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Tree man syndrome-A review

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ABSTRACT

Basis for review

Tree man syndrome, also known as epidermodysplasia verruciformis is recognized as an inherited disorder with widespread and persistent infection with human papilloma virus causes defect in cell-mediated immunity. In addition, there is a link with squamous cell carcinoma. A tree man syndrome is a rare disorder happening more frequently at a younger age in the general population. It is an unusual genodermatosis characterized by an autosomal recessive inheritance pattern. Mutations in the family of genes called EVER genes cause the tree man syndrome. Non-surgical therapies and surgical therapies are known to treat this disorder, but it is known to be lifelong disorder. Recent surgery was performed in Bangladesh which is known to be successful.

INTRODUCTION

The tree man syndrome was first described by Lewandowsky and Wilhelm Lutz in 1922 where both are dermatologists. It was first described as an epidermal nevus in 1922. Hence this disease is also known by two names they are,

- Epidermodysplasia verruciformis and
- Lewandowsky Lutz dysplasia. [1]

It is a rare skin disease characterized by disseminating, refractor, Pityriasis, Versicolor like lesions as well as flat wart like lesions.[2]

DEFINITION

Tree man syndrome or tree man illness is usually a genodermatosis which is mainly characterized by persistent "HUMAN PAPILLOMA VIRUS (HPV)" infection.[3]

- These diseases carry higher risk of skin cancer (squamous cell carcinoma) when exposed to the sun.
- More than 200 cases were reported so far.
- Particularly, it is an autosomal recessive genetic disorder.[1]

FIGURE NO.1: MAN WITH TREE MAN SYNDROME

EPIDEMIOLOGY

It has no definite age of onset but most probably seen in children to young adults ranging from 1 year old to 20 years old.

It is malignant when occurs at the age of 40-50 years.[4] The percentages of cases seen, according to age group are the following:

- Infancy- 7.5%
- Childhood- 61.5%
- Puberty- 22%[5]

As the progresses, some lesions get disappear while new ones may appear. The largest series of disease is seen mainly from Eastern Europe, Poland and Latin America, which includes 195 cases. [6]

SIGNS AND SYMPTOMS

- The hands and feet were enlarged and also they were difficult to use
- Deform, yellow-brown branches extending up to 3 feet may appear
- Increased Skin becomes hard and thickened[8]
- Susceptibility to human papilloma virus on skin
- Flat red, brown macules
- Skin bumps on back of head [9]
- Patients can also develop Tinea versicolor-lesions on the trunk and
- It may usually begin in childhood with various types of flat wart like lesions on the skin as well as inside the skin. [7]

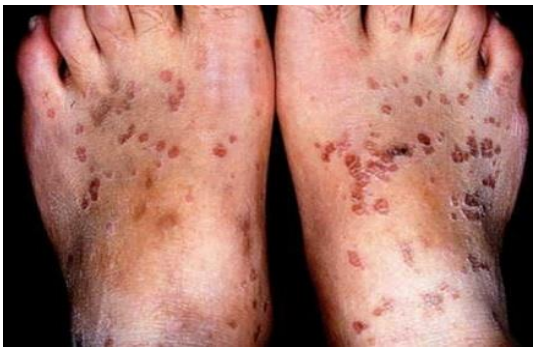

FIGURE NO.2: YELLOW BROWN PATCHES

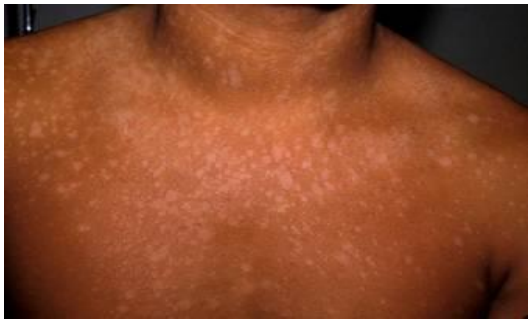

FIGURE NO.3: TINEA VERSICOLOR

PATHOPHYSIOLOGY

Two forms of the syndrome are recognized they are as follows:

- This type of syndrome shows multiple plane warts. This form was widely distributed and it does not show any malignancy transformations.
- The second form of the syndrome is induced by HPV-5 and sometimes HPV-8, 9, 14, 20,24,28,47 and others. The most familial history is with an autosomal recessive inheritance. Malignant transformations occur in about 25% if patients on the oncogenic potential of viruses.[10]

The tree man syndrome can be caused due to loss in function mutations in either of 2 adjacent genes they are:

- EVER1/TMC6 (or)
- EVER2/TMC8

These genes are located on chromosome 17

These chromosomes 17 codes for membrane proteins which form a complex with Zn transporter protein (ZnT-1) in the endoplasmic reticulum of keratinocytes.

These mutations in genes lead to susceptibility to infection with specific HPV subtype, including HPV-5, 8,9,10,12,14,15,17,19-25,36-38,47 and 49 which are harmless to healthy individuals.[5]

RISK FACTORS

Genetic

Many researches shown that predisposed deficiencies in cutaneous immunity make patients of this disease more vulnerable to HPV infection

Any mutations in the family of genes called EVER genes who's deficiency cause TREE MAN SYNDROME.

Acquired

- Squamosal cell carcinoma,
- Acrokeratosis verruciformis,
- Tinea versicolor,
- Generalized verrucosis, [5]
- Immunosuppression,
- Profound CD8+T cell lymphocytopenia [1]

TREATMENT

Medical care

The tree man syndrome is known to be lifelong disease, but lesions can be treated or removed as these lesions will continue to appear throughout life in these patients. Currently there is no treatment to prevent new lesions from occurring. But it is important to reduce the risk of skin cancer.

The main important factor is strict sun avoidance and protection [11].

A non-surgical therapy for skin cancer management includes the following:

- Topical imiquimod and 5- fluorouracil,
- Systemic retinoids and
- Interferon and 5-aminolevulinic acid Photodynamic therapy
  - HPV related carcinomas include
- Combination of 13-cis retinoid acid and interferon alpha (or) cholecalciferol analogues
  - For localized multiple malignant lesions Auto transplantation of skin, which reported to be successful in preventing further development of cancer.[12]

Surgical care

Electro surgical removes and cryotherapy are used in the treatment of benign and premalignant skin lesions. If skin grafting is necessary, the graft should be from sun protected skin.[12]

RECENT SURGERIES

Tree man was nicknamed to a Bangladeshi Abul Bajandar from the southern district of khulana for massive bark like warts on his hands and feet where it was first begun appearing 10 years ago. Abul bajandar was undergoing for surgery preparations to cut all the outgrowths where doctors weighed it about 5 kgs/11 lbs. Bajandar said that,

"Initially, I thought that they are harmless", the 26 year old told at the Dhaka medical college hospital. But slowly I lost all my abilities to work.There are now dozens of 2-3 inch root in both my hands and there are some small ones in my legs, the one who was forced to quit working as a bicycle puller. The Doctor's team will perform surgery at the Dhaka medical college hospital named as top hospitals in Bangladesh for free of cost. "The warts, which started appearing in teenage, began spreading rapidly after 4 years and diagnosed it as a tree man syndrome or epidermodysplasia verruciformis" a doctor said.

It was a successful operation. All the warts were removed from all the five fingers of his right hand and left hand and proper medications were given.[13

Figure no.4: Abul Bajandar before surgery

Figure No.5: Abul bajandar after right hand surgery

In March 2007 Dede koswara was diagnosed with tree man illness. He is an Indonesian man. He said that "It all started as a simple wart on his knees, but it turned his life upside down." The

warts covered his body with HPV. He had more than 6 kgs of warts. He underwent a surgery in august 2008.usually the surgery involves three steps they are:

- The first step includes removal of massive horns and thick carpet of warts on hands.
- Second step includes removal of small warts on his torso, head and feet and
- The last step includes covering of hands with grafted skin.

Unfortunately the warts reappeared. Doctors said that there is a need of at least 2 surgeries a year to make the condition manageable. His condition is still miserable.[14]

Figure No.6: Dede Koswara before surgery

Figure No.7: Dede koswara after surgery

CONCLUSION

The tree man syndrome is known to be a lifelong disorder, but lesions can be treated or removed. Many of the malignant lesions were seen only on sun exposure. UV-B and UV-A exposure and X-ray irradiation is to be avoided as this may cause or promotes the occurrence of most aggressive skin cancer. Hence precautions should be taken. Treatment should be taken as soon as possible to prevent the further complications of the disease.

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