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Cancer Prone Disease Section

Mini Review

Epidermodysplasia verruciformis

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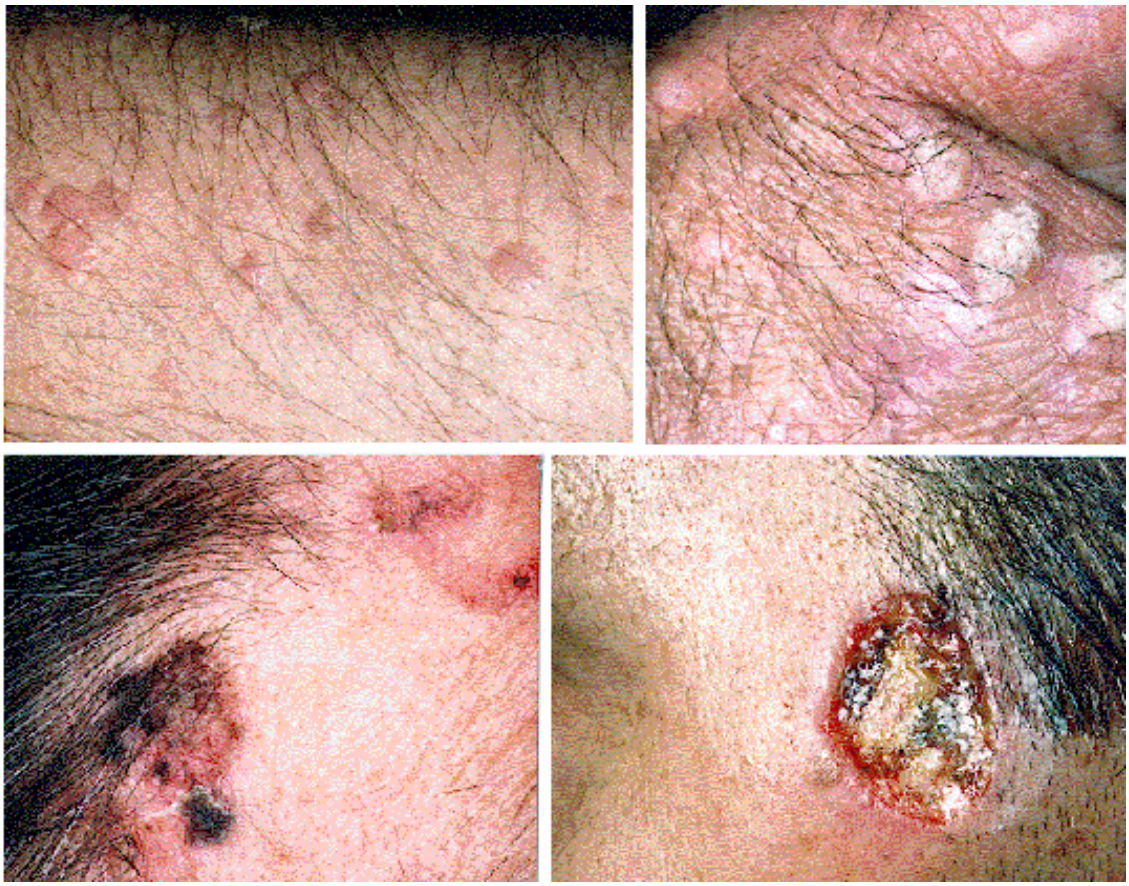
Identity

Other names: Levandowsky-Lutz syndrome
Inheritance: autosomal recessive in 10 to 20 % of the patients; other cases are sporadic; rare disease.

Clinics

Epidermodysplasia verruciformis (EV) is a model of malignant transformation from benign cutaneous viral lesion.



Top left: numerous papule-like warts on the skin of an hand; right: other aspect of papule-like warts on one hand; bottom left: basocellular carcinoma of the face developped from epidermodysplasia verruciformis lesions; right: carcinoma of the face developped from epidermodysplasia verruciformis lesions - Courtesy Daniel Wallach.

Phenotype and clinics

Age at onset is variable; more frequently: young adults or children. Two types of elementary cutaneous lesions are observed:
- Persistant papule-like warts, isolated, or confluent with a psoriasic aspect;
- White spots, pityriasis versicolor-like.

Both types of lesions are localized mainly on the outer part of the hands, on foreharms, legs, face, trunk and perianal zone.
Immunodeficiency: decreased immune response of T lymphocytes to mitogens, decreased humoral response to Human Papilloma Virus antigens; EV lesions have been described in renal transplant recipients.
Various Human Papilloma Virus (HPV) subtypes are regularly detected in the cutaneous lesions: HPV 5, 8, 9, 12, 14, 15, 17, 19, 25, 36, 38, 47, 50.
Patients are simultaneously affected by differents HPV subtypes, according to disease localisation; these subtypes are different from those observed in common warts (HPV 2, 3, 4, 10).

Neoplastic risk

Risk of malignant transformation of cutaneous lesions is within a delay of 20 to 30 yrs (very slow process comparable to the genital carcinogenesis associated with high risk HPVs).
Cytology: squamous cell carcinoma (spinocellular or basocellular carcinoma, Bowen disease).
Correlation with oncogenic subtypes of HPV found in the transformed lesions: HPV 5, 8, 14; the most frequent subtypes are HPV 5 and 8 (90% of cases).
Benign familial forms are associated with HPV 3, without malignant evolution.
HPV-5 is present in the macular lesions.
Probable potentialisation by UV ligth: 25 to 30% of malignant lesions localised to the face and forehead (hypothetic role of P53 mutations).
Protein E6 and/or E7 (tumour suppressor function) from HPV seem to be involved in the malignant transformation.

Treatment

Surgical resection of localized lesions; chemotherapy with Acitretin (25 mg/j) for multifocal lesions.

Evolution

Local recidives, enhanced by UV exposition.

Genes involved and Proteins

Complementation groups:
Genes and proteins are unknown.

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