Pityriasis rubra pilaris type 6: A case report in an AIDS patient

Abhilasha Williams, Anisha George, Emy Abi Thomas, Jency Maria Koshy[1]
Departments of Dermatology and [1]Medicine, Christian Medical College, Ludhiana, Punjab, India

Address for correspondence:
Dr. Abhilasha Williams, Department of Dermatology, Christian Medical College, Ludhiana - 141 008, Punjab, India.
E-mail: abhilasha.williams@gmail.com

Abstract

A 24-year-old woman, known to be human immunodeficiency virus positive for 6 years, presented with an itchy rash on the body. She had dull erythematous to hyperpigmented scaly plaques over the body, with extensor predominance. Inflammatory papules and nodules were noted on the face. Follicular hyperkeratotic papules were seen on the shins, giving a "nutmeg grater" feel. All her nails were dystrophic. Histopathology was consistent with the clinical diagnosis of pityriasis rubra pilaris. CD4 counts had dropped to 192 cells/μl, so she was started on antiretroviral therapy along with acitretin to which she responded well within 2 months.

Key words: Acitretin, human immunodeficiency virus, pityriasis rubra pilaris

INTRODUCTION

Pityriasis rubra pilaris (PRP) is a papulosquamous disorder which was originally classified by Griffith into types 1-5.[1] With the rise of the human immunodeficiency virus (HIV) pandemic, PRP associated with it was categorized as PRP type 6.[2] It has an atypical presentation; the most characteristic features are acne conglobata and filiform pattern of keratosis on the trunk and face.[3,4] It is more severe, resistant to treatment,[5] relapses and has a poor prognosis.[3]

CASE REPORT

A 24-year-old woman, known to be HIV positive for 6 years, presented with an itchy rash on the body for the same duration. The lesions began on the face and spread in a cephalocaudal manner to involve the limbs and the trunk. Three months before the presentation, she developed pimples on the face along with an exacerbation of the existing skin lesions.

Cutaneous examination revealed inflammatory papules and few nodules on the face on a background of closed and open comedones [Figure 1a]. Many dull erythematous to hyperpigmented scaly plaques were noted over the extremities, trunk, and buttocks, with extensor predominance [Figure 1b-d]. Follicular hyperkeratotic papules were seen on the shins giving a "nutmeg grater" feel [Figure 1b]. Auspitz's sign was negative. Yellow waxy palmoplantar keratoderma was present [Figure 2a and b]. Scalp showed diffuse scaly plaques. All her nails were dystrophic, with nail plate thickening and subungual hyperkeratosis but no pits [Figure 2c]. Mucosae were normal.

On investigation, routine tests were normal. CD4 count was 192 cells/ml. Skin biopsy revealed alternating hyperkeratosis and parakeratosis with irregular moderate acanthosis and papillomatosis. Hair follicles were dilated and filled with keratotic material. The upper and mid-dermis showed a mild perivascular lymphocytic infiltrate [Figure 3a and b].

She was started on antiretroviral therapy (ART) in the form of zidovudine, lamivudine, and nevirapine along

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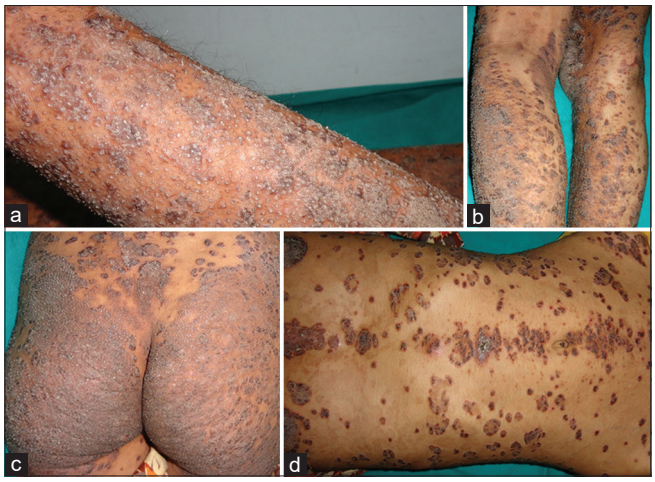

Figure 1: Extensor predominance of hyperkeratotic dull erythematous grouped papules noted over the a) extensor forearm, b) cubital fossa, c) buttocks d) back

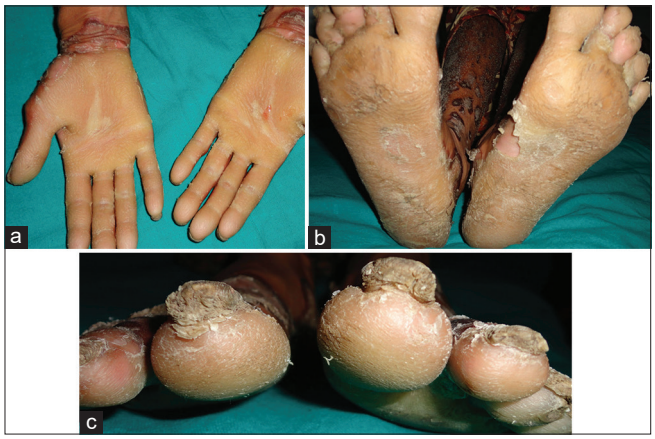

Figure 2: (a and b) Palmo-plantar keratoderma and c) nail dystrophy

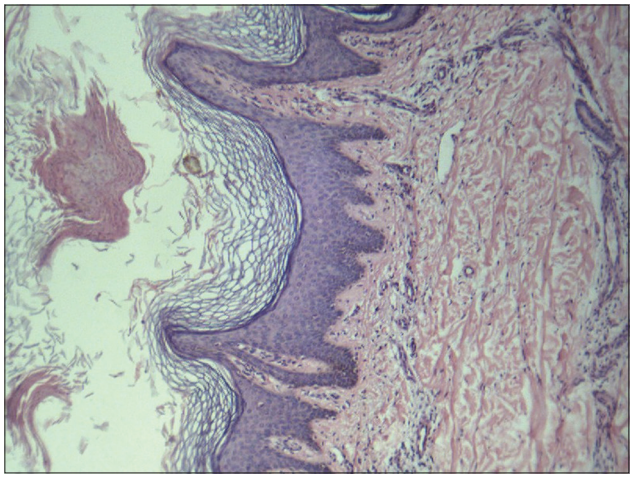

Figure 3: Haematoxylin and eosin stain; original magnification 10×, alternating hyperkeratosis and parakeratosis

with capsule acitretin 25 mg daily. Few weeks later, acne on the face worsened with the appearance of multiple new nodules and cysts. However, ART and acitretin were continued, and complete resolution of the skin lesions was seen after 2 months.

DISCUSSION

PRP type 6 was first reported to be associated with HIV infection by Miralles *et al*. as is well-documented in the literature.[2] However, the description of PRP type 6 and its therapeutic response is mainly based on case reports.

Unlike the skin lesions seen in classical PRP, our patient presented with extensive skin lesions and acne conglobata during the course of her illness. She did not have the other peculiar findings of PRP type 6, namely filiform keratosis. The skin biopsy supplemented the clinical diagnosis of PRP.[6] Our patient developed PRP at the time of diagnosis of HIV. Even though PRP type 6 is said to be recalcitrant to conventional treatment, with the advent of ART, resolution of symptoms and complete regression of the skin lesions has been reported.[7] Our patient's response to ART along with acitretin was excellent with clearing of all the skin lesions within 2 months of starting treatment. No drug interaction between ART and acitretin was noted.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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