

## Epidermodysplasia Verruciformis in A HIV Patient –A Rare Case Report

Doogana Vishali<sup>1</sup>, V. Nivedita Devi<sup>2</sup>

<sup>1</sup>(Department of DVL, Rangaraya Medical College/ Dr. NTR University of Health Sciences, India)

<sup>2</sup>(Department of DVL, Rangaraya Medical College/ Dr. NTR University of Health Sciences, India)

Corresponding Author: Doogana Vishali

**Abstract:** Epidermodysplasia Verruciformis (EDV), is a rare autosomal recessive genodermatosis with cases linked to chromosome X, and attributed to infection by specific types of Human Papilloma Virus (HPV) in immunologically deficient individual. There is an increased prevalence and decreased clearance of HPV infections in HIV infected patients. Malignant change occurs in about 20-30% cases mainly on sun exposed areas. Clinically presents as plane warts, reddish verrucous plaques or pityriasis versicolor like lesions mostly on sun exposed sites. There is no effective treatment. We present an 19 years old married HIV positive female case with asymptomatic hypopigmented macules and flat topped papules on face, neck, trunk and upper extremities since 4months. Punch biopsy of lesion on forearm showed epidermal acanthosis, hypergranulosis, hyperkeratosis, mild papillomatosis, enlarged, and altered keratinocytes with purple-blue cytoplasm suggestive of EDV. After clinico-histopathological correlation, the diagnosis of HIV associated EDV was made. We kept her on sunscreen with SPF 30.

**Keywords :** Epidermodysplasia Verruciformis, Human Papilloma Virus, Pityriasis versicolor

Date of Submission: 28-01-2019

Date of acceptance: 11-02-2019

### I. Introduction

Epidermodysplasia Verruciformis (EDV) was described by Lewandosky and Lutz in 1922<sup>[1]</sup>. It was a rare autosomal recessive genodermatosis with cases linked to chromosome X, attributed to infection by specific types of Human Papilloma Virus (HPV) in immunologically deficient individual<sup>[2]</sup>. HPV strains responsible for this condition were  $\beta$ -strains and include HPV-3, -5, -8, -9, -10, -12, -14, -15, -17, -19-25, -36-38, -47 and -50<sup>[3]</sup>, the most common being 5 & 8 in HIV associated EDV and EDV associated with malignancy. There is an increased prevalence and decreased clearance of HPV infections in HIV infected patients compared with immunocompetent patients<sup>[1]</sup>. Malignant change occurs in about 30-70% cases mainly on sun exposed areas between 20 and 40 years of age. The most common malignancies were Squamous cell carcinoma and Bowen's disease<sup>[4]</sup>.

### II. Case Report

A 19 year old HIV positive married female presented with asymptomatic skin lesions which were started on face since 4 months. She was diagnosed as HIV reactive 7 months back and was on Tenofovir, Lamivudine, and Efavirenz therapy since then. Her baseline CD4 count was 227cells/cu.mm. Her past and family history were nil significant. On physical examination, there were numerous, about 1-3 mm diameter hypopigmented macules and flat topped non scaly papules and few plaques. Koebnerization was seen in few lesions. Sites involved in this case were face, neck, anterior and posterior aspect of trunk and upper extremities (Fig 1a,1b,1c). There was no generalized lymphadenopathy. There were no signs of malignancy. Based on these clinical findings we kept differential diagnosis as Epidermodysplasia Verruciformis, Plane warts, Tinea versicolor. Bed side investigations like KOH mount was done which was negative. A 4mm punch biopsy was done from lesion on forearm. Histopathology showed epidermal acanthosis, hypergranulosis, hyperkeratosis, mild papillomatosis, enlarged, and altered keratinocytes with purple-blue cytoplasm suggestive of EDV (Fig 2). Other investigations like chest X ray, ultrasound abdomen were done to rule out malignancy. After clinico-histopathological correlation, the diagnosis of HIV associated EDV was made. We kept her on sunscreen with SPF 30. Further follow up could not be given as she didn't attend our OPD.

### III. Discussion

EDV may occur in either a classical often hereditary form or in association with various hereditary and acquired immunodeficiencies<sup>[3]</sup>. The classical form commonly presents in infancy or childhood<sup>[5]</sup>. There was loss of function mutations in EVER1 and EVER2 genes located on chromosome 17q25.3. These genes encode

integral transmembrane proteins in the endoplasmic reticulum involved in zinc homeostasis in cells which is recognised as cause of susceptibility to HPV subtypes. It alters zinc levels in the cell, leading to increased activity of transcription factors necessary for HPV replication. Mutations also downregulate cell-mediated immunity by decreasing the ability to present certain HPV antigens to T lymphocytes and disrupts control of the apoptosis/survival balance in keratinocytes<sup>[6]</sup>. There was no gender or racial predilection<sup>[7]</sup>.

It has a polymorphic clinical presentation<sup>[5]</sup>. Clinically the lesions have either the appearance of flat warts or flat scaly red-brown macules that resemble pityriasis versicolor, particularly if they occur on the trunk<sup>[8]</sup>. They occur on sun exposed areas but may be generalised all over the body. They may remain unchanged for many decades<sup>[9]</sup>. At present, there is no known effective treatment for EDV, and no effective preventative strategy for malignant transformation other than perhaps limiting sun exposure<sup>[8]</sup>. Topical 5-fluorouracil, imiquimod, cimetidine, systemic interferon, and oral retinoids are often used as monotherapy or combination treatments<sup>[6]</sup>. Cryotherapy and electrosurgery can be done. Skin grating may be necessary in malignant lesions and graft should be taken from sun protected skin<sup>[8]</sup>. Unfortunately in HIV-associated EDV, control of HIV infection with Highly Active Anti Retroviral Therapy (HAART) may not lead to clinical improvement in the EDV<sup>[6]</sup>. As our patient is in reproductive age group we did not try oral retinoids and just gave sun protection to prevent malignancy.

#### IV. Figures



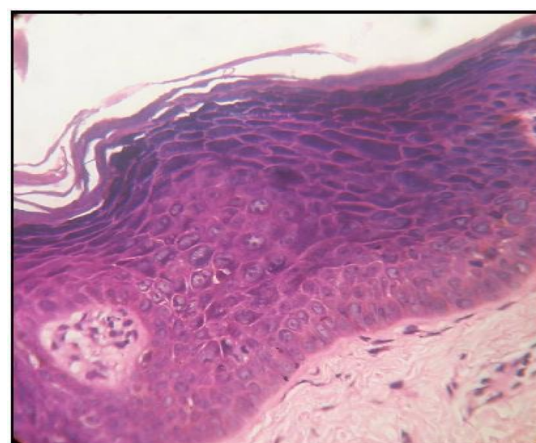
**Figure 1a:** Multiple hypopigmented non- scaly macules on back of the neck



**Figure 1b:** Similar lesions on anterior aspect of neck



**Figure 1c:** Similar lesions on the lower half of face



**Figure 2:** Epidermal acanthosis, hypergranulosis, enlarged and altered keratinocytes with purple blue cytoplasm

#### V. Conclusion

HIV-associated EDV is a relatively rare condition that clinically and histopathologically mirrors the inherited form of EDV. Although the immunologic abnormalities of an HIV infected individual clearly play a large role, there must also be a certain genetic predisposition that increases the chance. Though the risk of malignancy is less in acquired EDV compared to inherited EDV, they must be followed up to check for any development of malignancies and strict sun protection must be advised to them.

### References

- [1]. Kaushal A, Silver S, Kasper K, Severini A, Hamza S, Keynan Y. Epidermodysplasia verruciformis in an HIV-infected man: a case report and review of the literature. *Topics in Antiviral Medicine*. 2012 Dec;20(5):173.
- [2]. Sterling JC. Viral Infections. In Griffiths CEM, Barker J, Bleiker T, Chalmers R, Creamer D, editors. *Rook's Textbook of Dermatology*. New Delhi: Wiley and Blackwell; 2016.
- [3]. Berk DR, Bruckner AL, Lu D. Epidermodysplasia verruciform-like Lesions in an HIV patient. *Dermatology online journal*. 2009 January; 15(1): p. 1-5.
- [4]. Sharma S, Barman KD, Sarkar R, Manjhi M, Garg VK. Efficacy of oral zinc therapy in epidermodysplasia verruciformis with squamous cell carcinoma. *Indian dermatology online journal*. 2014 Jan;5(1):55.
- [5]. Cutlan JE, Rashid RM, Torres-Cabala C, Tyring SK, Thomas V. Epidermodysplasia verruciformis after cutaneous T-cell lymphoma: Periungual presentation. *Dermatology online journal*. 2010 Aug 1;16(8).
- [6]. Champagne C, Moore L, Reule R, Dyer JA, Rady P, Tyring SK, North JP. Cornoid lamella-like structures in HIV-associated epidermodysplasia verruciformis: a unique histopathologic finding. *The American Journal of Dermatopathology*. 2015 Dec;37(12):929
- [7]. Sá NB, Guerini MB, Barbato MT, Giunta GD, Nunes DH. Epidermodysplasia verruciformis: clinical presentation with varied forms of lesions. *Anais brasileiros de dermatologia*. 2011 Aug;86(4):57-60.
- [8]. Seetharam KA. Viral infections. In Sacchidanand S, Oberai C, Inamadar AC, editors. *IADVL Textbook of Dermatology*. 4th ed. Mumbai: Bhalani Publishing House; 2015.
- [9]. Androphy EJ, Kirnbauer R. Human Papilloma Virus Infections. In Goldsmith LA, Katz SI, Gilchrest BA, Paller AS, Leffell DJ, Wolff K, editors. *Fitzpatrick's Dermatology in General Medicine*. 8th ed. New Delhi: Mc Graw Hill; 2012.

Doogana Vishali. "Epidermodysplasia Verruciformis in A HIV Patient –A Rare Case Report." *IOSR Journal of Dental and Medical Sciences (IOSR-JDMS)*, vol. 18, no. 2, 2019, pp 05-07.