# Epidermodysplasia Verruciformis

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**Introduction:**

Epidermodysplasia verruciformis (EV) is an inherited disorder in which there is widespread and persistent infection with HPV, giving rise to a characteristic combination of plane warts, pityriasis versicolor-like lesion and reddish plaques. Malignant change is very common but metastasis is rare.

**Case Report**

Seven years old male Saudi patient, referred to Derma Clinic at A.C.H. with one-year history of persistent, slowly progressive, asymptomatic skin lesions on the face, neck, back, chest, shoulders and upper limbs. Patient has three brothers and two sisters. All were normal. Child was product of consanguineous marriage.

General examination, child looks in good health, vital signs were stable, no cyanosis, jaundice or lymphodenopathy, chest, CVS, CNS and abdomen were normal.

Cutaneous examination showed multiple, small, irregularly outlined, slightly scaly yellowish, erythematous, brownish and hypopigmented macules present on the face, neck, back, chest and shoulders (Fig.1, 2). Widespread, slightly elevated, flat smooth papules present on the upper extremities, mainly on the dorsal aspect of forearms and hands (Fig.3).

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Clinical impression was differential diagnosis and conditions to consider include epidermodysplasia verruciformis (EV), EV-like syndrome, Lichen planus, flat warts coexistent with pityriasis versicolor.

Investigation, routine laboratory investigations included Complete blood count, Liver function test, urea and creatinin, electrolytes, chest x-ray, Electro Cardio Gram, abdominal ultrasound. All were within normal limits. Two skin biopsies were taken from the chest and from the right forearm.

Dermatohistopathology showed epidermal changes similar to verruca plana. Epidermis is hyperkeratotic and slightly acanthosis. Vacuolated cells are present in the summer stratum malpighii and granular layer (Fig.4).

Affected keratinocytes are swollen and irregularly chapped and showed Basophilic cytoplasm which contain basophilic keratohyalingranules and some nuclei appear pyknotic, other appear large, round and empty (Figs. 5 & 6).

The final diagnosis was epidermodysplasia Verruciformis (EV). An arrangement was done with the patient for Skin biopsy – for detection of viral particles by electron microscopy, and molecular hybridization procedures for characterization of EV HPVs. Hematological investigation for lymphocyte population (T helper & suppressor lymphocytes). But unfortunately patient fails to attend the appointment so far.

Patient was put on systemic treatment of Retinoids (Acitretin 1mg/kg daily), Sunscreens and avoids exposure to sunlight.

**Discussion:**

EV first described in 1922 by Lewandowsky (as genodermatosis with predisposition to malignant transformation)[1]. EV is rare, lifelong, familial recessive gene, though autosomal dominant[2] and probable x-linked dominant patterns[3] have been reported. Impairment of cell-mediated immunity, notably T-helper-cell number and function, is commonly but not invariably found[4]. The increased natural killer (NK) cell activity in EV patient infected with oncogenic HPV type is interpreted as an appropriate response[4].

There is no impairment of DNA repair[5] and no association with HLA-A or B antigens[6]. There are at least 20 HPV types characteristic of EV, including types 5, 8, 9, 12, 14, 15, 17, 19-25, 28, 29, 36, 38, 47 ad 50. In addition, HPV-3 and 10, which cause ordinary plane warts are found in EV[7,8].

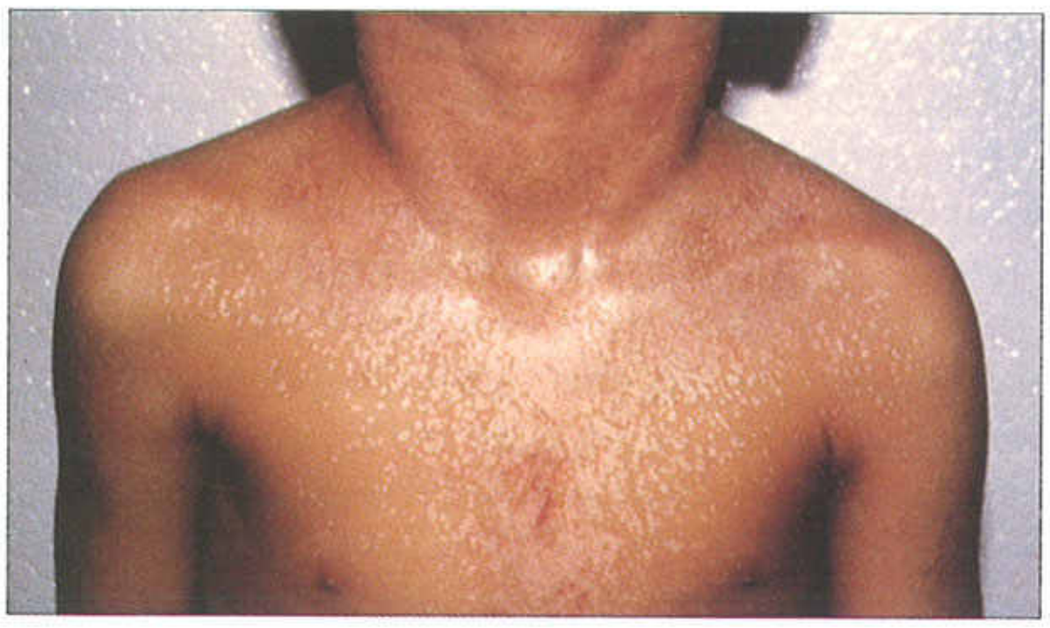

*Fig.1 Multiple, small, slightly scaly yellowish, erythematous, brownish and hypopigment macules present on the Neck ad chest of seven years old male patient.*

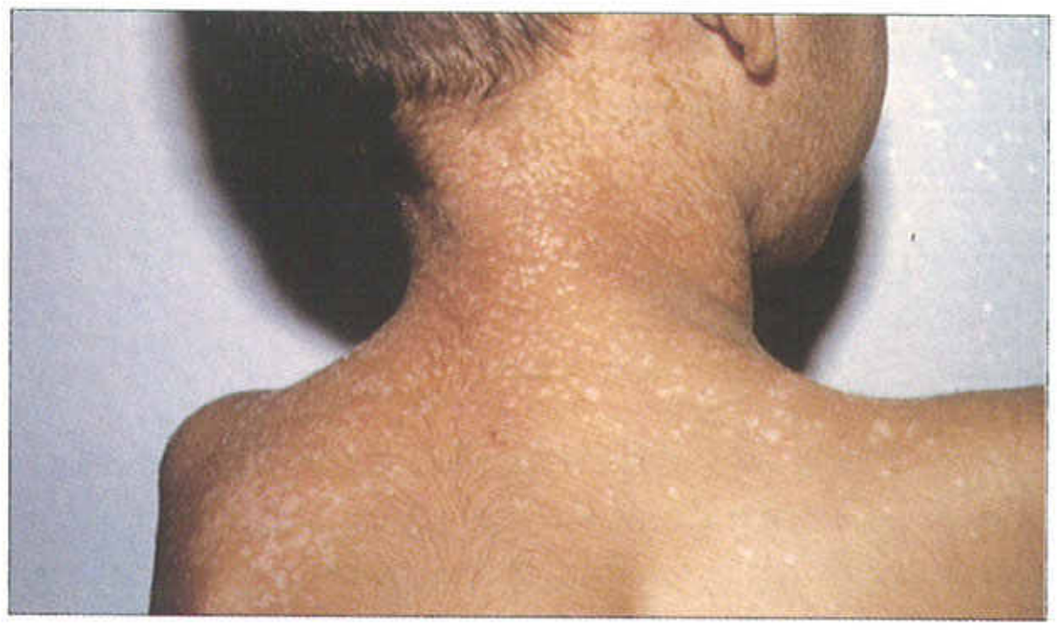

*Fig.2 Same patient of Fig.1, showed skin lesion on the chin, neck, and shoulders.*

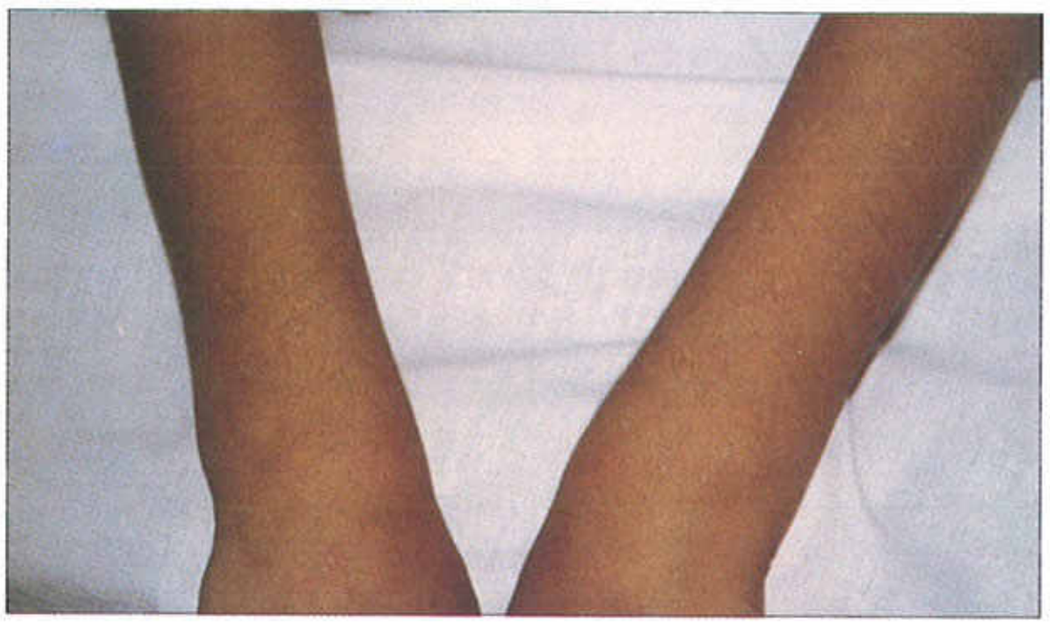

*Fig.3 Widespread, slightly elevated, flat, smooth papules present on the dorsal aspect of forearms and hands.*

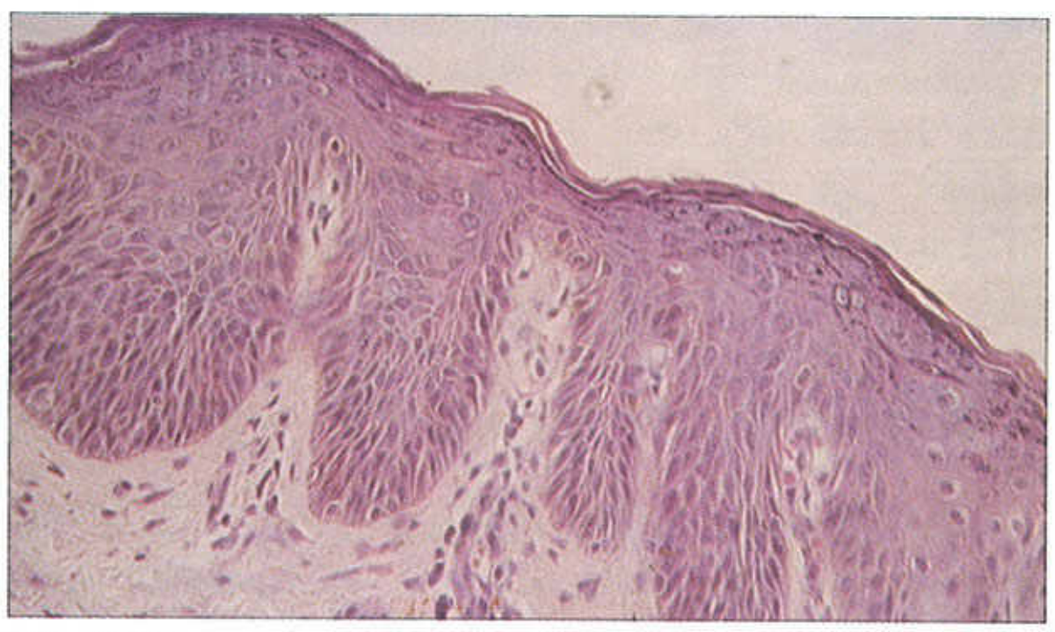

*Fig. 4: Photomicrograph of the lesions shows multiple basaloid cells arranged in solid and adenoid patterns with peripheral palisading (H & E stains x 250).*

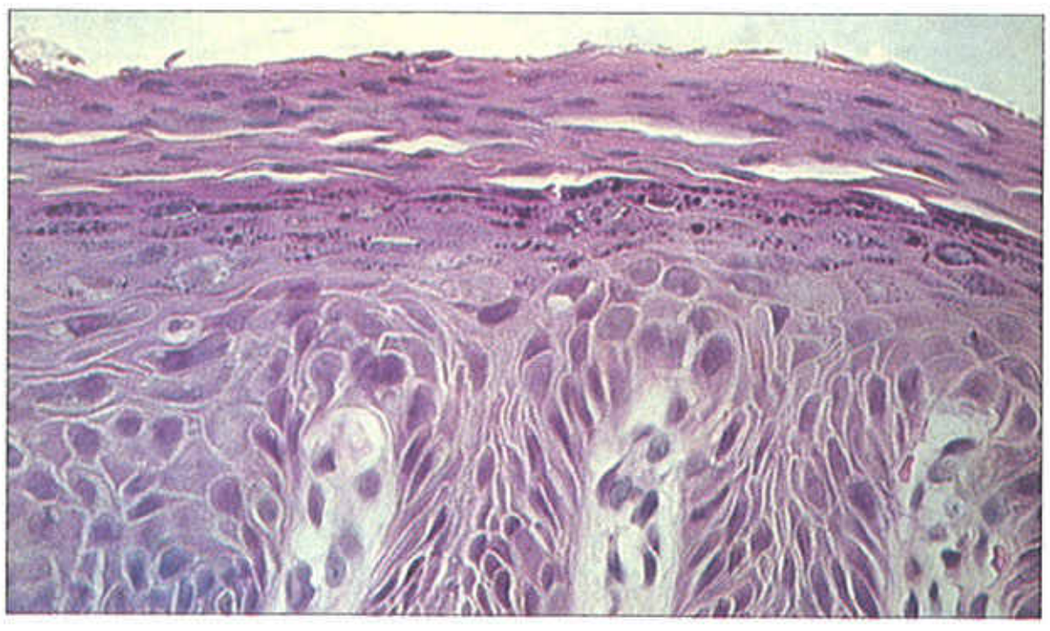

*Fig.5 Affected keratinocytes are swollen and irregularly chapped and showed Basophilic cytoplasm which contain basophilic keratohyalingranules and some nuclei appear pyknotic, other appear large round and empty.*

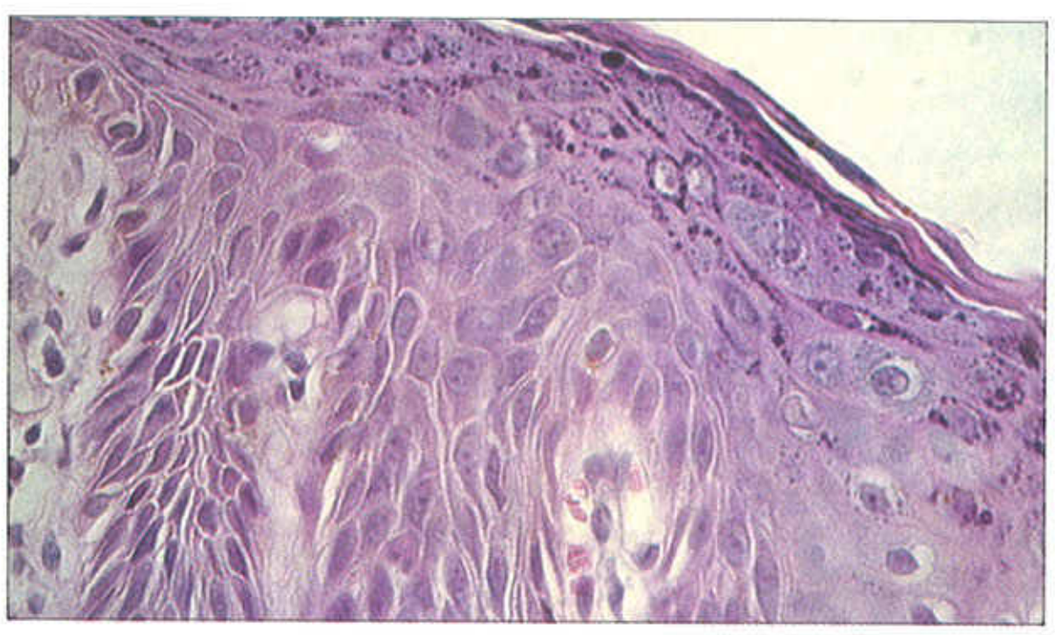

*Fig.6 Same as Fig.5 but in higher magnification.*

Histopathology of skin lesions shows hyperkeratosis and acanthosis. However vacuolation in the keratinocytes is more extensive and may affect the upper half to the three-quarters of the malpighian layer.

The affected keratinocytes are swollen ad irregularly chapped and show, basophilic cytoplasm, which contain basophilic keratohyalingranules, some nuclei appear pyknotic, other appear large, round and empty.

Lesions on the trunk and limbs they tend to be larger, and of two main types. Scaly macular lesions closely resemble pityriasis versicolor, showing depigmentation or varying degrees of brown pigmentation [10]. The warts usually develop rapidly in childhood but may first appear at any age. They are most numerous on the face, neck, backs of the hands and feet, but there are often scattered lesions elsewhere and the warts may be generalized over the body [11].

EV is remarkably persistent and may remain unchanged for decades. Dysplastic and malignant changes occur most often on exposed skin, commonly as actinic keratosis and Bowen's disease, suggesting that ultraviolet is an important factor.

Squamous carcinoma has ultimately developed in one or more lesions in about 20% of reported cases, even before the age of 20 and when the lesions have been present for less than 10 years [12,13,14]. However, metastasis is rare.

The role of etretinate in EV treatment is not clear. Substantial clinical improvement is often achieved at dose of 1mg/kg/day. The effect is dose dependent and relapse occurs if the drug is stopped [15]. Patient should be observed for the development of carcinoma and premalignant lesions. Avoidance of excessive sun exposure, with intelligent use of effective sunscreen, should be advised.

**Conclusion:**

We present a rare case of a Saudi male child with typical Epidermodysplasia verruciformis.

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