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Review article

TREE MAN SYNDROME - A REVIEW

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ABSTRACT

Tree man syndrome, also known as epidermodysplasia verruciformis is recognized as an inherited disorder, which is a widespread and persistent infection with human Papilloma virus and cause defect in cell mediated immunity. This disorder was first described by Lewandowsky and Lutz in 1922 as an epidermal nevus. It mainly occurs between the age of one to twenty. The main etiology behind this genetic disorder is an inactivating pH mutation in either EVER1 or EVER2 genes, which are located adjacent to one another one chromosome. Tree man's syndrome is a lifelong disease and only the lesions can be treated or removed as they appear, which will continue to develop throughout the life. Epidermodysplasia verruciformis diagnosis must be initiated upon the appearance of verrucous lesions . No treatment is available to prevent the development of new lesions. The disease can only be managed by the combination of medical and surgical treatment along with patient counselling and education. The patients are advised not to get exposed to sunlight as it may cause the development of skin carcinoma.

Keywords: Tree man syndrome, Epidermodysplasia verruciformis, Human Papilloma virus.

INTRODUCTION

Tree man's syndrome or Epidermodysplasia verruciformis is an extremely rare autosomal recessive genetic hereditary skin disorder associated with a high risk of carcinoma of skin. The disease is characterized by high susceptibility to human papilloma virus of the skin. The condition mainly occurs between the age group of 1 and 20, but in some cases present in middle age [1].

Tree man syndrome is also known as Epidermodysplasia verruciformis or Lewandowsky Lutz dysplasia, named after the physicians Felix Lewandowsky Wilhelm Lutz de who first documented the syndrome. The resulting uncontrolled HPV infections leads to the growth of scaly macules and papules, mainly on the hands and feet of genetically susceptible individual. The HPV type 5 and type 8 are the most common infection found in about 80% of the normal population producing asymptomatic infections [2].

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ETIOLOGY

The main etiology of this condition is the inactivation of pH mutation, in either the EVER1 or EVER2 genes which are found adjacent to one other in a chromosome. These genes are responsible for regulating the distribution of zinc in the cell nuclei [3]. Epidermodysplasia verruciformis caused due to HPVs infection can be mainly divided into two types:

□ One group having high oncogenic potential (HPV types 5, 8, 10 and 47). More than 90% of cases are associated with skin cancers and contain these virus types.

□ The other group has low oncogenic potential (HPVtypes 14, 20,21and 25) and are usually detected in benign skin lesions [1].

CLINICAL PRESENTAIONS

The common symptoms are lesions that resemble the appearance of warts , extensive skin outbreak that are even to papillomatous in nature and reddish to brownish pigmented plaques located on various parts of the body . Development of thick skin which get hardened. The upper and lower extremities get enlarged and the patients become unable to use their hands and feet of patients are described as like contorted, yellow brown branches extending up to 3 feet.

The skin seems to be like tree bark or tree roots and hence the name Tree man syndrome [2]

TYPES OF LESIONS IN EPIDERMODYSPLASIA VERRUCIFORMIS;

(a) Flat types : which is presented as flat or even lesion with flat topped papules, resembling a wart and verruca plana with rough surface and hypo or hyper pigmented elongated patches .prominent large [patches are also seen , which are formed by the fusion of small patches. They may be reddish to brown in color.

(b) Seborrheic like verrucous types :it is similar to a wart and form a linear column in sun exposed areas of the body like upper and lower extremities ,neck, face, and even the ear lobes.

The lesions also appear in the genital areas, soles of feet, as well as in the axillae or in the under arm area [2].

PATHOPHYSIOLOGY

The tree man syndrome is mainly caused due to the loss of function mutation in two adjacent genes mainly:

- □ EVER1/TMC6
- □ EVER2/TMC8

The two genes are located in chromosome 17 which codes for membrane proteins, and forms a complex with Zn transporter protein (ZnT-1) which is present in the endoplasmic reticulum of keratinocytes.

Due to this mutation in gene the susceptible individual become more prone to HPV infection including the subtypes HPV-5,8,9,10,12,14,15,17,19,25,36,38,47 and 49.

TREATMENT

Tree man syndrome is a lifelong disease and the lesions can be treated and removed once they appear. The main important factor is the strict avoidance of sun exposure and skin protection as these may lead to skin cancer [4]

Pharmacological treatment

This helps to improve the quality of life of the patient [5]. The management includes the following drugs, topical imiquimod and 5-aminolevulinic acid photodynamic therapy

HPV related carcinomas include: combination of 13-cis retinoid acid and interferon alpha or cholecalciferol analogues.

Auto transplantation of skin is reported to be successful in preventing further development of cancer [4].

Surgical treatment

Surgical removal of lesions is found to be more effective than the radical pharmacological treatment [5]. Benign and premalignant skin lesions can be treated by electro surgical removal and cryotherapy.

Skin grafting can also be done and the graft used must be from sun protected skin [4].

CONCLUSION

Long term exposure to sunlight, outdoor occupations and not using sunscreen precipitate malignant degeneration of epidermodysplasia verruciformis lesions[5].

Exposure to UV-B and UV-A and X-ray irradiation must be avoided as this may cause promotion and the occurrence of most aggressive forms of skin cancer. Treatment should be taken as soon as possible and this will prevent further complications associated with the disease[4]. Adequate education and counselling must be given to the patient. A good hygienic condition must be provided to the patient.

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Nil

CONFLICT OF INTEREST

Nil

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