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HIV-associated "Pityriasis Rubra Pilaris" type 6: First case report from Turkey

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Abstract

A 22-year-old newly diagnosed HIV male, homosexual, presented with rash and thin scaly peeling on bilateral knees and elbows. There were multiple orange-brown scaly plaques on the extensor surfaces of his extremities. Histopathologic examination was consistent with the clinical diagnosis of Pityriasis Rubra Pilaris. CD4+ T lymphocytes were 522/µl and virus load was 19 100 copies/µl. Acitretin and antiretroviral therapy was started and he responded well within 3 months.

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Background

Pityriasis Rubra Pilaris (PRP) is a papulosquamous disease of unknown etiology that progresses from keratoderma to erythroderma on the palms and soles. It is classified into 5 subtypes. The form associated with HIV disease is defined as type 6 [1]. PRP Type 6 is characterized by the presence of HIV infection, usually immunosuppression findings, poor prognosis, poor response to etretinata, and variable association with acne conglobata, hidradenitis suppurativa and lichen spinulosus lesions [2].

Case presentation

A 22-year-old male patient, homosexual, presented to the infectious diseases outpatient clinic with rashes and thin crusted peeling on bilateral knees and elbows. He had no known disease in his medical history. He was not taking any medication. Physical examination revealed multiple orange-brown scaly plaques on the extensor surfaces of the extremities. Palms, soles, and oral mucosa were normal. Nails were regular and there was no lymphadenopathy. The HIV test requested due to homosexual history was positive. Biopsy was performed from lesions with hyperemic appearance on admission and TAF/FTC/BIC treatment was started. There was no serious immunosuppression. Routine tests were normal, CD4+ T lymphocytes were 522/µl, virus load was 19,100 copies/µl, there was no opportunistic infection or malignancy, no prophylactic treatment was recommended. VDRL and TPHA tests were negative. In the 1st month of treatment, the hyperemic appearance in the knees and elbows continued, although decreasing. In the 2nd month of treatment, desquamative follicular papules spreading from the back to the anterior trunk, chest, neck, and scalp, and peeling on the hands and feet bilaterally developed. A punch biopsy taken from the right knee revealed superficial dermatitis with alternating parakeratosis, follicular plug formation, acanthosis, and focal hypogranulosis. A punch biopsy taken from the back revealed acanthosis in the epidermis and mild fibrosis in the dermis and was compatible with "Pityriasis Rubra Pilaris". Dermatology started acitretin 25 mg/day from retinoid drug class.

Discussion

PRP usually presents as a burning, scaling and painful erythrodermic rash consisting of follicular papules that coalesce into plaques. It frequently affects young homo or heterosexual men. The pathogenesis of PRP type 6 may be related to an abnormal immune response to antigenic triggers, namely HIV, and follicular inflammation caused by infection of the hair bulge area by HIV. Vitamin A deficiency, retinol binding protein deficiency and genetic factor may also be the cause [3]. There is no single diagnostic laboratory test or genetic marker for the diagnosis of PRP. As seen in our patient, the diagnosis can be made with clinical suspicion and dermatopathologic correlation. Since nodulocystic folliculitis differentiates this disorder from classical PRP. HIV-related PRP cases have been reported [3-5]. In the reported cases, the clinical onset of PRP was observed to occur shortly after or concurrently with the diagnosis of HIV infection in patients. In this case, the patient was unaware of his HIV diagnosis and had no other known immunosuppressive disease. PRP is one of the diseases that should definitely be considered in HIV-positive individuals and mostly male homosexual or more rarely heterosexual male patients in case of follicular and/or papular rash. The main treatment modality in PRP type 6 is antiretroviral drugs, which can alleviate the symptoms and even completely regress in many patients, as seen in our patient after 3 months, and the importance of effective and concerted antiretroviral therapy comes to the forefront.

Conclusion

With this case report, we aimed to draw attention to this HIV-associated skin rash because PRP type 6 is rare, it is the first case in Turkey, it can be seen independently of immunosuppression and in some cases it responds late to retinoid.

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