Hypopigmented skin lesions in a young boy

Douieb AK, MD[1], Bayoumi Eassa, MD[2]

[1]Consultant, Department of Dermatology, HPLM, Larache, Morocco

[2]Department of Dermatology, Farwaniya Hospital, Kuwait

Consulted for an asymptomatic hypopigmented and scaly eruption. On examination, he was presented with widespread hypopigmented and mild scaly lesions simulating pityriasis versicolor on the trunk and multiple plane warts on hands.

A 12 year-old boy presented with a history of multiple asymptomatic hypopigmented and scaly eruption since he was 3 years old. There was no family history of skin cancer or similar lesions and no history of consanguinity. Physical examination revealed numerous flat-topped mild scaly papular lesions on face, neck, upper back, extensor surfaces of the forearms, and thighs (Fig. 1 A, B, C, D). Some of them appeared verrucous and other pityriasis versicolor-like. The mucous membranes, hair, and nails were unaffected. The lesions were refractory to ketoconazole cream.

Laboratory findings including complete blood count, urinalysis and serum glucose level were within normal limits. Hepatitis, tumor markers and anti-human immunodeficiency virus (anti-HIV) antibodies were negetive.

The biopsy of one of these hypopigmented lesions showed an acanthotic epidermis with a prominent granular layer with dense keratohyaline granules and keratinocytes with clear cytoplasm, vesiculous nuclei, and perinuclear halos.

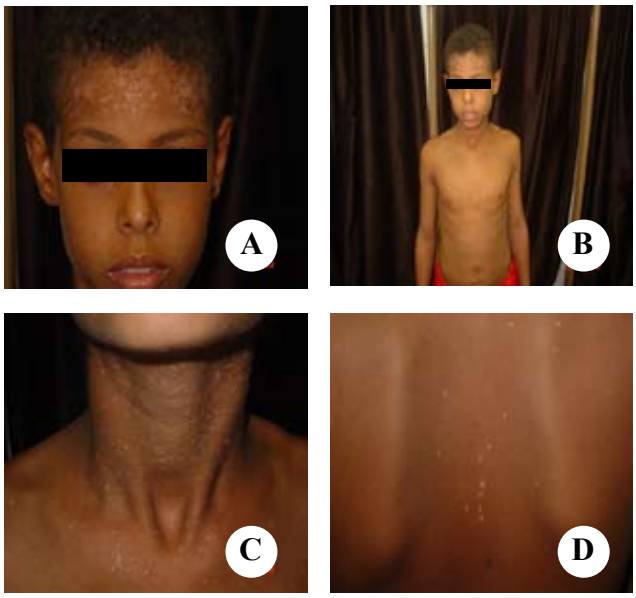

Fig. 1 A, B, C, D. Multiple flat-topped mild scaly papular lesions on face, neck and upper back

What's your diagnosis?

1. Verruca plana
2. Tinea versicolor
3. Epidermodysplasia verruciformis
4. Benign squamous papillomas

The diagnosis is epidermodysplasia verruciformis (EV).

DISCUSSION

Epidermodysplasia verruciformis is a rare lifelong cutaneous disorder associated with a high-risk of skin cancer. Cutaneous lesions are highly polymorphic and consist of pityriasis versicolor–like lesions; wart-like lesions; red macules; and seborrheic keratosis (SK)-like plaques, which usually

Correspondence: *Douieb AK, MD, Consultant, Department of Dermatology, HPLM, Larache, Morocco , E-mail : douieb@menara.ma*

manifest on the face, trunk, and extremities.

EV results from an abnormal susceptibility to specific human papillomavirus (HPV) genotypes and to the oncogenic potential of some of them, particularly HPV5 and HPV8.[1] Patients are unable to reject their lesions, and squamous cell carcinomas and Bowen's disease can develop on sun-exposed areas.

EV is usually an autosomally recessive disorder.[2] Approximately 50% of the patients carry mutations in the EVER 1, 2 genes on chromosome 17q25. There is a resulting defect in cell-mediated immunity, leading to an abnormal susceptibility to HPV infections[3-4] and a propensity to developing skin cancer. Implicated HPV strains include serotypes 3, 5, 8-10, 12, 14, 15, 17, 19 to 25, and 47. [5-6]

Histopathology of the lesions exhibited typical enlarged vacuolated keratinocytes with pale cytoplasm and round keratohyaline granules in upper epidermal layers. A malignant transformation of skin lesions occurs in more than half of the patients in the fourth and fifth decades of life. The preferential location of premalignant lesions, the forehead and other sun exposed areas, as in our patient, is suggesting a role for ultraviolet light–induced carcinogenesis.[7]

In reviewing the literature for treatment options, there was no evidence of a definitive treatment. Monotherapy with interferons, or in combination with retinoids or imiquinod, has been of value in the treatment of EV as well as surgical treatment.[8–13]

REFERENCES

1. Orth G, Jablonska S, Favre M, et al . Characterization of two types of human papillomaviruses in lesions of epidermodysplasia verruciformis. Proc Natl Acad Sci USA 1978; 75:1537-41.
2. Lutzner MA. Nosology among the neoplastic genodermatoses. In: Mulvihill JJ, Miller RW, Fraumeni JF Jr, eds. Genetics of Human Cancer. New York: Raven Press, 1977:145-67.
3. Ramoz N, Taieb A, Rueda L. Evidence for a non-allelic heterogeneity of epidermodysplasia verruciformis with two susceptibility loci mapped to chromosome regions 2p21-p24 and 17a25. J Invest Dermatol. 2000;114:1148-53.
4. Ramoz N, Rueda LA, Bouadjar B. Mutations in two adjacent novel genes are associated with epidermodysplasia verruciformis. Nature Gene. 2002;32: 579-81.
5. Lowy DR, Androphy EJ. Warts. In: Freedberg IM, Eisen AZ, Wolff K, editors. Fitzpatrick's Dermatology in General Medicine. 6th Ed. McGraw-Hill: New York; 2003. 2119-31.
6. Pfister H. Human papillomavirus and skin cancer. J Natl Cancer Inst Monogr. 2003;31: 52-56.
7. Majewski S, Jablonska S. Epidermodysplasia verruciformis as a model of papillomavirus-induced genetic cancer of the skin. Arch Dermatol 1995;131: 1312-18.
8. Gubinelli E, Posteraro P, Cocurccia B, et al. Epidermodysplasia verruciformis with multiple mucosal carcinomas treated with pegylated interferon α and acitretin. J Dermatol Treat 2003; 14: 184-88.
9. Anadolu R, Oskay T, Erdem C, et al. Treatment of epidermodysplasia verruciformis with a combination of acitretin and interferon α-2a. J Am Acad Dermatol 2001; 45: 296-99.
10. Nimura M. Application of β-interferon in virus-induced papillomas. J Invest Dermatol 1990; 95:132-37.
11. Gaspari AA, Zalka AD, Payne D, et al . Successful treatment of a generalized human papillomavirus infection with granulocyte-macrophage colony-stimulating factor and interferon γ immunotherapy in a patient with a primary immunodeficiency and cyclic neutropenia. Arch Dermatol 1997; 133: 491-96.
12. Baskan EB, Tunali S, Adim SB, et al . A case of epidermodysplasia verruciformis associated with squamous cell carcinoma and Bowen's disease: a therapeutic challenge. J Dermatol Treat 2006; 17:179-83.
13. Sanclemente G, Garcia JJ, Gomez LF, et al . An unusual presentation of human papillomavirus (HPV) infection in a black epidermodysplasia verruciformis (EV) patient. Int J Dermatol 2007; 46:199-201