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"Uncommon Manifestation: Epidermodysplasia Verruciformis Mimicking Pityriasis Versicolor"

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Abstract-
Background- *epidermodysplasia verruciformis (edv) is a rare, inherited disorder that is characterized by an increased susceptibility to infection with specific human papilloma viruses (hpv) . It is characterized by hypo- or hyperpigmented macular lesions, pityriasis versicolor-like lesions, and an early tendency to transform into skin cancer.*
Case summary*-a 32 year old female presented with asymptomatic wart like multiple lesions of different sizes on face, trunk, upper limbs since childhood. Histopathology was suggestive of ev and our patient was given oral isotretinoin 20mg/day and advised for strict photoprotection.*
Conclusion- *we report a case of edv in a 32-year-old female patient with confluent wart like lesions, pityriasis versicolor like lesions and reddish plaques. The case is being reported in view of rarity of disease*
Keywords- *epidermodysplasia verruciformis, hpv, autosomal recessive, pityriasis versicolor.*

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I. Introduction-

Epidermodysplasia Verruciformis (EV) is a rare, autosomal recessive inherited disorder. It is characterized by increased susceptibility to infection with human papilloma virus (HPV) of certain types. 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 start to appear in infancy and persist throughout life mainly over the sun exposed areas of face, neck, trunk and extremities. Lesions on sun exposed sites are more prone for increased risk of malignant transformation. EV-like syndrome has been recently described in patients who have compromised cell-mediated immunity, and the term "acquired EV" (AEV) has been established. AEV has been described secondarily in immunocompromised states, including patients who have immunodeficiency virus (HIV), organ transplantation, lepromatous leprosy, systemic lupus erythematosus (SLE), Hodgkin's disease, warts, immunoglobulin M deficiency, adult T cell leuk emia, lymphedema, and graft versus- host disease. Prevalence of malignancy is very high in immunocompromised patients.30-60% of EV patients the lesions may transfer into skin cancer mostly intraepidermal squamous cell carcinoma and basal cell carcinoma. Malignant tumors are usually found in 30 -50 years of age.

II. Case Presentation-

A 32 year old female born to non- consanguineous marriage presented to the dermatology OPD with complaints of asymptomatic wart like lesions since childhood .The lesions first appeared on the leg gradually increased in number and size finally spread to hands upper back ,lower back , abdomen , chest . No history of similar complaints in the family members .Patient had visited many dermatologist for the same reason. Physical Examination revealed multiple brownish, slightly scaly, verrucous flat growths of varying size smallest being 1x1 to largest being 6x8 cm on upper back, neck, face, feet, abdomen areas (Fig 1,2,3,4) .No history of similar lesions in family members. Biopsy showed koilocytes with marked hyperkeratosis with basket weave appearance noted in the epidermis. Patient was started on tab. ISOTRETINOIN 10mg once daily for 2 weeks along with topical sunscreen was advised. Along with strict sun protection.

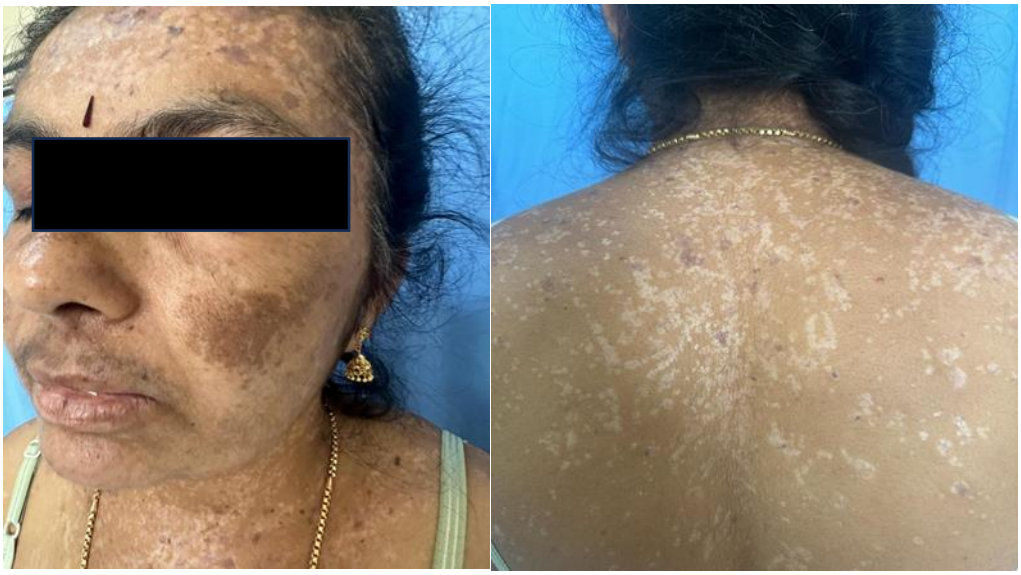

Fig 1 & 2 shows multiple hypopigmented patches and verrucous plaques on face,neck, upper back.

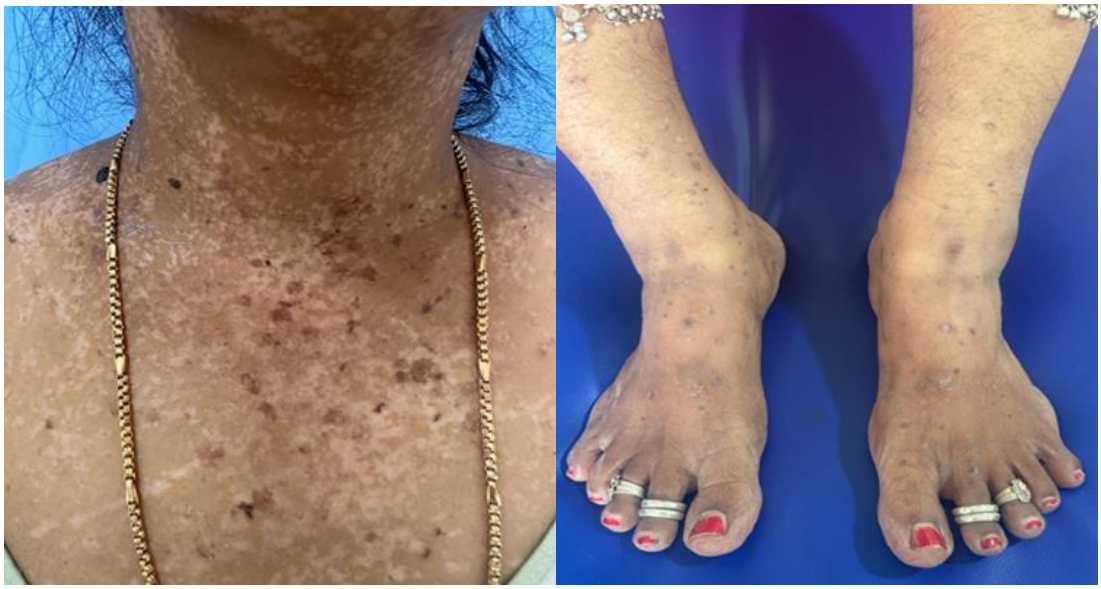

Fig 3 & 4 shows multiple hypopigmented to verrucous plaques on neck and dorsal aspect of feet.

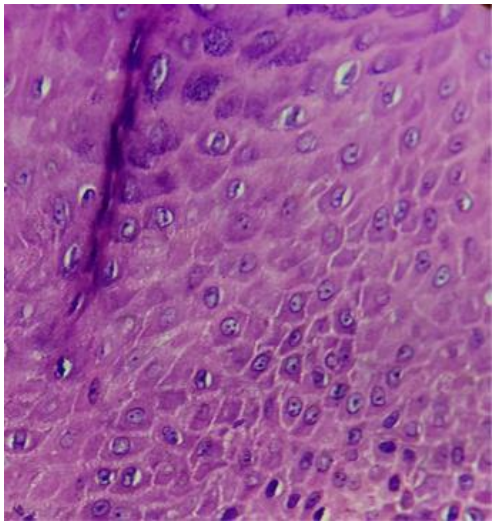

Fig 5. Histopathology shows presence of koilocytes in epidermis.

III. Discussion-

Epidermodysplasia Verruciformis (EV) is usually transmitted as an autosomal recessive disorder with mutations in EVER1/TMC6 and EVER2/TMC8 genes located on chromosome 17q 25.A HPV-specific defect of cell mediated immunity is present. There is no gender predisposition. suspected. Histologic examination of lesions and detection of HPV DNA proves the diagnosis. The gene involved are EVER1 (Epidermodysplasia Verruciformis Enhancing Region) and EVER2. Mutation in these gene causes an individual to be susceptible to develop EDV. Mutation in genes and infection by HPV14, HPV20, HPV21 and HPV25 is usually associated only with the development of benign lesions while HPV5, HPV8, HPV10 and HPV47 were found in more than 90% of EV-associated malignant neoplasms. 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. The histopathological features are hyperkeratosis, acanthosis, vacuolation in perinuclear area of keratinocytes which involves the upper half of three fourth of the spinous layer. Classic clear cells are not found in all lesions.2 There is a high risk of development of cutaneous squamous cell carcinomas in early adult life.2 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. The combination of etretinate plus IFN-α may also produce a useful clinical effect.4 Oral isotretinoin can also reduce the benign lesions. Other treatments that have been tried include topical imiquimod,7 topical vitamin D analogue,6 topical immunotherapy with squaric acid dibutylester8 and oral cimetidine.

IV. Conclusion-

The pattern of inheritance of EV is autosomal recessive although some patients can exhibit sporadic appearance. In the case described above ,the patient had parents that were not in a consanguineous marriage and were without EV lesions. In addition, she did not have family members with the same lesions and displayed a sporadic appearance. Extended exposure to sunlight and not using sunblock can precipitate the malignant transformation of EV lesions .Our patient did not use sunblock ,and we recommended that she did so . regular follow-up is also mandatory for her

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