



## Case Report

# Extensive Epidermodysplasia Verruciformis in an HIV Infected Man: A Case Report

Amina Shuaibu<sup>1\*</sup>, Faiza Sadauki<sup>1</sup>, Sara Sani<sup>1</sup>, Abdullahi Muhammad Ahmad<sup>2</sup>

<sup>1</sup>Department of Medicine, Dermatology unit, Aminu Kano Teaching Hospital, Kano

<sup>2</sup>Department of Pathology, Aminu Kano Teaching Hospital, Kano

### Abstract

Epidermodysplasia Verruciformis (EV) is a rare genodermatosis that may be hereditary or acquired, characterised by an abnormal predilection to infection by certain types of Human papilloma virus (HPV). The risk of acquired epidermodysplasia verruciformis increases with immunosuppression, as seen in HIV infected individuals, organ transplant recipients, patients with lymphoma, lupus patients and those on certain biologic therapy. There is an increased susceptibility to non-melanoma skin cancer, especially squamous cell carcinoma. There is no definitive treatment for epidermodysplasia verruciformis, therefore, early diagnosis, skin surveillance for malignancy and adequate sun protection is very pertinent. This is a case report of epidermodysplasia verruciformis in an HIV infected individual.

**Keywords:** Epidermodysplasia verruciformis, Human Immunodeficiency virus, Human Papilloma Virus, Genodermatosis, Nigeria

### INTRODUCTION

Epidermodysplasia Verruciformis (EV), also known as Lutz-Lewandowsky disease, is a rare genodermatosis first described by Lutz and Lewandowsky in 1922.<sup>1,2</sup> EV is characterised by an abnormal susceptibility to certain variants of the Human papilloma virus (HPV), resulting in disseminated skin lesions with a warty appearance and an increased risk of developing non-melanoma skin malignancy.<sup>2</sup> EV can be inherited (genetic) in an autosomal recessive pattern or acquired (sporadic). Acquired immune deficiency is said to play a role in EV due to the link between immunosuppression (HIV, organ transplant, chemotherapy) and acquired phenotypes of EV.<sup>2</sup> We hereby report a rare case of disseminated EV in an HIV-infected man.

### CASE REPORT

This is a report of a 21-year-old man with a recent diagnosis of HIV infection 4 months prior to presentation, and who has been on HAART for 2 months. He presented with asymptomatic generalised skin lesions, which initially started on the sun-exposed parts of his body (face, neck, shoulders and extensor surfaces of the upper limbs), which then disseminated to involve the flexural parts of the body. Lesions started at about 10 years of age. There was no family history of similar lesions. Both parents were deceased.

**\*Correspondence:** Dr Amina Shuaibu, Department of Medicine, Dermatology unit, Aminu Kano Teaching Hospital, Kano, Nigeria.  
Email: [aminashchu86@gmail.com](mailto:aminashchu86@gmail.com); Tel: +2347078198814

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Dermatological examination revealed multiple skin colored to hyper-pigmented confluent lesions. Some of the lesions, especially on the forearm also appear warty (Figs 1, 2, 3). A punch biopsy was done and histopathological examination revealed hyperkeratosis, epidermal hyperplasia with papillomatosis, several swollen, bluish keratinocytes within the granular and spinous layer (Fig 4). Laboratory examinations revealed a viral load of 20 copies/ml, anaemia, raised ESR, elevated liver enzymes AST and ALT, normal electrolytes, normal chest X-ray, and non-reactive HBsAg/HCV-ab.

The patient was counselled about the diagnosis and treatment options were discussed. Treatment with oral retinoid (Isotretinoin) was prescribed but due to the persistent elevation in liver enzymes, it was postponed until liver function improved. Hence, a trial treatment with topical retinoid (0.05% tretinoin) was initiated with no significant improvement on subsequent follow up visits.



Fig 1: Clinical micrograph showing numerous hyperpigmented flat topped papules and plaques, interspaced with few hypopigmented macules seen on the trunk and bilateral upper limbs



Fig 2: Clinical micrograph showing multiple hypopigmented macules and hyperpigmented slightly verrucous papules seen on the trunk posteriorly



Fig 3: Clinical micrograph showing extensive hyperpigmented, lichenified, flat-topped, scaly, verrucous papules and plaques seen on the lower limbs bilaterally

## DISCUSSION

Epidermodysplasia verruciformis (EV) is a rare skin disorder that occurs due to an abnormal susceptibility to infection with certain beta-types of Human papilloma virus.<sup>1</sup> EV is an autosomal recessive skin condition,

although, acquired cases have been reported due to impaired cell mediated immunity in conditions such as HIV, organ transplant, lymphoma, leprosy, and systemic lupus erythematosus.<sup>3</sup> Some reports have linked EV with consanguinity and other factors (genetics, viruses, immunologic and environmental).<sup>4</sup>

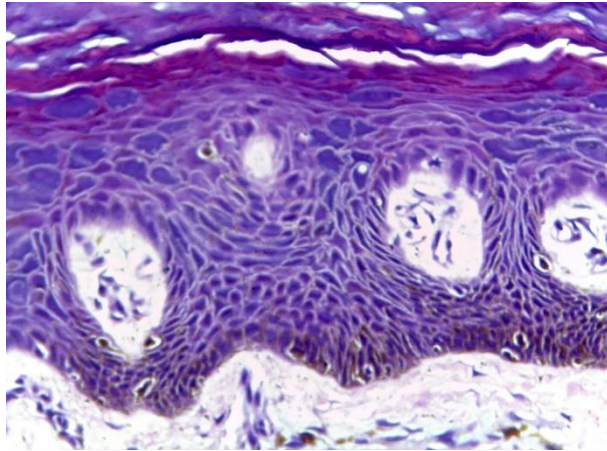


Fig 4: Photomicrograph of the skin biopsy showing hyperkeratosis, with several swollen, bluish keratinocytes within the granular and spinous layer (Haematoxylin and Eosin x400)

HIV-infected individuals have increased prevalence and reactivation of HPV infection in comparison to the immunocompetent persons.<sup>13</sup> In addition, the rate of clearance of HPV is reduced by these patients.<sup>13</sup> EVER 1 and EVER 2 genes located on chromosome 17q25 were first identified in 2002 to be the loci for EV susceptibility.<sup>5</sup> These genes encode transmembrane proteins found within the endoplasmic reticulum that are vital in zinc homeostasis.<sup>5</sup> The mutation in these loci may down regulate cellular immunity and increase the expression of E6 and E7 pro-oncogenic viral genes.<sup>5</sup> The beta-types of HPV isolated globally include HPV-5,-8,-12,-14,-15,-17,-19,-25,-36,-38,-47, and -50.<sup>6</sup> Amongst these HPV-types, HPV 5 and 8 have been found to be more common in HIV-patients.<sup>6</sup> Generally, the data available on EV and HIV is not as that found on HPV associated oral and penile diseases amongst HIV-patients.<sup>7</sup> EV was found to occur in HIV-patients irrespective of their viral load and CD4 count.<sup>3</sup> Additionally, the commencement of HAART did not seem to change the lesions according to some studies.<sup>3</sup> EV has also been considered to be part of the immune reconstitution syndrome (IRIS), as certain reports described appearance of lesions with the initiation of HAART.<sup>8</sup>

The lesions of EV are described as pityriasis-like or flat-wart like macules, papules or plaques found on sun exposed areas (face, neck, chest, upper back and limbs), rarely become generalised, which maybe discrete or confluent, hypo- or hyperpigmented or erythematous. Lesions may involve the anogenital region to a lesser extent, but the mucosa is spared.<sup>9</sup> Clinical features and histology aid in the diagnosis of EV, with polymerase chain reaction (PCR) used in detecting the viral sequence.<sup>10</sup>

Epidermodysplasia verruciformis has no definitive treatment and the HPV is not possible to eradicate. Treatment includes adequate sun protection, cryotherapy, use of pharmacological agents such as retinoids (topical and oral), cidofovir 1%, topical glycolic acid, imiquimod and interferons. Combination therapy has produced prolonged remission in certain patients.<sup>11,12</sup>

## CONCLUSION

Epidermodysplasia verruciformis can be extensive in HIV patients and the risk of malignancy is higher. Hence, the need for early diagnosis, proper skin cancer surveillance, prompt treatment of cases and adequate follow up.

## INFORMED CONSENT

Patient examination and written informed consent were obtained in-line with the principles of the Declaration of Helsinki. The authors certify that all the appropriate consent for the clinical information and images were gotten from the patient to be included in the journal.

## CONFLICT OF INTEREST

There was no conflict of interest

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