Epidermodysplasia Verruciformis misdiagnosed as Pityriasis Versicolor

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(Received: December, 2016) (Accepted: January, 2017)

ABSTRACT

Epidermodysplasia verruciformis is an inherited autosomal recessive disorder characterized by widespread and persistent infection with HPV commonly HPV3 and 10, giving rise to a characteristic combination of plane warts, pityriasis versicolor like lesions and reddish plaques like lichen planus. It is an autosomal recessive condition though autosomal dominant and X-linked dominant cases has also been reported. In EV, there are mutations in the EVER1 or EVER2 genes on chromosome 17q25, which, due to a defect of cell-mediated immunity, lead to an abnormal susceptibility of the patients to a specific group of HPV genotypes known as EV HPV. The disease usually begins in childhood with equal prevalence in both sexes affecting persons of all races. Malignant changes like squamous cell carcinoma, basal cell carcinoma occurs in 30-70% of cases on the sun exposed parts in the age group 20-40 years, but metastasis is rare. EV diagnosis must be suspected when persistent verrucas are present in large body areas and are thus difficult to treat.

We report a case of a 20 years old male patient who came to our OPD with the complaints of asymptomatic white patches over upper chest, upper back, bilateral upper limbs and neck since 1½ year.

KEY WORDS: autosomal recessive disorder; epidermodysplasia verruciformis (EV); human papilloma virus (HPV); pityriasis versicolor (PV).

INTRODUCTION:

Epidermodysplasia verruciformis (EV) is an inherited autosomal recessive disorder representing a unique susceptibility to cutaneous human papilloma virus (HPV) infection characterized by widespread and persistent infection with HPV commonly HPV3 and 10, giving rise to a characteristic combination of plane warts, pityriasis versicolor like lesions and reddish plaques similar to lichen planus. Malignant changes like squamous cell carcinoma and basal cell carcinoma occurs in 30-70% of cases on the sun exposed parts though metastasis is rare. The condition usually has an onset between 1–20 years of age but can occasionally present in middle-age. The exact prevalence of EV is unknown.

We report a case of EV in a 20 years old male

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A 20 years old male patient presented with complaints of hypopigmented to skin macules over bilateral upper limb, upper back and upper part of chest & neck since 1½ year and six months. Lesions were asymptomatic. On close examination skin coloured to erythematous plaques was found in between the macular lesion (Figure 1 & 2). Koebnerization was seen at some places (Figure 3). Patient was diagnosed and was taking treatment for Pityriasis versicolor at private clinics before and was on topical and systemic antifungal agents since last two months, but there was no relief in his symptoms.

KOH examination was performed for fungus element, which was negative. Histopathological examination of skin biopsy revealed epidermal hyperplasia, thickening of granular layer and...
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vacuolation of keratinocytes in upper spinous layer, feature similar to verruca plana or flat wart (Figure 4 & 5). Involvement of large areas of the body with warts and failure to clear the lesions despite treatment were the typical indications to consider the diagnosis of EV.

Patient was started with topical tretinoin 0.05% gel and oral acitretin in a dose of 1mg/kg/day with sun protection for 4 months and then dose of acitretin was reduced to 0.05mg/kg/day for next 2 months. A slight improvement in skin lesions was observed, but lesions recurred upon discontinuation of treatment.

DISCUSSION:
Epidermodysplasia verruciformis (EV) is a rare, lifelong, cutaneous, autosomal recessive genetic disorder of the immune system manifested by increased susceptibility to cutaneous HPV infection beginning in the early years of life. This disorder was first described by Lewandowsky and Lutz in 1922 as an epidermal nevus. In 1939, Sullivan and Ellis described a close relationship between EV and high risk of skin cancer. Autosomal dominant and x-linked dominant pattern has also been reported. In EV, there are mutations in the EVER1 or EVER2 genes on chromosome 17q25, which, due to a defect of cell-mediated immunity, lead to an abnormal susceptibility of the patients to a specific group of HPV genotypes known as EV HPV. EVER proteins are members of transmembrane channel-like (TMC) family. They are encoded by 8 genes (TMC1-8). EVER1 and EVER2 correspond to TMC6 and TMC8, respectively.

Therefore the recent literature focus on the mutation finding the culpable gene. There are at least 20 types of HPV characteristic of EV most common among them are 5, 8, 9, 12, 14, 15, 17, 19-25. Type 5&8 are commonly associated with malignancy. Thus, EV is in essence a genetic cancer of viral origin, and could also be regarded as a model of cutaneous HPV oncogenesis.

Lesions usually develop in childhood. Lesions on face and neck are like flat warts. On trunk and limb they tend to be larger and are of two types. Scaly macular lesions like p. versicolor or thicker plaques which may be dull pink, violet or brown in colour resembling seborrhoeic keratosis.
warts are often present on sides of fingers and on palms and soles. Malignant changes occur on sun exposed skin mainly in the form of actinic keratosis, Bowen’s disease, Squamous cell carcinoma.\[7\]

EV needs to be differentiated from some clinically similar looking disorders. In Acrokeratosis verruciformis, lesion are present on dorsum on hands, feet, but histopath doesent show any vacuolation. In Lichen planus, the lesion are pruritic, pink or violaceous in colour with mucosal lesions and distinctive histology. Pversicolor sholud also be ruled out.\[8\]

Several nonsurgical treatment modalities have been tried for EV, including oral and topical retinoids, interferon, immunotherapy, electrodesiccation and cryotherapy. However, all of these treatments have either been ineffective or have had temporary results. These lesions generally require surgical excision and reconstruction of the defects, which seems to be the most effective treatment and the only way to increase survival.\[9\]

Oral retinoids have been used in early premalignant lesions. These have an antiproliferative and differentiation-inducing effect that is believed to prevent the occurrence of malignant lesions. Another proposed agent is interferon; however, the patient must have an intact immune system for clinical benefit. Imiquimod, another therapeutic agent, induces the production of cytokines from monocytes and macrophages, thus stimulating the T cell helper 1 response and most probably the cytotoxic T cell activation and B cell response.\[10\] Cimetidine, depresses mitogen-induced lymphocyte proliferation and suppressor T cell activity features.

CONCLUSION:

EV is a disfiguring and potentially stigmatizing condition, and also difficult to treat. It should be considered in patients of pityriasis versicolor not responding to treatment. Close dermatological follow up is always required to detect potential skin malignancies at the earliest.

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