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Case Report

Skin lesions in epidermodysplasia verruciformis and clinically mimicking many dermatological conditions

Rajani Chandika*, P. Sirisha, K. V. T. Gopal, P. V. Krishnam Raju

Department of Dermatology, Maharajah's Institute of Medical Sciences, Vizianagaram, Andhra Pradesh, India

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***Correspondence:**
Dr. Rajani Chandika,
E-mail: dr.rajanisekhar@gmail.com

ABSTRACT

Epidermodysplasia verruciformis (EDV) is a rare genodermatosis characterized by increased susceptibility to cutaneous human papillomavirus (HPV) infection, particularly HPV 5 and 8, with a risk of malignant transformation. A 35-year-old female presented with multiple hypopigmented papules and macules over the face and upper extremities, resembling pityriasis versicolor and plane warts. Various investigations, including hemogram, serum biochemistry, viral markers and histopathological examination were conducted. Based on history and thorough examination, the case was diagnosed as EDV. The patient was treated with isotretinoin, emollients, and sunscreen, resulting in moderate improvement. Early diagnosis, sun protection, and regular dermatological follow-up are essential to prevent malignant transformation.

Keywords: Epidermodysplasia verruciformis, Human papilloma virus, EDV, Warts, Malignancy

INTRODUCTION

Epidermodysplasia verruciformis (EDV) is a rare genodermatosis characterized by a unique susceptibility to cutaneous infection by human papilloma viruses (HPVs). The defect is usually inherited as an autosomal recessive trait and presents clinically with plane warts, pityriasis versicolor-like lesions, and reddish verrucous plaques. Dysplastic and malignant changes may occur in the form of actinic keratoses, Bowen's disease, squamous cell carcinoma (SCC), basal cell carcinoma, or rarely sweat apparatus carcinoma.[1]

Patients with this genodermatosis exhibit an abnormal susceptibility of the skin to HPV particularly oncogenic genotypes HPV 5 and HPV 8, as well as HPV 14, 17, 20 and 36 and other β-HPV types.[2] Here in a case of EDV in a middle-aged female who presented with multiple hypopigmented lesions over face and both upper extremities which clinically resembled pityriasis versicolor and plane warts.

CASE REPORT

A 35-year-old female presented with extensive hypopigmented macules over upper extremities and face of 2 years duration. Initially whitish papules developed over right forearm (Figure 1); over next few weeks lesions progressed to adjoining areas and similar lesions developed over left forearm (Figure 2) and over face (Figure 3). There was no history of exposure to excessive sunlight, no history of fever, no history of itching or scaling and no history of other illnesses. No relevant past history and family history non-contributory. She was an agricultural laborer by occupation and had no addictions. On dermatological examination, multiple flat hypopigmented papules coalescing to form plaques were seen over upper extremities. Multiple hypopigmented macules coalescing to form patches were seen over face. Koebner's phenomenon (Figure 4) was positive.

Based on the clinical picture, differentials considered were diffuse hypopigmented keratosis, EDV, pityriasis versicolor, and verruca plana. Complete hemogram and

serum biochemistry were normal, viral screening for HBsAg, HCV, HIV were found nonreactive VDRL was negative and imaging studies including chest X ray and ultrasound abdomen were normal. Histopathological examination showed the following changes in epidermis-prominent keratohyaline granules in the stratum granulosum, acanthosis with large swollen keratinocytes in stratum spinosum along with few koilocytes (Figure 5) which was suggestive of EDV. The patient was treated with Isotretinoin 20 mg daily along with Emollients and broad-spectrum sunscreen for 2 months resulting in moderate improvement. The patient was under review.

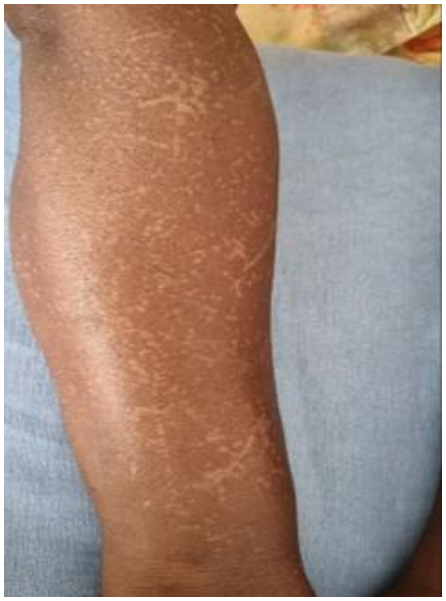

Figure 1: Hypopigmented macules arranged closely over dorsum of right forearm forming linear streaks.

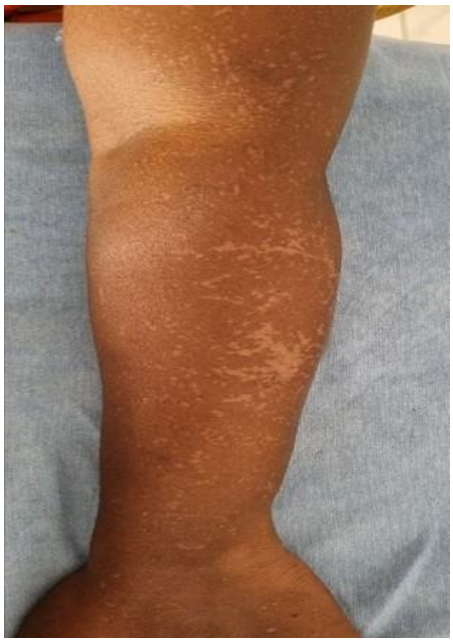

Figure 2: Hypopigmented macules arranged closely over dorsum of left forearm forming linear streaks.

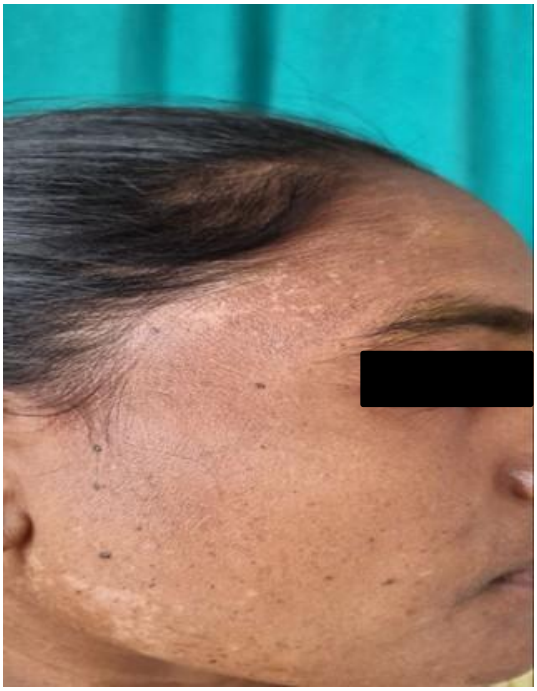

Figure 3: Hypopigmented macules coalescing to form patch over right mandible region and temple.

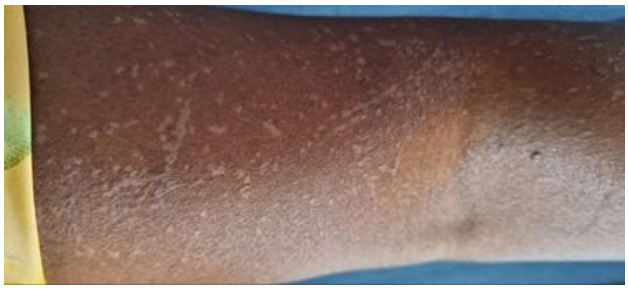

Figure 4: Koebner's phenomenon over right upper extremity.

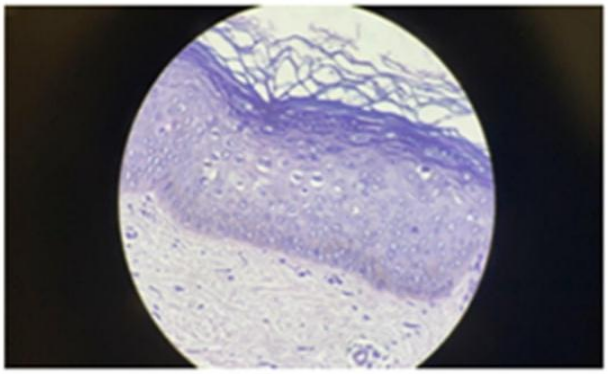

Figure 5: Thick epidermis with prominent keratohyaline granules and koilocytes.

DISCUSSION

EDV, also known as Lutz Lewandowsky syndrome or 'tree man disease' is a rare geno-dermatosis that

predisposes individuals to chronic infections by cutaneous HPV and an increased risk of skin cancer.[3] It follows an autosomal recessive inheritance pattern, with mutations affecting both innate and adaptive immunity. A new classification of EV has recently been proposed, distinguishing between three forms; classical genetic form (linked to EVER1 and EVER2 mutations), non-classical genetic form (associated with X-chromosome mutations), acquired form (observed in HIV-positive patients and organ transplant recipients).[4] In present case with no family history of EDV, which demonstrates the sporadic occurrence which is infrequently reported.

EDV usually begins in infancy or early childhood but may appear at any age. The cutaneous lesions are mainly flat warts, reddish-brown scaly macules resembling tinea versicolor or pityriasis rosea. The lesions mainly occur on the sun-exposed area but may be generalized all over the body. Oral mucosa is spared. EDV lesions are remarkably persistent and may remain unchanged for decades.[5] Malignant change occurs in about 20-30% of cases mainly on the sun-exposed areas, but metastasis is uncommon. The risk of neoplasia is more with HPV 5 and 8. Dysplastic and malignant changes occur most often on UV-exposed sites, commonly as actinic keratoses and Bowens disease suggesting that UV radiation is an important factor. SCC may develop on UV-exposed sites, particularly the forehead, in up to 60% of patients from the third decade onwards.[6] In the present case no clinical features suggestive of premalignant changes were observed.

Other skin diseases which mimic EDV are acrokeratosis verruciformis and lichen planus. In acrokeratosis verruciformis, flat warty papules occur on the back of hands and feet, and on knees and elbows. the palms are diffusely thickened and show punctiform breaks whereas in lichen planus the papules, which are usually pruritic, are pink or lilac in colour and distinctive mucosal lesions are often present. In the present case multiple hypopigmented macules were seen over face and bilateral upper extremities where final diagnosis is achieved with clinicopathological correlation.

Identification of beta-HPVs by PCR or *in situ* hybridisation within lesions can indicate the possibility of EV and this will be strengthened by detection of multiple types in both benign and, if present, malignant lesions. The characteristic histopathology findings for EDV are usually diagnostic. The histological picture is similar in the different clinical lesions. Characteristically, hyperkeratosis and acanthosis are associated with keratinocytes displaying hyperchromatic nuclei, coarse keratohyalin granules and vacuolation or ballooning with a blue-grey pallor in the perinuclear area of keratinocytes and may affect the upper half to three-quarters of the spinous layer.[7] In the present case histopathology showed prominent keratohyaline granules in the stratum granulosum, acanthosis with large swollen keratinocytes in stratum spinosum along with few koilocytes which was suggestive of EDV. Acrokeratosis verruciformis was ruled out because of presence of vacuolation and pityriasis versicolor by absence of hyphae in stratum corneum and pigment deposition.

Treatment of EDV is challenging. If a diagnosis of EDV is suspected or confirmed, avoidance of excessive sun exposure and UV-protective measures should be instigated. Monitoring for cutaneous malignancy is advisable. First line treatments include Imiquimod**,** photodynamic therapy, and cryotherapy. Second line treatments include acitretin and isotretinoin. Electrosurgery and cryotherapy are used in the treatment of benign and premalignant skin lesions. Skin grafting may be necessary for malignant lesions and the graft should be taken from the sun protected skin.[8] In the present case patient was treated with Isotretinoin, emollients and broad-spectrum sunscreen resulted in moderate improvement.

CONCLUSION

Clinical awareness of EDV is essential to make an early diagnosis in view of the presence of immunodeficiency in affected individuals and a high risk of development of cutaneous SCC in early adult life. Prompt treatment should be initiated in all diagnosed cases with oral retinoids to prevent malignant transformation. Dermatologists must be diligent in performing regular full body skin examination and educate the patients about the importance of daily sunscreen use and regular follow up. The case highlights sporadic occurrence of EDV in an immunocompetent adult who presented with predominantly hypochromic skin lesions. Patients of EDV should be kept under supervision to rule out malignancies.

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Ethical approval: Case Report

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