplantar keratoderma, and keratotic follicular papules. Familial occurrence has been recorded in upto 6.5% of patients, with the pattern of inheritance generally being autosomal dominant. Autosomal recessive has also been described in few cases. There are six types of PRP according to Griffith and Gonzales- Lopez classification.

CASE REPORT

A 9-year-old boy born to third degree consanguinity couple brought to skin OPD with complaints of scaly lesions all over the body from four years of age. Scaly lesion started over face at the age of 3 years and progressed to involve whole body. History of itching present. History of atopy present. No h/o photosensitivity. History of delay in developmental mile stone present. No similar complaints in the family. Systemic examination was normal. Local examination revealed scaly erythematous plaque all over the body involving face, chest, back, b/l hands and legs. Few papules were noted. Scaling and thickening present over the palms. Scaling present over the scalp. Oral cavity leucoplakia present involving tongue. Few pitting was noted in nails. Patient had slanting eyes, low set ears, synophrys. 3.5mm punch biopsy was taken from the left hand. Histopathology showed orthokeratosis and parakeratosis in both vertical and horizontal directions (checkerboard pattern), hyperkeratosis, hypergranulosis, spongiosis, perivascular lymphocytic infiltration is seen.

DISCUSSION

PRP is rare chronic papulosquamous disorder of unknown etiology. The incidence of PRP in india has been reported to be 1 in 50000.three distinct age groups of peak occurrences have been noted, early childhood up to 10 years, late childhood 11 -19 years, and adulthood 40 – 60 years. In adult both sex are equally affected, but in children males are more commonly affected.

Based on the classification proposed by Griffiths, six types of PRP can be distinguished: classic adult PRP (Type I) with generalized skin involvement and mostly spontaneous remission within 1 – 3 years; atypical adult (Type II) with generalized lesion distribution, but lond duration of the disease course; Classic Juvenile (Type III), which is similar to Type I in clinical presentation, but appears in year 1 or 2 of life; circumscribed juvenile(Type IV), which has localized skin involvement, but unpredictable prognosis; atypical juvenile (Type V), begins in first few years of life, accounts for most familial cases, chronic course; and human immunodeficiency virus-associated (Type VI).

Clinically PRP is characterized by wide spread, small, follicular acuminate, pinkish-yellow, scaly plaques distributed symmetrically on the trunk and limbs. The barnny and reddish orange erythema is present. On the scalp, diffuse erythema and scaling simulating seborrheic dermatitis occurs. Horny follicular papules on the back of the proximal phalanges of the fingers may feel like a nutmeg grater. Lesions usually starts from scalp and progress caudally. Sometimes erythroderma with islands of normal skin within affected areas (nappesclaires), which is diagnostic. The yellowish hyperkeratosis of the palms and sole is often known as "PRP sandal". Pruritis and burning sensation have been reported in 20% of the patients. Kaposi varicelliform eruption can occur.

The histopathalogic criteria for diagnosis of PRP include alternating orthokeratosis and parakeratosis in both vertical and horizontal directions (checkerboard pattern), focal or hypergranulosis, follicular plugging with perifollicular parakeratosis forming a shoulder effect, sparse superficial perivascular infiltration, mostly of lymphocytes.

Differential diagnosis includes erythrokeratodermas, psoriasis, lichen scrofulosorum, follicular ichthyosis, and othe follicular keratoses.

Treatment include topical emollients and mild keratolytics like 3% salicylic ointment or 20% urea may be beneficial. Oral retinoids are the first line of treatment. For children isotretinoin 1 -2.2 mg/kg/day for 4 to 6 weeks. Oral vitamin A 50,000 IU t.i.d is given. other options are extracorporeal photochemotherapy, cyclosporine, methotrexate, azathioprine, infliximab, efalizumab, etarnacept, and ustekinumab.

CONCLUSION

This case has been diagnosed as PRP both clinically and histopatho logically. This case is reported for its rare association with facial dysmorphism.

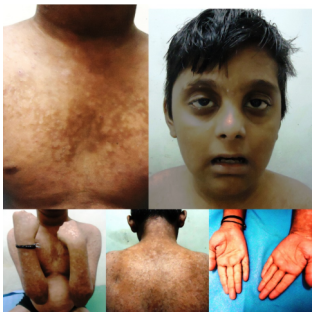

Figure 1

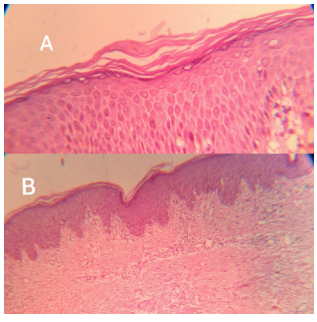

Figure 2

Figure 1: Showing scaly lesion over face chest, back, b/l upper limb. Scaly lesions over palms. slanting eyes, low set ears, synophrys.
Figure 2A: Orthokeratosis and parakeratosis in both vertical and horizontal directions (checkerboard pattern).
Figure 2B: Hyperkeratosis, hypergranulosis, spongiosis, perivascular lymphocytic infiltration is seen.

References

1. Griffiths W. Pityriasis rubra pilaris. Clin Exp Dermatol 1980;5:105-12.
2. Allison DS, El-Azhary RA, Calobrisi SD, Dicken CH. Pityriasis rubra pilaris in children. J Am Acad Dermatol 2002;47:386-9.
3. Vergilis-Kalner IJ, Mann DJ, Wasserman J, Petronic-Rosic V, Tsoukas MM. Pityriasis rubra pilaris sensitive to narrow band-ultraviolet B light therapy. J Drugs Dermatol 2009;8:270-3.
4. Kirby B, Watson R. Pityriasis rubra pilaris treated with acitretin and narrow-band ultraviolet B (Re-TL-01) Br J Dermatol 2000;142:376-7.