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Epidermodysplasia Verruciformis in a Young Female: A Case Report

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Abstract:

Introduction: Epidermodysplasia Verruciformis (EV) is a rare inherited genodermatosis characterized by an abnormal susceptibility to specific types of human papillomavirus infection. The disease manifests as persistent hypopigmented macules, flat wart-like papules, and plaques predominantly over sun-exposed areas, and carries a risk of malignant transformation.

Case Presentation: We report the case of a 21-year-old female presenting with asymptomatic hypopigmented lesions over the face, neck, chest, and upper arms for six years. Cutaneous examination revealed multiple hypopigmented macules and flat wart-like papules predominantly over sun-exposed areas, along with oral hyperkeratosis. Histopathological examination demonstrated epidermal thickening with enlarged keratinocytes exhibiting prominent perinuclear halos and grayish-blue cytoplasm, consistent with EV. The patient was advised to undergo strict photoprotection and oral isotretinoin therapy was initiated.

Conclusion: This case highlights the importance of the early recognition of EV and emphasizes the need for long-term follow-up owing to the potential risk of malignant transformation.

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Introduction

Epidermodysplasia Verruciformis is a rare autosomal recessive genodermatosis characterized by increased susceptibility to infection with specific types of human papillomavirus (HPV), resulting from impaired cell-mediated immunity [1]. Clinically, the disease presents with hypopigmented macules resembling pityriasis versicolor, flat wart-like papules, and seborrheic keratosis–like plaques [1,2].

The lesions typically begin during childhood and predominantly involve sun-exposed areas such as the face, neck, trunk, and extremities. Patients with epidermodysplasia verruciformis have an increased risk of malignant transformation, particularly to squamous cell carcinoma, especially in adulthood [1].

Herein, we report a case of epidermodysplasia verruciformis in a young female with characteristic clinical and histopathological findings.

Case Report

A 21-year-old female born to a non-consanguineous marriage presented with asymptomatic hypopigmented lesions on the face, neck, chest, and upper limbs since the age of 15 years. The lesions initially appeared over the neck, gradually increased in number, and subsequently spread to the face,

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chest, and upper arms. There was no history of similar complaints among the family members.

Dermatological examination revealed multiple hypopigmented macules on the forehead, posterior neck, and both forearms (Fig. 1). In addition, multiple flat wart-like papules were observed on the neck and upper arms (Fig. 2). Oral examination revealed hyperkeratotic plaques on the anterior aspect of the tongue (Fig. 3). Examination of the hair and nails was unremarkable.
A punch biopsy obtained from a hypopigmented macule on the right upper arm revealed epidermal thickening, with enlarged keratinocytes in the upper epidermis. These keratinocytes showed prominent perinuclear halos and grayish-blue cytoplasm. The stratum corneum demonstrated a basket-weave pattern, and some epidermal cells exhibited dysplastic changes (Figs. 4 and 5). These histopathological findings were consistent with those of EV.

Baseline laboratory investigations, including fasting lipid profile and liver function tests, were within normal limits. The patient was advised strict photoprotection and oral isotretinoin was initiated at a dose of 0.5 mg/kg/day. She was counseled regarding the chronic nature of the disease and advised regular dermatological follow-up for the early detection of malignant transformation.
After three months of therapy, partial clinical improvement was observed with a reduction in the number and prominence of the wart-like papules, whereas the hypopigmented macules persisted. No significant adverse effects were reported during treatment. The patient continues to remain under regular dermatological follow-up for monitoring and early detection of malignant transformation.

Discussion

Epidermodysplasia verruciformis (EV) is a rare lifelong genodermatosis characterized by defective cell-mediated immunity against infection with specific types of human papillomavirus, resulting in persistent viral infection and an increased risk of cutaneous malignancy [3,4]. The condition may occur in familial or sporadic forms, with familial cases being more commonly reported. However, in the present case, there was no positive family history of the disease. Acquired forms have also been reported in immunocompromised individuals including organ transplant recipients and patients with human immunodeficiency virus infection.

Mutations in the EVER1 and EVER2 genes (also known as TMC6 and TMC8) located on chromosome 17q25 play a crucial role in the pathogenesis of EV [5]. These genes regulate cellular zinc homeostasis and immune responses in HPV-infected keratinocytes. Mutations in these genes impair antiviral immunity, allowing persistent infection of keratinocytes with β-HPV types [5].

More than 30 HPV genotypes have been associated with EV, of which HPV-5 and HPV-8 are strongly associated with malignant transformation [1]. Approximately 35–50% of patients develop skin cancers between the ages of 40 and 50 years, most commonly Bowen's disease followed by invasive squamous cell carcinoma [1]. Ultraviolet radiation is considered an important cofactor in carcinogenesis in EV patients, possibly through mutations in tumor suppressor genes such as p53 [6,7]. Recent genetic studies have expanded our understanding of EV by identifying novel structural variants in EVER genes associated with familial cases of the disease [8]. Recent reports have described EV complicated by multiple synchronous squamous cell and basal cell carcinomas, highlighting the malignant potential of the disease and emphasizing the importance of long-term surveillance [9].

Clinically, EV lesions initially appear as hypopigmented or erythematous scaly macules resembling pityriasis versicolor and later evolve into flat wart-like papules and plaques involving sun-exposed areas such as the face, neck, trunk, and extremities [6]. Histopathologically, EV lesions are characterized by enlarged keratinocytes in the upper epidermis with abundant pale or grayish-blue cytoplasm and prominent perinuclear halos, representing viral cytopathic changes.
Currently there is no definitive cure for EV and management primarily focuses on prevention of malignant transformation. Preventive strategies include strict photoprotection, regular dermatological surveillance, and early treatment of premalignant lesions. Various therapeutic modalities have been reported including systemic

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retinoids, topical retinoids, imiquimod, interferons, and immunomodulatory therapies [1,2]. Systemic retinoids such as acitretin and isotretinoin are commonly used because of their antiproliferative and antiviral properties. Recent reports have also suggested that biomarkers such as p16 and Ki-67 may aid in identifying early dysplastic changes in EV lesions, thereby facilitating earlier detection of malignant transformation [10].

Overall, EV remains a rare but clinically significant genodermatosis because of its lifelong persistence and risk of malignant transformation. Early recognition, patient education, and regular follow-up are essential for optimal management and early detection of skin cancers.

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Figure Legends

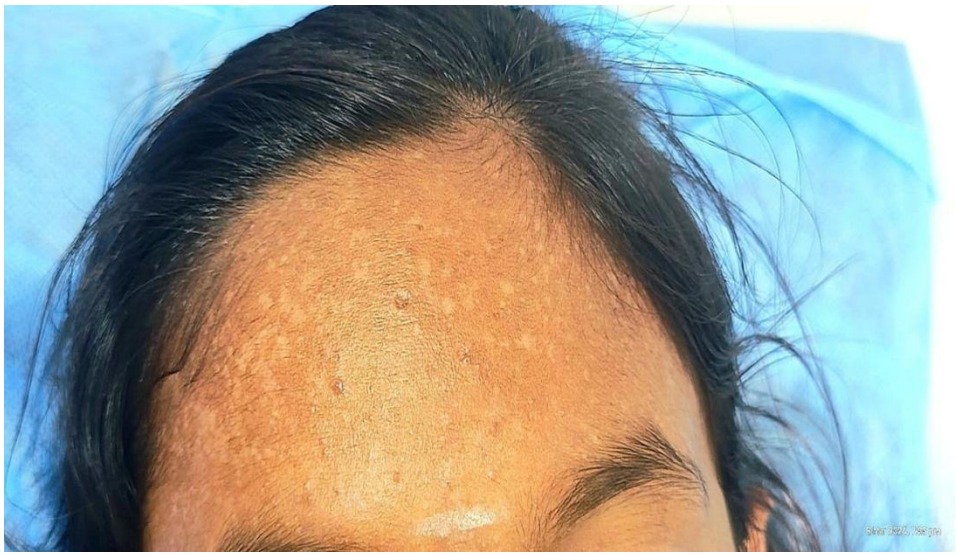

Figure 1: Multiple hypopigmented macules over the forehead resembling pityriasis versicolor.

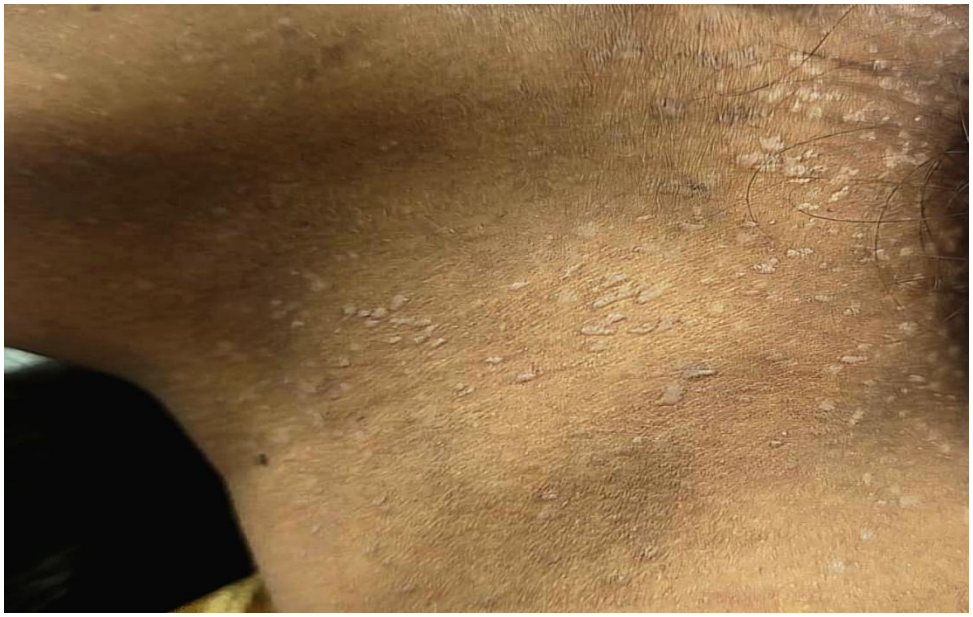

Figure 2: Multiple flat wart-like papules over the neck.

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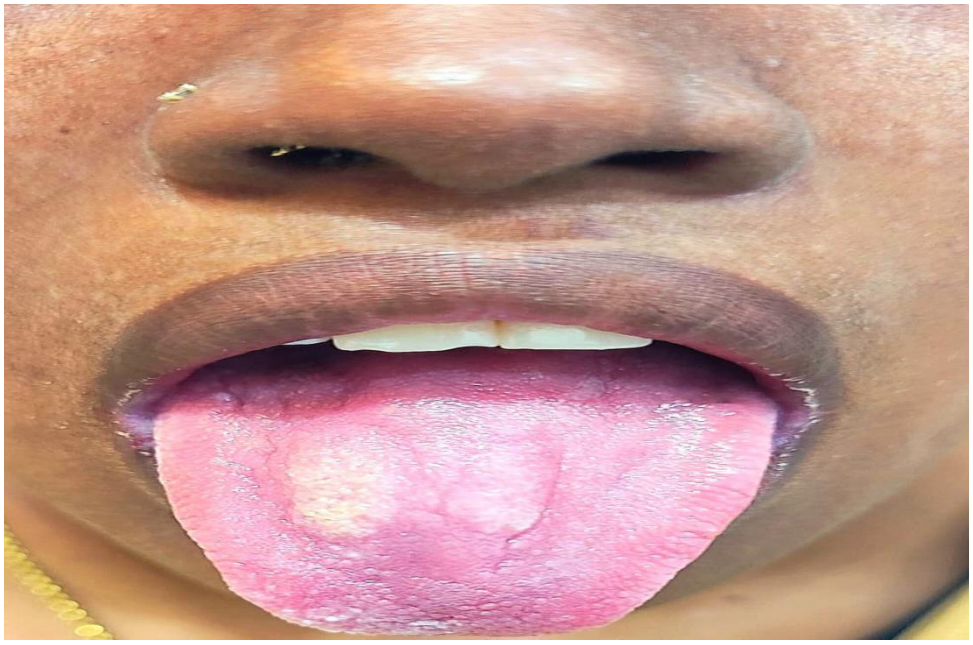

Figure 3: Oral hyperkeratosis on the anterior aspect of the tongue.

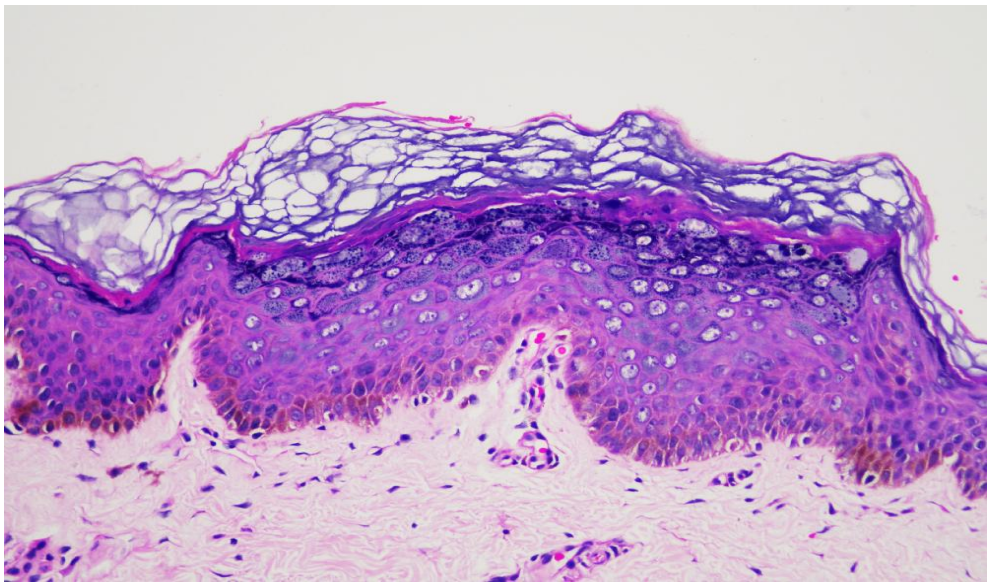

Figure 4: Histopathological section showing enlarged keratinocytes in the upper epidermis with prominent perinuclear halos and grayish-blue cytoplasm (H&E stain).

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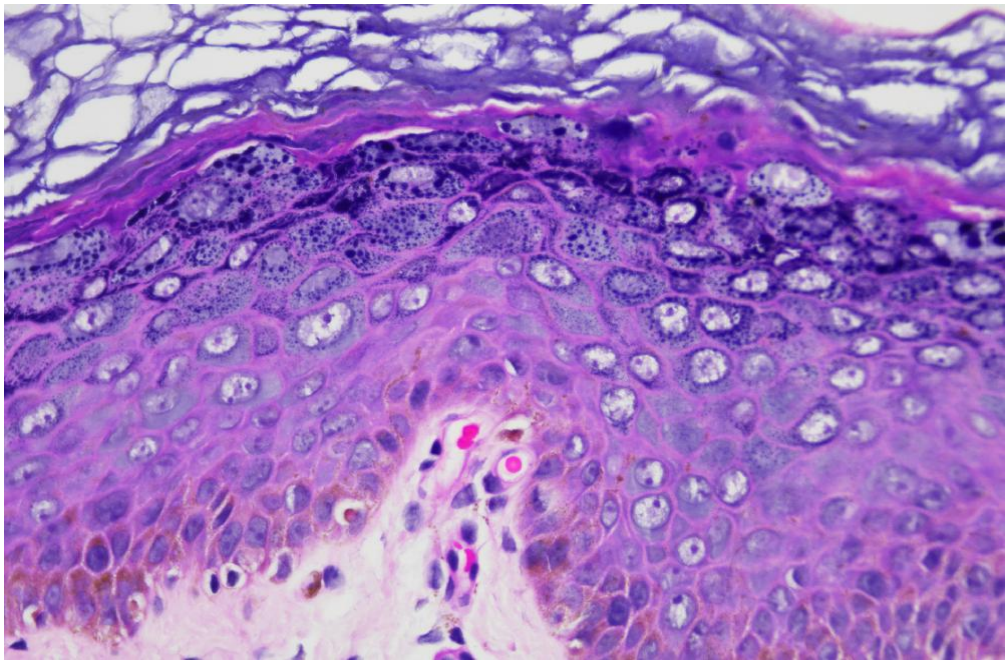

Figure 5: Histopathology demonstrating basket-weave pattern of the stratum corneum with dysplastic epidermal cells.