Case Report

Erythroderma Due to Pityriasis Rubra Pilaris

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Erythroderma can be due to a number of causes. Pityriasis Rubra Pilaris contributing to 1% of all the cases. Pityriasis Rubra Pilaris belongs to a rare erythematous squamous disorders of unknown etiology. We report here a case of Pityriasis Rubra Pilaris that progressed to erythroderma.

Key words: pityriasis rubra pilaris, PRP, HIV human immunodeficiency virus HAART, highly active anti retroviral therapy

Introduction

Erythroderma is generalized erythema of skin affecting over 90% of body surface area. It can be due to several causes. Pityriasis rubra pilaris contributes to one percent of all cases of erythroderma. Pityriasis rubra pilaris is a heterogenous group of disorders that have circumscribed follicular keratosis, brauny skin and orange red erythema. This is a rare disease with an incidence of one in 5000. It has a world wide distribution. It is one of the papulo squamous disorders that runs a chronic relapsing course. We report a case of pityriasis rubra pilaris which progressed into erythroderma. The erythroderma subsided by the use of topical emollients and oral retinoids.

Case Report

A 20 years old man presented with generalized erythema, itching and dryness of skin for last 2 months.

The patient history dates back to 3 years when he started having erythema, dryness and scaling of scalp and face which progressed to involve trunk and limbs. The rash was itchy and the skin was dry.

On examination there was generalized erythema of the body with an orangish tinge. There was mild bilateral pedal oedema. Cardiovascular and respiratory systems were normal. The rest of systemic examination was unremarkable. There was furaceous yellowish orange fine branny scaling on whole of the body more marked on back. The finger and toe nails were shiny.

Nails were thickened and yellowish in colour with splinter hemorrhages and whitish longitudinal bands. There was also subungual hyperkeratosis. Palms and soles were hyperkeratotic named as PRP sandals. Usually PRP is associated with HIV infection but our patient was negative for it.

A skin biopsy taken from arm showed hyperkeratosis, parafollicular parakeratosis, follicular plugging, prominent granular layer (Figure 4 & 5). Dermis showed mild capillary dilatation and mild lymphohistiocytic infiltrate. Some eosinophils were also noted.

A diagnosis of PRP was made and he was started on oral retinoids to which he responded well. Our patient on follow up visits showed marked improvement.

Discussion

Pityriasis rubra pilaris belongs to a group of rare erythematous squamous disorders of unknown etiology. It is characterized by:

- Pityriasis—prominent scaling
- Rubra—perifollicular redness
- Pilaris—follicular plugging

It may become erythrodermic and is associated with palmo-plantar hyper keratosis. Griffith described five subtypes and a new
sixth one has been described associated with HIV infection.

labelling is increased from an average normal 3% to 27%. The nail growth is increased as well but not as fast as in psoriasis

Type - I Classical Adult Form

The condition is severly itchy starts from head spreads towards distal parts of body. It has powdery scaling and pilosebaceous follicles are plugged with keratin.

They are surrounded by erythema, the overall colour is orangish yellow.

The nails show longitudinal ridges, subungual hyperkeratosis and splinter hemorrhages.

It may progress to erythroderma All these findings were present in our patient.

There is spontaneous remission with in three years without recurrence.

Type - II Adult Atypical Form

In this form scales are coarser and more lamellated. The legs are particularly involved. Five percent of the patients belong to this type.

Type - III Classical Juvenile Form;

It is not present at birth and appears at the end of first decade. One third of the patients may have family history. It may resemble adult PRP but may follow an acute infection.

It may clear spontaneously within one to two years.
Type IV – Juvenile Onset Circumscribed Type; Circumscribed persistent asymptomatic plaques of follicular plugging are present on elbows and knees. It may involve scalp. It remains localized to knees and elbows, does not progress further.

Type -V – Atypical Juvenile Variant It presents early in life or even at birth. There is erythema, hyper keratosis follicular plugging, palmoplantar keratoderma. It is wide spread and shows little tendency to clear. Some cases are familial.2

TYPE - VI PRP Associated with HIV; PRP like eruption can occur with HIV. It responds to HAART i.e highly active anti retro viral therapy for HIV.

It shows features of classical PRP with filiform keratosis of scalp face and trunk. it is also associated with acne conglobata.2

There are no definite laboratory tests for PRP.4 Skin histopathology shows basket weave hyperkeratosis, prominent follicular plugging, acanthosis with exaggerated follicular shoulders, spotty parakeratosis and mild upper dermal infiltrate.

In our patient, since he was erythrodermic a prominent scab was also noted above epidermis. As epidermal activity is increased so mitotic figures were also seen. The major differentials of the condition were psoriasis, but in an erythrodermic patient like ours other causes like eczema, drug eruption, Mycosis fungoides, and Sezary syndrome must also be kept in mind.

Since it is a disorder of keratinization , it responds well to oral retinoids. Like acitretin and isotertinoin12 can be used. Methotrexate is an alternative and as an adjunct to retinoids. CiclosporinA 10 and azathioprine have also been used. Emollients help in reducing scales. In circumscribed variety topical vitamin D analogues can also be used.

TYPE VI PRP responds to HAART.2 Photo chemotherapy may exacerbate disease but narrow band UVB, PUVA bath and extra corporeal photophoresis have been found to be beneficial.2

Our case presents classical PRP clinically with characteristic findings on skin biopsy. Thus PRP, although rare, should be kept in the list of differential diagnosis in cases of atypical erythrodermic and psoriasisiform disorders.

References
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