

Epidermodysplasia Verruciformis-like Lesions in a Patient with Systemic Lupus Erythematosus*

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Sir,

Epidermodysplasia verruciformis (EV) is a rare, frequently familial disease, caused by chronic infection with human papillomavirus (HPV). EV is characterized by numerous flat warts and red and brownish macules, which sometimes strongly resemble pityriasis versicolor. A high percentage of these lesions develop into squamous cell carcinomas. Lesions similar to those present in EV have been clinically and histologically reported in patients with immunosuppression. We present here a patient with systemic lupus erythematosus (SLE) and EV-like lesions associated with HPV-17 and HPV-20.

CASE REPORT

A 32-year-old woman presented with a 3-year history of warty papules on her hands suggestive of common warts. Over the subsequent years, she progressively developed asymptomatic brownish pityriasis versicolor-like macules on her upper chest and neck. Her parents were not consanguineous and there was no family history of EV lesions. The patient had been diagnosed with SLE 13 years earlier (malar rash, photosensitivity, renal and hematologic disease, elevated ANA and anti-DNA antibodies). Since then, she had been treated with 33 intravenous pulses of cyclophosphamide, 3 pulses with methylprednisolone, oral prednisone (15–30 mg daily) and oral azathioprine (75–150 mg daily). Different complications such as an aspergillus pulmonary infection, hemophilus influenza pneumonia, and salmonella diarrhea appeared during the course of her disease. At present, the SLE appears stable, controlled with oral azathioprine and prednisone.

Physical examination revealed some flat warts on the dorsum of her hands, and numerous disseminated, ill-defined, slightly scaling, macular pityriasis versicolor-like lesions on her neck and upper chest (Fig. 1). Laboratory tests including blood cell count, erythrocyte sedimentation rate, and serum biochemistry were within normal limits. IgA (87.5 mg/dl, normal 100–300) and IgM (20.2 mg/dl, normal 80–250) were decreased with normal IgG. There was also a low absolute peripheral blood lymphocyte count (652 cells/ μ l, normal 1,600–2,400), with decreased CD3+ cells (595 cells/ μ l, normal 1,100–1,700) and CD4+ cells (187 cells/ μ l,



Fig. 1. Macular pityriasis versicolor-like lesions on the patient's neck and upper chest.

normal 500–900). HIV serology and delayed hypersensitivity reaction to tuberculin were negative. A cutaneous biopsy from one of the neck lesions showed a slight acanthosis. The granular and upper spinous layers contained swollen keratinocytes with abundant blue-grey granular cytoplasm (Fig. 2). A slight nuclear variability was also present. The presence of HPV17 and HPV-20 in the cutaneous lesion was demonstrated by DNA amplification by PCR. HPV-20 was also found in a biopsy from the adjacent apparently healthy skin. The patient was given different treatment modalities (cryotherapy, electrosurgery, imiquimod), which achieved a partial clearing of the lesions.

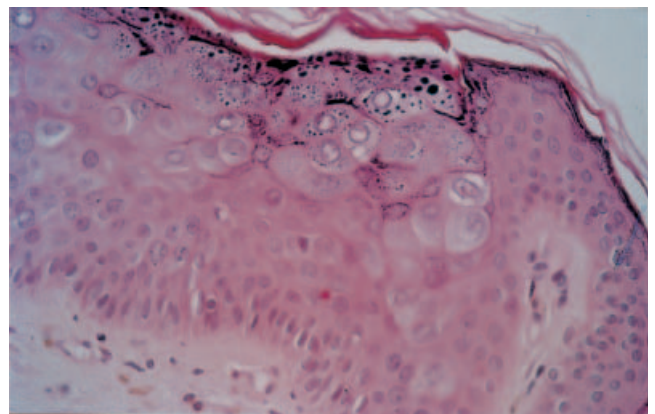


Fig. 2. The granular and upper spinous layers contained swollen keratinocytes with abundant blue-grey granular cytoplasm.

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DISCUSSION

The description of EV or an EV-like syndrome associated with immunodeficiency states such as transplantation, HIV infection, lymphoma, leukemia, lepromatous leprosy or thymoma is well described in the literature (1–3). In a review of the literature through MEDLINE records, we found only one previous patient associated with SLE (4). It is controversial whether these cases represent a true EV or a similar syndrome, as the clinical lesions and histopathologic features are indistinguishable from those seen in EV, except for the fact that in some patients with the EV-like syndrome the lesions are almost exclusively seen in UV-exposed skin (2). In our patient the immunosuppression induced by the long-lasting immunosuppressive therapy for SLE seems to have been the underlying cause of the genesis of her cutaneous lesions. To date, close to 80 HPV types have been identified. Over 20 of them, including HPV-17 and HPV-20, have been found in patients with EV. Similarly to clinical and histopathologic data, the typing of HPV does not discriminate among EV typical

lesions and EV-like patients associated with immunosuppressive states. The finding of HPV-20 in the healthy skin of our patient might be explained by the suggested wide distribution and latent state of the HPV among the immunosuppressed population.

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