

## CASE REPORT

### EPIDERMODYSPLASIA VERRUCIFORMIS WITH SQUAMOUS CELL CARCINOMA OF SCALP: A CASE REPORT

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**ABSTRACT:** Epidermodysplasia verruciformis (EV) is a rare genodermatosis characterized by an unique susceptibility to cutaneous infection by human papilloma viruses (HPVs). Pityriasis versicolor like lesions persists from childhood.

**KEYWORDS:** Human papilloma virus, Genodermatosis, Autosomal recessive, Acitretin.

**INTRODUCTION:** Epidermodysplasia Verruciformis (EV) is a rare genodermatosis first described by Lewandowski and Lutz in 1922. EV patients show an unique susceptibility to cutaneous infection by a group of phylogenetically related human papilloma virus types referred to as EV types. The (HPV) disease usually manifests in childhood with highly polymorphic and numerous lesions. These patients present with flat wart like and pityriasis versicolor like lesions.<sup>[1]</sup> After 30 years or so approximately half of the patients may show a malignant transformation. We hereby report a case of EV with well differentiated squamous cell carcinoma of scalp.

**CASE REPORT:** A 40 years old male born of non consanguinous marriage presented with multiple asymptomatic light and dark coloured lesions over extremities, abdomen, trunk, face and neck since childhood, from 7 years of age. Lesions progressively increased in size and number. Patient developed a large nodule over the scalp 6 months ago which ulcerated. H/O pityriasis versicolor like lesions present in the elder brother.

**ON EXAMINATION:** Single non healing ulcer (4X3 cm) with everted edges, firm in consistency seen over the scalp. Multiple flat topped, scaly, hypopigmented macules seen over the extremities, abdomen, chest, trunk with dorsum of hands resembling pityriasis versicolor. Bilateral preauricular, submandibular lymph nodes were enlarged (Hard, non tender and fixed). Nails and mucous membrane were normal. On investigating complete blood counts, serum biochemistry, urinalysis, HIV serology were normal.

FNAC of (Rt) submandibular lymph node showed cluster of tumor epithelial cells, pleomorphism, hyperchromatic nuclei, high nuclear cytoplasmic ratio.

#### Skin Biopsy of Scaly Lesion from Back Showed:

1. Stratum corneum (Basket weave appearance).
2. Focal hypergranulosis, mild acanthosis.
3. Vacuolization of cells with central nucleus, keratohyaline granules, blue gray cytoplasm (KOILOCYTES).
4. Flattening of rete ridges seen.

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**Scalp Biopsy:** Well differentiated squamous cell carcinoma Patient underwent a wide local excision for scalp (SCC) followed by local flap and split skin graft under general anaesthesia.

For the skin lesions patient was started on topical sunscreens with oral retinoids (Acitretin 25 mg/day). Genetic counseling was done and he was asked to come for regular follow-up in view of surveillance for evolving malignancies, for early diagnosis and management of malignant and premalignant lesions.<sup>[2]</sup>

**DISCUSSION:** Epidermodysplasia verruciformis is a rare genodermatosis characterized by persistent and widespread infection with HPVs. About two dozen EV specific HPV types have been described. A subset of which (mainly 5, 8) is seen in EV associated skin cancers.<sup>[3]</sup> Mode of inheritance is autosomal recessive. The Human keratinocytes are susceptible to HPV in EV due to truncating mutations in (EVER1/TMC 6), (EVER 2/TMC 8) genes on chromosome 17q25.<sup>[4]</sup> Tumours are generally of low metastatic potential and develop mainly in sunexposed areas implicating UV irradiation as an important co- carcinogen.

The diagnosis is usually clinicopathological. To identify HPV types, DNA amplification is done which could not be done in our case due to financial constraints. This case highlights the need for educating the patients with EV about minimizing sunexposure and the importance of screening in family members for similar lesions.<sup>[5]</sup> A regular follow-up of EV patients is mandatory.

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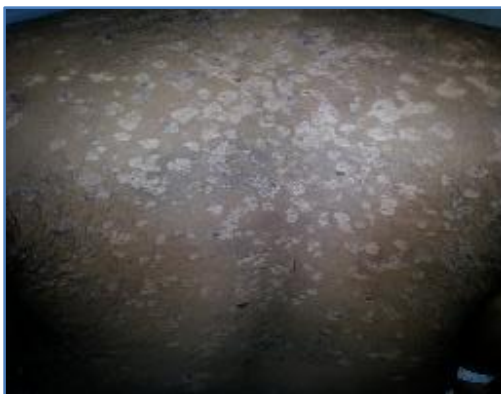
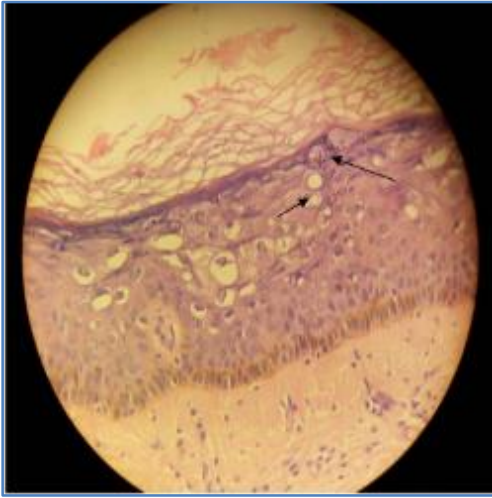


Fig. 1: Back



Fig. 2: Chest and Abdomen

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**Fig. 3: Skin biopsy showing Koilocytes**



**Fig. 4: Ulcer- Scalp**

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