

## Case Report

# Epidermodysplasia Verruciformis (Tree Man Syndrome): A Brief Review with Case Reports

Naga Jyothi A<sup>1</sup>, Pavan Kumar E<sup>1</sup>, Bhavani M<sup>1</sup>, Rudra JT<sup>1</sup>, Rajavardhana T<sup>1</sup>, Reddenna L<sup>1\*</sup>, Mastanaiah J<sup>1</sup>, Chakrapani B<sup>1</sup>, Madhavi Latha C<sup>2</sup> and Sreedhar V<sup>1</sup>

<sup>1</sup>Department of Pharmacy Practice, Balaji College of Pharmacy, Andhra Pradesh, India

<sup>2</sup>Department of Pharmacology, Jagan's College of Pharmacy, Andhra Pradesh, India

## Abstract

Epidermodysplasia verruciformis (tree man syndrome), is a rare autosomal recessive hereditary skin disorder related with a high risk of skin cancer. Uncontrolled infections result in the growth of scaly macules and papules similar to tree bark, mainly on the hands and feet. Patients here with slightly scaly, flat, red-brown macules on the neck, face and body, chronic particularly around the penial area, seborrheic keratosis-like lesions or verruca-like papillomatous lesions, and pinkish-red plane papules on the hands, upper and lower extremities, and face. Clinical diagnostic features are enduring eruptions of flat wart-like papules, pityriasis versicolor-like macules, cutaneous horn-like lesions, and development of cutaneous carcinomas. No curative treatment has been established yet. Here, we present a brief description Epidermodysplasia verruciformis with case reports.

**Keywords:** Epidermodysplasia verruciformis; Tree man syndrome; Uncontrolled infections

## Introduction

Epidermodysplasia verruciformis (tree man syndrome), is a rare autosomal recessive hereditary skin disorder related with a high risk of skin cancer and characterized by abnormal vulnerability to Human Papillomaviruses (HPVs) of the skin and result in the development of scaly macules and papules like to tree bark, primarily on the hands and feet. It has an onset between the ages of one and 20, but it can sporadically be present in middle age. Its cause is by inactivating PH mutation in either the *EVER1* or *EVER2* genes, which are positioned neighboring to one another on chromosome 17. Other genes (ras homolog gene family member H) have also infrequently been allied with this condition [1-3].

Patients here with scaly, red-brown macules on the neck, face and body, particularly around the penial area, seborrheic keratosis-like lesions and pinkish-red plane papules on the hands, upper and lower extremities and face. The preliminary form presents with just flat, wart-like lesions over the body, while the malignant form shows an advanced rate of polymorphic skin lesions and progress of multiple cutaneous tumors. Generally, cutaneous lesions are spread over the body and in some cases limited to one extremity (Figure 1) [4,5].

Clinical features are enduring eruptions of papules, pityriasis versicolor-like macules, and cutaneous horn-like lesions. No curative

treatment has been established yet. Numerous treatments have been recommended, and acitretin 0.5 to 1 mg/day for 6 months' duration is the main valuable treatment owing to antiproliferative and differentiation-inducing effects. Interferon's can be used efficiently along with retinoid. Cimetidine was efficient because of its miserable mitogen-induced lymphocyte proliferation and regulatory T cell activity features [6].

## Case Presentation

### Case 1

A 7 years boy came to the outpatient dermatology department with complaints of asymptomatic lesions over the forehead for 2 years, which were increasing and extend to the forearms and neck. There were no family histories. On examination, multiple hypopigmented maculopapular lesions were present over the forehead. Systemic examination reveals normal. A punch biopsy showed hyperkeratosis and acanthotic epidermi, this indicates EV. The patient was on strict photo-protection, ongoing on oral zinc, topical 0.025% retinoic acid over the lesions, and was advised regular follow-up (Figure 2) [7].

### Case 2

A 14-year-old boy residing in a rural area in India presented to the dermatology clinic with complaints of numerous, non-itchy hypopigmented papules and plaques that were dispersed over the

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**\*Corresponding author:** Reddenna L, Department of Pharmacy Practice, Balaji College of Pharmacy, Rudrampeta, Anantapur, Andhra Pradesh, India-515001, Tel: 9866124220; E-mail: reddennapharmd@gmail.com



**Figure 1:** A patient suffer from Epidermodysplasia verruciformis (tree man syndrome).

forehead, trunk, neck and abdomen. They ranged from 0.5 cm to 2 cm in diameter and initial erupted at the age of 6 months. The lesions slowly progressed in size and number. There were no family histories of EV. Punch skin biopsies were taken from the lesions on the trunk and abdomen (Figure 3) [8].

### Case 3

A 15-year-old boy presented to the dermatology outpatient department with several hypopigmented non-itchy papules ranging in size from 0.2 cm to 0.5 cm over the trunk and both hands that happening 7 years previously. Similar lesions were not here in any family members. The patient had consulted a doctor for his skin ailment. There was no other associated disease. Punch skin biopsy was taken from the hand and trunk (Figure 4) [9].



**Figure 2:** Multiple hypopigmented macules and papules- left side of the face & right side of the face.



**Figure 3:** Nonitchy hypopigmented papules.



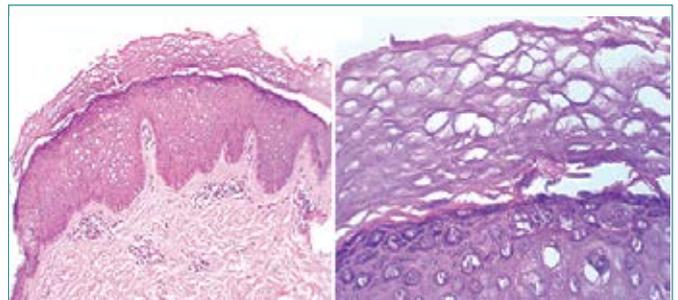
**Figure 4:** Hypopigmented non-itchy papules.

### Case 4

A 42-year-old woman presented with a 2 cm ulcerated plaque over the left clavicular region that had erupted 2 years past. The plaque was flat, but it steadily developed a crust and during the previous month it had started ulcerating. Numerous flat, wart-like lesions changeable in size from 0.5 cm to 1 cm were present on the abdomen, back and upper and lower limbs. Excisional skin biopsy and a punch biopsy were taken from a lesion (Figures 5 and 6) [10].



**Figure 5:** Numerous flat, wart-like lesions.



**Figure 6:** Hyperkeratosis and acanthotic epidermis with the vacuolated cells.

## Discussion

Punch skin biopsies from different sites were performed in four different cases, and all of them showed comparable histopathological (hyperkeratosis, acanthosis, and distended vacuolated keratinocytes) findings. The clinical presentation was distinguishing in all the cases and the histo-pathological features were diagnostic of EV. Most cases of EV are autosomal recessive, cross-linked inheritance and sporadic mutations have also been documented in the literature. The ancestral form of EV is more common, but none of our cases elicited any family history of the disease and hence we presume that they had a sporadic appearance. Mutations of these genes have been found only in EV and are said to be accountable for the development of the disease. Patel et al. have shown a relation between the *EVER2* gene and SCC with HPV5 and HPV8 as causative agents in 90% of cases [11].

## Conclusion

It is a rare autosomal recessive hereditary skin disorder characterized by abnormal vulnerability to Human Papillomaviruses (HPVs) of the skin. Surgical removal and cryotherapy are used in the treatment. Auto transplantation of the skin from simple skin for localized multiple malignant lesions. Education of the patient and expurgation of the tumoral lesions take predilection to avoid the development of cutaneous tumors.

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