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Epidermodysplasia Verruciformis: A Case Report

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Abstract
Epidermodysplasia verruciformis(EV) is a rare, hereditary, genodermatoses characterized by susceptibility to certain types of human papillomavirus(HPV) infections and a heightened risk of malignant skin tumors. We present a case of 25-year-old male from South India exhibiting multiple asymptomatic wart-like lesions on sun-exposed areas with no family history of EV. Clinical examination and histopathological analysis confirmed the diagnosis. Treatment involved oral isotretinoin and photoprotection. The discussion highlights the clinical manifestations, genetic factors and management options, emphasizing the importance of sun protection and regular follow-up to prevent malignant transformation.

Keywords: *Epidermodysplasia verruciformis, Human Papilloma virus, sun-exposed areas, pityriasis versicolor, verruca-like lesions.*

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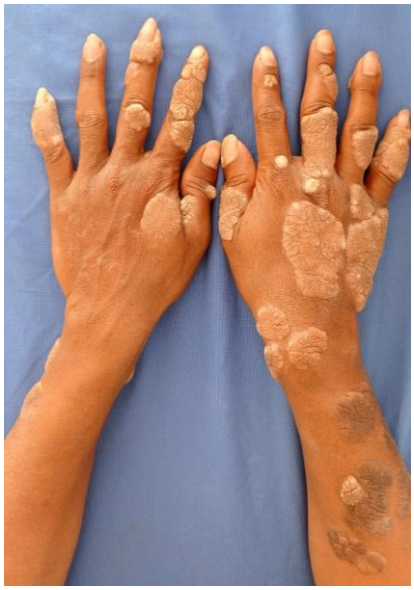
I. Introduction

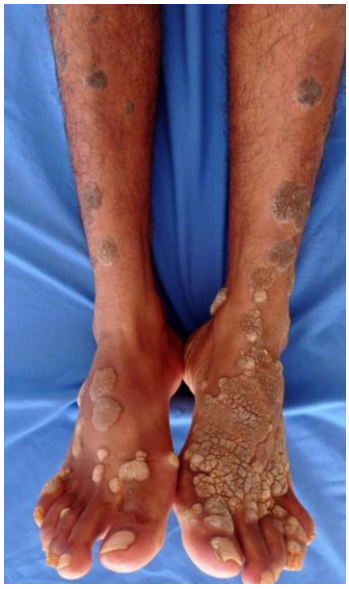
Epidermodysplasia Verruciformis (EV) is a rare, lifelong hereditary autosomal recessive genodermatoses. It is characterized by unusual susceptibility to infection with Human Papilloma virus (HPV) of certain types and with an increased tendency to develop malignant skin tumors. Lewandowski and Lutz first described the disease in 1922. Clinically the disease is characterized by macular and raised flat lesions like pityriasis versicolor and seborrheic keratosis like plaques that begin in childhood. The lesions usually occurs in infancy and continue appearing throughout life mainly on the sun exposed areas of face, neck, trunk and extremities. Lesions on sun exposed sites are more prone for increased risk of malignant transformation. Prevalence of malignancy is very high in immunocompromised patients. Common malignancies are Bowen's type carcinoma in situ or invasive squamous cell carcinoma.[1,2] We report a Epidermodysplasia Verruciformis (EV) case and briefly document it.

II. Case Synopsis

A 25 year old male from South India born to a non-consanguineous marriage presented to the dermatology OPD complaining of asymptomatic wart like lesions since 3 years. The lesions first appeared on dorsal aspect of hands gradually increased in number and spread to the legs, dorsum of feet, lower back and abdomen areas. Patient had visited Ayurvedic specialist and was prescribed some tablets and topical medications. He is otherwise healthy with no other medical illnesses. Physical examination revealed multiple verrucous firm flat growths of varying sizes ranging from 1x1 cm to 10x20 cm on the dorsum of hands, anterior aspect of legs, dorsum of feet (Figure 1a & 1b). No history of similar lesions among the family members. Histopathology showed marked hyperkeratosis with basket weave appearance and koilocytes in epidemis (Figure 2a & 2b) proving the diagnosis. Patient was advised to follow strict photoprotection and started on oral Isotretinoin 20mg OD. He was advised to have regular follow- up in the OPD.



Firgure (1a) Multiple flat verrucous growths on dorsum of hands.



Firgure (1b) Multiple flat verrucous growths on dorsum of legs and feet.

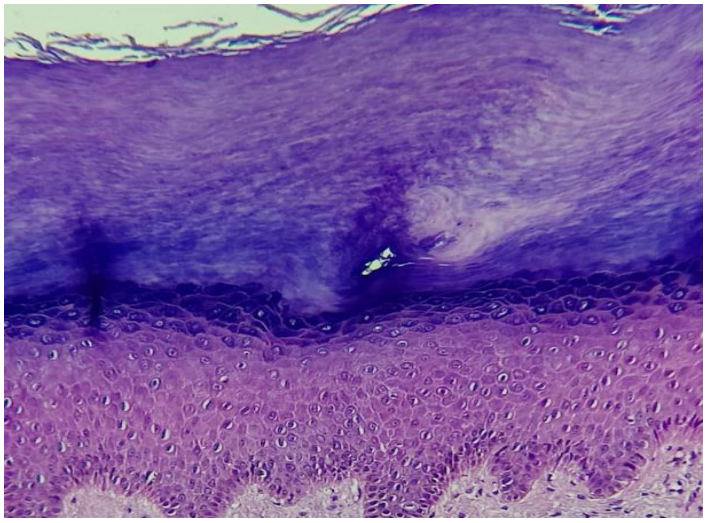


Figure (2a) Marked hyperkeratosis with basket weave appearnace of epidermis.

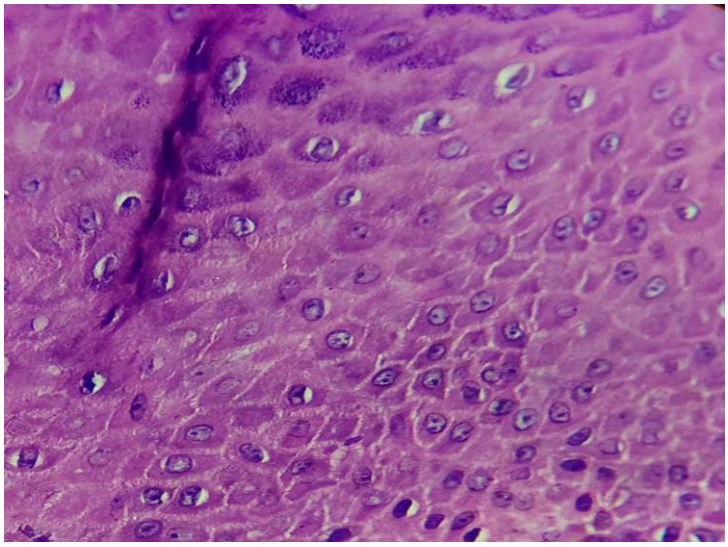


Figure (2b) Koilocytes in epidermis.

III. Discussion

Epidermodysplasia Verruciformis (EV) is usually suspected when generalized wart like lesions appear very early in life. In EV, there are mutations in the EVER1 or EVER2 genes located on chromosome 17q25. The disease has no preference for gender or race. Histology shows marked hyperkeratosis with basket weave appearance with koilocytes in epidermis. Genetic factors are thought to cause EV, clinically there are two types of manifestations viz pityriasis versicolor like and seborrheic keratosis like, prolonged sun exposure leads to malignant transformation of the lesions. Nearly half of all patients with EV develop cutaneous malignancies resulting in Bowen type carcinoma and squamous cell carcinoma which usually occur after the sun exposure in the 4th or 5th decade of life. Metastasis is quite rare in EV. Malignant transformation is observed earlier in the course of the disease among patients usually working outdoors and /or living in the high-altitude places. Oncogenic EV HPV serotypes are 5,8,9,12,14,15,17,19-25. Acitretin and Isotretinoin are the drugs of choice for extensive lesions along with strict photoprotection.[1,2]

IV. Conclusion

The inheritance pattern of Epidermodysplasia Verruciformis is autosomal recessive in most patients. Although many case reports have been published earlier, [3,4,5] we report a case probably of sporadic occurrence. In the case described here, parents of the patient were not related by blood and had no EV lesions. In addition, he had no family members with the same lesions. Our patient did not use sunscreen and we recommended him to use sunscreen. Regular follow up is mandatory for him.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient has given his consent for his images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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