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Case Report

Pityriasis Rubra Pilaris: An Uncommon Presentation

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ABSTRACT

Human immunodeficiency virus (HIV) associated pityriasis rubra pilaris (PRP) or PRP Type VI is a distinctive entity reported in HIV patients. We report a patient with skin Type V (based on the Fitzpatrick scale), and HIV infection, presenting with an atypical clinical manifestation (non-acneiform), and with an uncommon distribution of this entity but characteristic histological findings of Type VI PRP. We review the clinical features, pathology and possible aetiology of this entity.

Keywords: Human immunodeficiency virus infection, Pityriasis rubra pilaris, Antiretroviral therapy

INTRODUCTION

Numerous skin conditions have been documented in individuals with human immunodeficiency virus (HIV) infection. The most commonly reported non-infectious skin manifestations include seborrheic dermatitis, psoriasis, pityriasis rosea, Reiter's syndrome, palmoplantar keratoderma, acquired ichthyosis and lichen planus.[1]

However, during the period from 1995 to 1996, a variant of pityriasis rubra pilaris (PRP) emerged in individuals with HIV infection, termed as Type VI PRP. It had a similar clinical presentation as Type I PRP, including diffuse erythematous and scaly plaques with a cephalocaudal evolution, follicular hyperkeratosis and islands of healthy skin. However, it showed a poor response to conventional treatment and was often observed to coexist with acneiform dermatoses, as well as prominent follicular obstruction with spicule formation (reported as lichen spinulosus-like eruption).[2]

We present an atypical case of HIV-associated PRP (Type VI), emphasising its significance in differential diagnosis for HIV patients with Type V skin phototype. This is an attempt to contribute to the available literature in these regards.

CASE REPORT

A 45-year-old woman with skin Type V based on Fitzpatrick scale and HIV infection (clinical stage 2) since 1999, but without treatment 13 months before hospital admission, reported a 2-month history of asymptomatic dermatosis. Cutaneous examination revealed a disseminated, bilateral and symmetric dermatosis, which affected the anterior area of the feet, knees and forearms, characterised by hyperpigmented plaques with a cartographic appearance, with fine

and furfuraceous scale on the surface [Figure 1]. At the time of the assessment, viral load was 88,000 copies/mL and CD4 count was 229 cells/mm^3. In the histopathological study of the skin, we observed psoriasiform dermatitis and hyperkeratosis with horizontally and vertically alternating foci of parakeratosis ("*checkerboard" pattern*) [Figure 2].

Antiretroviral therapy was started with fostemsavir 600 mg bid, darunavir 600 mg bid, ritonavir 100 mg bid, dolutegravir 50 mg bid, maraviroc 600 mg bid and prophylaxis with trimethoprim-sulfamethoxazole (160 mg/800 mg) on Mondays, Wednesdays and Fridays. Clinical dermatological evolution was satisfactory 6 weeks after highly active antiretroviral therapy (HAART) was started.

DISCUSSION

PRP is an inflammatory skin disorder, clinically characterised by the acute appearance of follicular papules and scaly salmon-coloured plaques, alternating with islands of unaffected skin, and it is divided into subtypes.[3,4]

In 1980, Griffiths[5] categorised PRP into five types: Type I: Classic adult; Type II: Atypical adult; Type III: Classic juvenile; Type IV: Circumscribed juvenile and Type V: Atypical juvenile; and in 1996, HIV-associated PRP was added to the classification as a separate type (Type VI).[2]

The dermatosis of PRP Type VI is similar to Type I; the difference lies in the treatment response and the association with other skin diseases, as mentioned above.[6] However, in our case, the lesions appear as hyperpigmented, not salmon coloured as in the typical presentation, and there are no hyperkeratotic papules as we would expect to find. This makes the case different. Nonetheless, during our literature review, we encountered a case report of PRP in a patient with a recent HIV infection diagnosis and similar clinical characteristics to the ones we report here (cartographic-looking hyperpigmented plaques in extensor areas).[6,7] The similarity between cases in terms of clinical appearance, phototype and HIV infection history is noteworthy.

Although the pathogenesis of Type VI PRP is still unknown, given its association with HIV infection and its response to HAART, a pathogenic role of HIV in skin inflammation is hypothesised.[2] According to reported findings, immunosuppression is not a known risk factor for the development of these disorders.[7] That is to say that not only the decline in CD4+ T cell counts[8] but also the shift toward a Th2 cytokine profile,[9] the molecular mimicry and the over-expression of superantigens/xenobiotics,[10] could play a decisive role in the development of Type VI PRP.

A well-described clinical evolution for Type VI PRP is still lacking, and various clinical courses, including spontaneous resolution, chronic management with etretinate and improvement with HAART, such as the one observed in this case, have been documented.

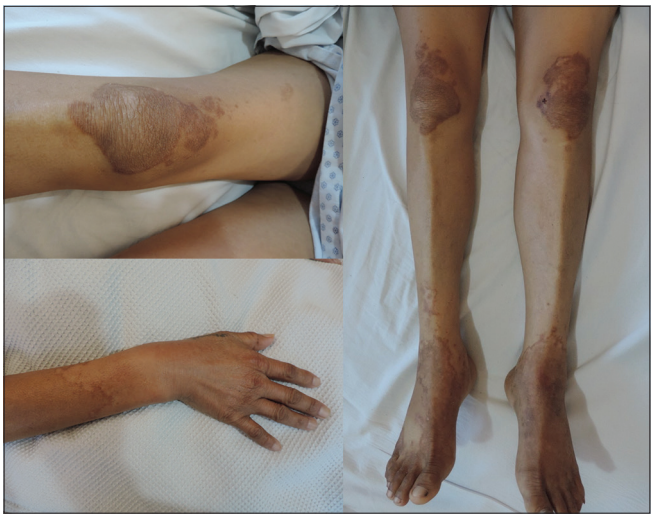

Figure 1: Disseminated, bilateral and symmetric dermatosis, affecting the anterior area of the feet, knees and forearms, characterised by hyperpigmented plaques with a cartographic appearance, with fine and furfuraceous scale on the surface.

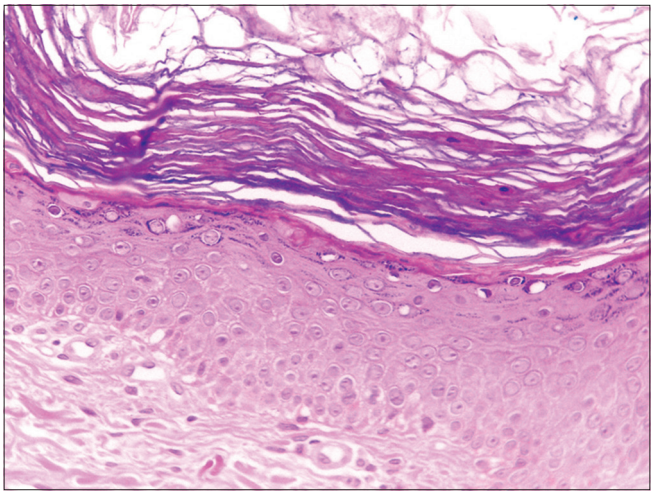

Figure 2: The sections showed a focal psoriasiform dermatitis accompanied by a checkerboard pattern, of alternating parakeratosis with hyperkeratosis with orthokeratosis, in a vertical and horizontal distribution. Some necrotic keratinocytes are present within the superficial epidermis (stratum granulosum). A mild superficial lymphocytic infiltrate is seen. Stain: hematoxylin and eosin ×40.

CONCLUSION

In conclusion, the clinical presentation and course of the disease is variable. However, with an increasing number of case reports sharing similar variables, it will become clearer whether there is an association between patient's, backgrounds and the onset and progression of the disease.

Ethical approval

Institutional Review Board approval is not required.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Conflicts of interest

There are no conflicts of interest.

Use of artificial intelligence (AI)-assisted technology for manuscript preparation

The authors confirm that there was no use of artificial intelligence (AI)-assisted technology for assisting in the writing or editing of the manuscript and no images were manipulated using AI.

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