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INTERNATIONAL JOURNAL OF ADVANCED RESEARCH (IJAR)

Article DOI: 10.21474/IJAR01/18751
DOI URL: http://dx.doi.org/10.21474/IJAR01/18751

*RESEARCH ARTICLE*

# AN UNCOMMON COMPLICATION OF EPIDERMODYSPLASIA VERRUCIFORMIS

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## Manuscript Info

***Manuscript History***
Received: 20 March 2024
Final Accepted: 27 April 2024
Published: May 2024

***Key words:-***
Epidermodysplasia Verruciformis, Basal Cell Carcinoma, Dysplasia, Malignancy

## Abstract

Epidermodysplasia verruciformis (EV) is a rare autosomal recessive genetic disorder. which patients have a reduced immunological ability to defend against and eliminate certain types of human papillomavirus (HPV), resulting in persistent infection and an increased lifetime risk of developing cutaneous dysplasia and malignancy. mostly squamous cell carcinoma was seen on sunlight-exposed areas, especially on the extremities, and forehead. Malignant transformation began in the form of actinic keratosis and early micro- invasive squamous cell carcinoma of Bowen's type, rarely basal cell carcinoma. We report the case of a young patient with a history of EV who presented with a basal cell carcinoma that developed over a short period of time. The patient was successfully treated surgically.



## Introduction:-

Epidermodysplasia verruciformis (EV) is a rare autosomal recessive genetic disorder. which patients have a reduced immunological ability to defend against and eliminate certain types of human papillomavirus (HPV), resulting in persistent infection and an increased lifetime risk of developing cutaneous dysplasia and malignancy. EV was first described by Lewandowsky and Lutz in 1922 (1) There are broadly 2 forms of EV, the classic form is the inherited or primary type, which is inherited in an autosomal recessive pattern, whereas a separate acquired or secondary type is a clinically almost indistinguishable condition observed mainly in HIV-infected or immunosuppressed individuals (2).

We report the case of a young patient with a history of EV who presented with a basal cell carcinoma that developed over a short period of time. The patient was successfully treated surgically.

**Cas Report:**

A young man of 30 followed in the service of dermatology for EV, which evolves since the age of 4 years, characterized by the progressive appearance of wart at the level of the hands and the feet, during its regular follow-up, the patient reported the appearance of a lesion at the level of the back increasing gradually in size, bled in contact, Physical examination revealed multiple hyperkeratotic lesions, pinkish red and particularly papillomatous on both extremities hands and back, and multiple macules of pityriasis versicolor like and flat plaques of seborrheic keratosis ( Fig A), on the trunk, and an erythematous plaque, surmounted by an erosion. Dermoscopy evidence of ovoid nests, Spots and globules, chrysalis and telangiectasis( Fig B), a BBC was suspected, a biopsy was performed which returned of a basal cell carcinoma nodular, then an excision was performed with a successful evolution.

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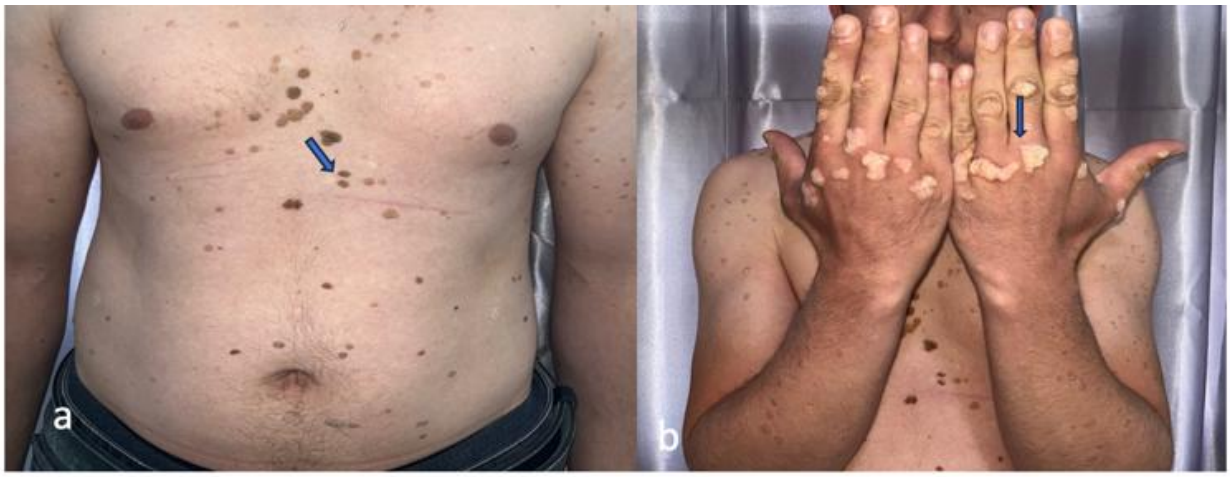


Fig A: multiple macules of pityriasis versicolor like and flat plaques of seborrhoeic keratosis on the trunk (a), Multiple hyperkeratotic, pinkish-red papillomatous lesions in the back of the hands

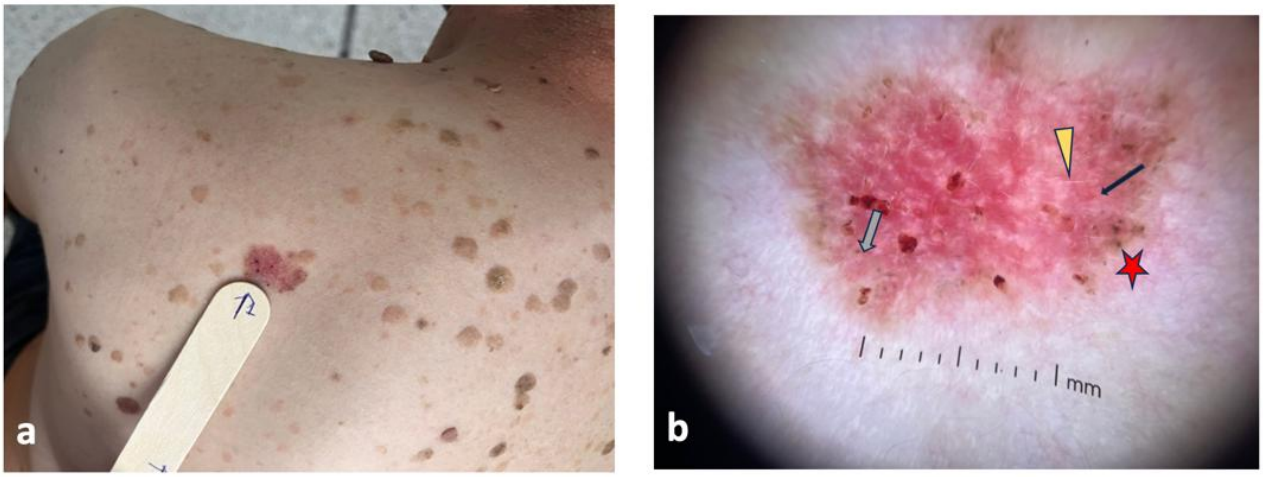


**Fig B:** (a) Clinic : An erythematous plaque, surmounted by an erosion
(b) dermoscopy , Spots and globule , crystalline structures , Spoke-wheel pigmentation , Linear and arborising telangiectasia.

## Discussion:-

Epidermodysplasia verruciformis (EV) is a rare autosomal recessive genetic disorder of the immune system manifested by increased susceptibility to cutaneous human papillomavirus (HPV) infection beginning from the early years of life.

This genodermatosis manifests mainly as verrucous cutaneous lesions such as multiple persistent verrucae, pityriasis versicolor-like lesions, warty cutaneous lesions.

There are broadly 2 forms of EV, the classic form being the inherited or primary type, inherited in an autosomal recessive pattern, while a separate acquired or secondary type is a clinically almost indistinguishable condition that is observed mainly in HIV-infected, immunocompromised, or immunosuppressed individuals.(2) Patients with typical EV develop EV lesions early in childhood and continue to develop new lesions throughout life and This autosomal recessive condition in its primary , is caused by a mutation of TMC6/EVER1 or TMC8/EVER2, which is believed to impart a defect in the ability to mount an immune response to certain HPV types within keratinocytes characterized by the persistence of the HPV virus in skin lesions, with a lifelong increased risk of developing skin dysplasia and malignant tumors. The most identified HPV types found in EV related cancers are HPV5 and HPV 8 (3), Malignant transformation was reported to occur in about 30 to 50% of patients and non- melanoma skin cancers, mostly squamous cell carcinoma was seen on sunlight-exposed areas, especially on the extremities, and forehead. Malignant transformation began in the form of actinic keratosis and early micro- invasive squamous cell carcinoma of Bowen's type, rarely basal cell carcinoma (4)

Histopathological examination of EV lesions shares many features with verruca-type lesions. Hyperkeratosis and parakeratosis, acanthosis with koilocytic cellular atypia and pale eosinophilic cytoplasm are characteristic of EV-type lesions (5). The histological finding of "blue cells" is pathognomonic for HPV-infected keratinocytes in EV, and the "blue" appearance of these cells can be used as an indication of EV-related disease. This "blue" appearance refers to cells with a pale blue cytoplasm with the presence of abundant basophilic keratohyalin granules.

## Conclusion:-

Epidermodysplasia verruciformis (EV), is a rare genetic disease, the patient has a high risk of developing malignant tumors including squamous cell carcinoma, Bowen's, and rarely basal cell carcinoma, the particularity of our case is that the patient has developed a basal cell carcinoma, through our observation we insist on a careful examination and dermoscopy is recommended at each consultation in patients being followed for EV.

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