EPIDERMODYSPLASIA VERRUCIFORMIS WITH BOWENOID CHANGES

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A 21-year - old female presented with epidermodysplasia verruciformis.Histopathological study of skin lesions showed Bowenoid changes. This case clearly indicates that subclinical Bowenoid changes in epidermodysplasia verruciformis may develop and should be confirmed by skin biopsy.

Key words: Epidermodysplasia verruciformis, Bowenoid changes

Introduction

Epidermodysplasia verruciformis (EV) is a rare cutaneous infection due to human papilloma virus. It was first described in 1922 by Lewandowsky and Lutz[1]. Familial occurrence has been reported. Twenty to thirty percent of lesions turn malignant to either Bowen's disease, basal cell carcinoma or squamous cell carcinoma.[2]

Case Report

A 21-year-old woman presented with multiple asymptomatic skin lesions over the body since the age of 6 . She was the 4th sibling of a non-consanguinous marriage. Her brother had similar lesions since the age of eight. The lesions initially appeared on the face and later involved the trunk and extremities. There was no history of spontaneous regression.

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Examination revealed multiple discrete, violaceous, flat-topped, warty papules with irregular borders, varying in size from 1 - 10mm on the trunk, extremities and face. Some of the papules showed scaling. Papules over the neck, cubital and popliteal fossae coalesced to form plaques. Koebner phenomenon was present. Multiple hypopigmented scaly macules, some of which coalesced to form patches with polycyclic configuration were distributed over face, neck, trunk and extremities. Scalp, genitalia, palms and soles were spared. Nails were normal. Oral mucosa was normal.

Routine haemogram and urinalysis were normal. Biopsy of a violaceous papule showed hyperkeratosis, acanthosis and mild papillomatosis of epidermis. In the superficial layer of the acanthotic epidermis were seen dyskeratotic cells with prominent keratohyaline granules. Deeper down in the epidermis were seen large cells with hyperchromatic nuclei and cytoplasmic vacuolation. Some of the cells showed intracytoplasmic keratinisation. Superficial dermis showed nonspecific inflam-

mation. A few of the keratinocytes showed large nuclei with clearing of chromatin. The overall picture was consistent with Bowenoid changes in EV.

Discussion

Epidermodysplasia verruciformis is a rare, life-long generalised infection with human papilloma virus. It is characterized by the development of cutaneous carcinomas often at an early age. The cancers are usually in situ with Bowenoid features.[1] Multiple Bowenoid foci have been reported in seborrhoeic keratosis-like lesions in epidermodysplasia verrucifomis.[3] The oncogenic potential of the virus,[2] the decreased cell mediated immunity,[4] ultraviolet light [5] and impaired DNA repair,[6] may be factors contributing to increased inidence of malignancy in epidermodysplasia verruciformis.

Clinically the occurrence of Bowen's disease is heralded by the appearance of pigmented, crusted lesions. However, in our patient crusted lesions suggestive of Bowen's disease were not seen clinically. Histopathological examination brought out the Bowenoid transformation, thereby emphasising the importance of biopsy in the early detection of Bowenoid changes in EV.

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