

Epidermodysplasia Verruciformis

Also known as ... Treeman syndrome, Lewandowsky-Lutz syndrome, Lewandowsky-Lutz dysplasia

What is Epidermodysplasia Verruciformis?

Epidermodysplasia verruciformis (EV) is a rare inherited skin condition that predisposes a person to infections caused by the human papillomavirus (HPV).

What causes Epidermodysplasia Verruciformis?

EV is an inherited disorder. In most cases, it is inherited in an autosomal recessive manner. This means a person must have both copies of mutated EV genes to be affected. Most people can carry a copy of the mutated gene but not show signs nor symptoms. However, there has been some cases of atypical inheritance reported.

'Acquired epidermodysplasia verruciformis' or EV-like eruptions can result from having an impaired immune system i.e. HIV infection, organ transplantation or cancer.

What does Epidermodysplasia Verruciformis look like?

The disease usually manifests in childhood with multiple pink or tan flat bumps (papules), flat pinkish white or brown spots that resemble pityriasis versicolor or thicker warty lesions that resemble seborrheic keratosis.

Lesions are most numerous on the face, neck and backs of the hands and feet.

What other problems can occur with Epidermodysplasia Verruciformis?

A major concern is that patients with EV develop skin cancers, especially squamous cell carcinoma. Exposure to sunlight increases the risk of the EV lesions transforming into skin cancers.

How is Epidermodysplasia Verruciformis diagnosed?

It is diagnosed based on clinical and skin biopsy findings. HPV subtype can be detected within the cells using specialised techniques (PCR).

How is Epidermodysplasia Verruciformis treated?

EV is a lifelong disease. Treatment options are available for of individual lesions. These include:

- Cryotherapy
- Curettage and electrodessication
- Laser ablation
- Topical imiquimod cream
- Topical 5-fluorouracil cream
- Oral retinoids i.e. acitretin and isotretinoin
- Photodynamic therapy (PDT)
- Surgical excision this is the treatment of choice for skin cancers

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Prevention of skin cancers is important. This can be achieved by strict sun protection and regular skin checks.

What is the likely outcome of Epidermodysplasia Verruciformis?

EV has a favourable prognosis, given skin cancers appear progressively and are usually treated before they spread.

This information has been written by Dr Ruby Lee and Dr Heba Jibreal Published: October 2020