

EPIDERMODYPLASIA VERRUCIFORMIS

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A 14-year-old female presented with multiple hypopigmented macules and papules over trunk, face and neck of 10 years duration. She also had verrucous plaques over the extremities. Biopsy of the lesion was consistent with the diagnosis of epidermodysplasia verruciformis. Some of the lesions flattened following placentrex injections. At present, the patient does not have any malignant or premalignant lesion and she is under observation.

Key word : Epidermodysplasia verruciformis

Epidermodysplasia verruciformis (EV) is a rare, lifelong, sporadic or familial, genetic, autosomal recessive disease associated with human papilloma viruses. The disseminated wart-like or pityriasis versicolor-like lesions persist from early childhood and in about one-third of patients, cutaneous carcinoma develops in adult life.

Case Report

A 14-year-old female patient, presented with asymptomatic, generalised raised skin lesions of 10 years duration. It started at the age of 4 as tiny hypopigmented macules over the elbows. The lesions gradually increased in number and size and spread to involve trunk, face, neck and extremities over a period of 5

years. Some of them over the extremities became verrucous. She had received some form of native treatment for 3 months, but without any improvement. For the last 5 years, the lesions had remained the same. On examination, the patient had multiple closely set, non-scaly, hypopigmented macules and papules over the face, neck, trunk and extrmities (Fig.1). Some of them

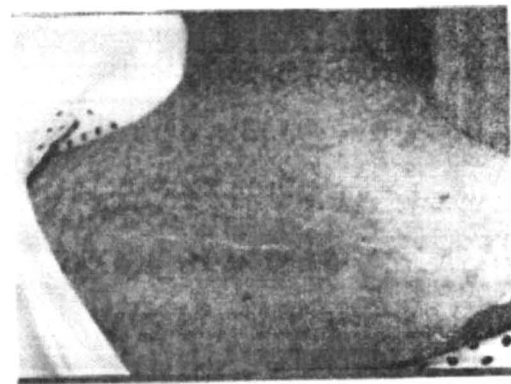


Fig. 1 . Back showing multiple hypopigmented macules and papules, some of which have coalesced.

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had coalesced to form patches. She also had multiple well-defined, irregular verrucous plaques of varying sizes over both upper and

lower extremities (Fig. 2). There were

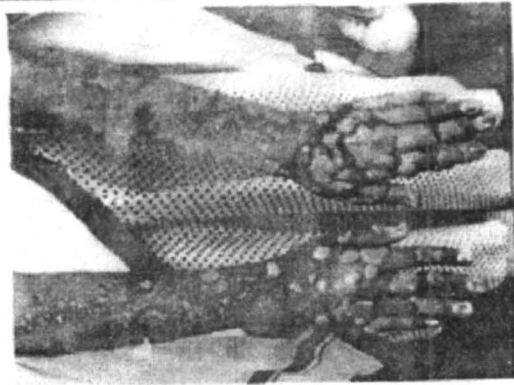


Fig. 2 . Multiple, discrete, irregular, verrucous plaques over both forearms.

periungual verrucae over the hands along with involvement of palms and soles. Mucous membranes were spared. There was no evidence of premalignant or malignant change in any of the lesions.

Routine hematological assessment and skiagrams of chest and extremities were normal. Biopsy taken from edge of a verrucous plaque and tinea versicolor-like macule showed irregular hyperkeratosis, parakeratosis, acanthosis and papillomatosis. There were vacuolated cells extending into the deeper layers of stratum malpighii and prominent keratohyaline granules within some of the cells. The findings were consistent with the diagnosis of EV. Patient was given 20 intramuscular injections of placentrex (1 amp dialy). It was observed that some

of the verrucous plaques flattened. Patient is now under observation.

Discussion

Epidermodysplasia verruciformis was first described by Lewandowsky and Lutz in 1922. It is probably a polygenic multifactorial disease involving genes, infectious agents and environmental factors. The disease is characterised by defect in cell-mediated immunity as inhibition of specific response of T cells and NK cells to EV specific HPV encoded antigens. Dysplastic and malignant changes occur most often on the exposed skin and include actinic keratosis, Bowen's disease, squamous cell carcinoma, basal cell carcinoma and rhabdomyosarcoma.

In our patient, the genetics could not be traced. She rapidly developed warts in childhood as commonly described. Generally, these lesions are resistant to various modalities of immuno modulatory treatment, because CMI is deficient only against the HPV antigens. The partial regression of the lesions following placentrex injection may be a chance occurrence or may be due to immunomodulation. If due to the latter, it provides one more agent which may be useful in the treatment of verruca. Even though at present our patient does not have any dysplastic or neoplastic change, because of their increased occurrence in the patients, it is imperative that she is kept under regular observation.

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