Epidermodysplasia Verruciformis acquired in an HIV-positive patient

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Abstract

The disease epidermodysplasia verruciformis is a rare genodermatosis, autosomal recessive in most cases, characterized by susceptibility to infection by types of HPV of the genus β (EV-HPV) that do not occur in immunocompetent clinics. With the increased survival of immunosuppressed patients, especially with AIDS, a form of the disease called epidermodysplasia verruciformis acquired from a clinician similar to the genetic disease has been reduced, but challenging to manage, since an antiretroviral therapy leads to a cutaneous one, making patients more resistant to any treatment modality.

Keywords: Epidermodysplasia verruciformis, immunocompetent, hyperpigmented macular lesions

1. Introduction

Epidermodysplasia verruciformis was described in 1922 by Lewandowsky and Lutz, who thought it was a nevus lesion. It is now known that it is a rare genodermatosis of autosomal recessive inheritance, characterized by susceptibility to infection by types of HPV of the genus β, who do not produce clinical lesions in immunocompetent individuals[1,2, 3].

There are more than 150 HPV serotypes, and recently these have been grouped into five genera, two of which encompass the vast majority of HPV types: α and β HPV. The types of HPV associated with EV correspond to the beta-HPV genus (special emphasis on HPV types 5 and 8) and do not produce lesions in immunocompetent individuals. On the other hand, Alpha-HPV types include agents that cause genital, oral, and skin diseases[4].

This susceptibility to infection by the HPV virus of the genus β is due to the genetic mutation related to EV that consists of the alteration of two genes TMC6 and TMC8, that encode transmembrane proteins that form a complex whose function is to interact with the Zinc transporter 1 and affect the intracellular distribution of zinc. The intracellular level of zinc leads to reduced transcription factors, essential for the replication of HPV of the genus β. Consequently, when this complex is mutated, the transcription factors increase and allow β-HPV to replicate[5].

EV usually starts in childhood, manifesting mainly in exposed areas. The clinical picture is usually insidious, polymorphic, and maybe of the type flat warts, macular pityriasis Versicolor lesions, psoriasiform erythematous lesions, atrophic lenticular lesions, and may present a wide spectrum of shades[5,6].

With the most prolonged survival of immunosuppressed patients, a form of the disease called Epidermodisplasiaverruciforme acquired from clinical behavior similar to genetic disease has been described[2,3,7]. Most of the time, these patients will develop their lesions in adulthood. Most of them are HIV-related immunocompromised, rarer patients with a history of organ transplantation, Hodgkin's disease, SLE, leprosy, and severe atopic dermatitis[8,9,10].

2. Case Report

We report a case of a 41-year-old female HIV positive undergoing irregular treatment, with hyperpigmented macular lesions on the keratosis surface, tending to confluence in the face region, cervical region, and upper trunk with evolution in 2 months (Figure 1). We chose to perform a skin biopsy, which revealed a fragment of skin with an epidermis with irregular acanthosis and blocks of cells with a broad, grayish, and/or bluish cytoplasm with clear vesicular nuclei corresponding to epidermodysplasia verruciforme (Figure 2 and 3). Given the disease's appearance 2 months ago and the patient's important immunosuppressive status (CD4 193), the diagnosis of acquired epidermodysplasia verruciforme was inferred.

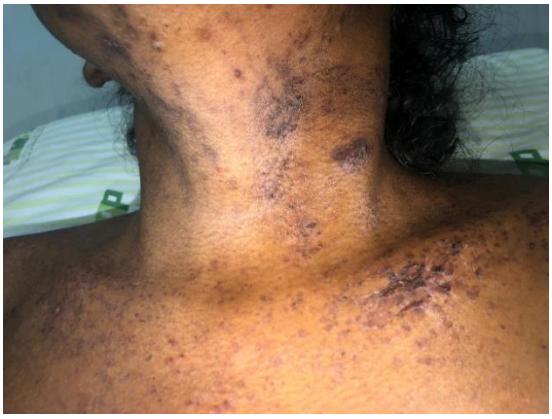

Figure 1 - Macular lesions on the keratosis surface in the cervical region and upper trunk.

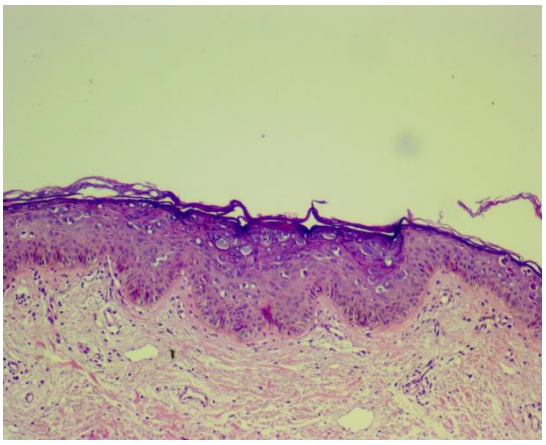

Figure 2 - histopathology of the case. Hematoxylin & Eosin. 100X. Observe epidermis with irregular acanthosis and keratinocyte blocks with broad, grayish cytoplasm.

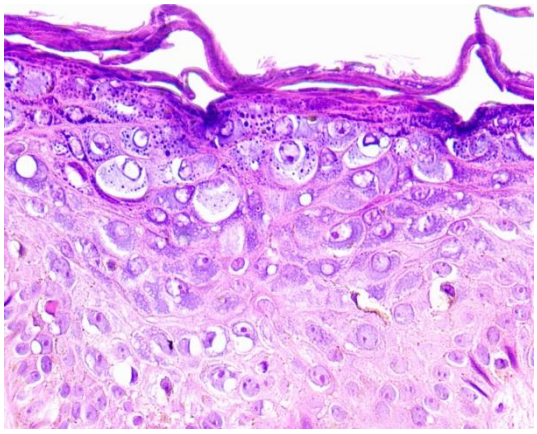

Figure 3 - histopathology. Hematoxylin & Eosin. 400X. Observe the bluish-gray keratinocytes in the epidermis with enlarged cytoplasm, fine granulations, and vesicular nuclei with the evident nucleolus.

Most patients with acquired EV associated with HIV do not have the TMC gene mutation. However, polymorphism of a single nucleotide in the TMC gene has been reported in these patients with a likely modulating role in the disease[11]. An important consideration regarding the treatment of these patients with acquired EV associated with HIV, antiretroviral therapy does not lead to a decrease in skin lesions, which makes patients more resistant to any therapeutic modality[6].

3. References

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