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Review Article
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A Detailed Study on Epidermodysplasia Verruciformis

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Epidermodysplasia verruciformis is also known as "A tree man syndrome". EV is an autosomal recessive genetic hereditary skin disorder. The purpose of this review was to provide information for the patient and caretakers or families for better understanding of the disease so that they are in a better position to help epidermodysplasia verruciformis patients with many problems they faced.

INTRODUCTION: EV is a skin disease which begins in childhood and remains within the individual for their lifetime. It is characterized by widely spread, refractory skin lesions which resemble flat warts. Individuals with EV are highly susceptible to human papillomavirus (HPV), and if infected with HPV, will go on to develop these physical symptoms. However, scientists hypothesize that they have roles regarding zinc distribution within the nuclei of cells. Disruption of the chemical and elemental balances with a cell can have catastrophic complications.

SYMPTOMS:

Symptoms can appear at any age, including infancy. In more than half of EV cases, symptoms first appear in children between the ages of 5 and 11. For nearly a quarter of people with EV, symptoms first emerge during puberty.

Symptoms may include a mix of:

- flat-topped or bumpy lesions
- small, raised bumps known as papules
- large patches of raised and inflamed skin, known as plaques
- small, raised brown lesions that resemble scabs

Flat lesions are more common in areas exposed to sunlight, such as:

- hands
- feet
- face
- ears

Many of these lesions take on the appearance of tree bark or tree roots. Because of this, EV is sometimes referred to as "tree-man disease" or "tree-man syndrome."

Growths may be limited to a small cluster of just a few warts or extend to more than 100 warts covering much of the body.

Plaques usually develop on the:

- neck
- arms
- armpits
- palms of the hands
- trunk
- legs
- soles of the feet
- external genitals

CAUSES:

EV is a type of genetic disease known as an autosomal recessive inherited disorder. That means a person must have two abnormal EV genes — one from each parent — to develop EV. In most cases of EV, the genetic mutation is sporadic, meaning it developed when the sperm or egg first formed. These genes can still be passed on to future offspring.

About 10 percent of people with EV have parents who were blood relatives, meaning they shared a common ancestor.

People who have EV have a normal immune response to non-HPV infections. However, for reasons still not completely known, these individuals are more prone to infection with certain HPV subtypes. There are over 70 HPV subtypes that can cause warts. But the subtypes most often found in EV are not the same as the those that most commonly cause genital warts and cervical cancer.

People with EV may have a poor immune response to HPV or other wart viruses. There have been about 30 HPV subtypes identified as triggers for warts and plaques in people with EV.

Although symptoms primarily appear by puberty, EV can develop at any age. Males and females appear to be equally at risk.

DIAGNOSIS:

Diagnosing a rare disease of any kind can be a challenge. If warts or unusual lesions appear, you should see a dermatologist, even if the symptoms appear to be mild.

Your doctor should know your medical history and have an understanding of your symptoms: what they are, when they started, and if they've responded to any treatment. Your doctor will also examine your skin.

If your doctor suspects EV or any other condition, they may take small tissue samples for a biopsy. An EV skin biopsy will include tests for HPVs and other signs that might point to EV. Skin cells called keratinocytes can sometimes reveal HPVs associated with EV.

TREATMENT:

There is no cure for EV, so treatment is primarily to alleviate symptoms. Although surgery to remove the lesions can be successful, it may only be a temporary solution. Lesions can develop again, though they may never come back or may take years to return.

One surgical option available is curettage. It involves the use of a spoon-shaped device called a curette. Your surgeon uses the curette to carefully scrape away a lesion. The goal is to preserve as much healthy skin under and around the lesion as possible.

EV warts may be treated as you would other types of viral warts. These treatments include:

- chemical treatments, such as liquid nitrogen
- topical ointments such as Verrugon that contain salicylic acid
- cryotherapy, in which the wart is destroyed by freezing it

Another important aspect of treatment is limiting sun exposure and using sunscreen to help preserve the health of your skin. People with EV are at an increased risk of developing skin cancer. Following your dermatologist's advice about skin care and sun protection is very important. In fact, up to two-thirds of people with EV will develop skin cancer, usually in their 20s or 30s.

OUTLOOK:

EV is a lifelong condition. Although surgery can usually remove lesions temporarily, they often return.

One of the greater concerns with EV is the increased risk of skin cancer. As many as 50 percent of people with EV have lesions that turn cancerous. Research suggests that this typically occurs between the ages of 40 and 50.

Squamous cell carcinoma and intraepidermal carcinoma are most likely to develop.

Your skin cancer risk may be based on the type of HPV infection involved. Most of the skin cancers associated with EV contain HPV types 5, 8, 10, and 47. HPV types 14, 20, 21 and 25 are usually benign.

If you have EV in any form, it's crucial that you work with your doctor or dermatologist to reduce your risk of skin cancer. This includes regular screening and daily use of sunscreen.

PREVENTION:

Because EV is an inherited condition, there is little anyone with the abnormal genes can do to prevent the disorder.

If you have a family member with the disease or know that your parents carry the abnormal EV gene, talk with your doctor. They can test your genes and determine your next steps.

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