EPIDERMODYSPLASIA VERRUCIFORMTS

M L Khatri, M Shafi

A Libyan male child aged 10 years having coeliac disease and moderately retarded growth developed epidermodysplasia verruciformis (EV) since the age of 4 years and later (8 years age) developed a lesion of porokeratosis of Mibelli on the cheek. Such an association has not been reported earlier.

Key Words : Epidermodysplasia verruciformis, Coeliac disease, Porokeratosis of Mibelli

Introduction

Epidermodysplasia verruciformis (EV) is a rare, life long disease characterized by persistent, refractory HPV infection manifesting as disseminated, flat wart-like lesions and erythematous, hypopigmented or hyperpigmented macules and development of skin cancer in about one third of the patients. It was first decribed in 1922 by Lewandowsly and Lutz. It is believed to be a multifactorial disease with viral, genetic, immunologic and extrinsic factors playing a role it its expression.[1] At least 23 HPV types have been isolated in EV patients, 21 of these occur exclusively in EV patients (HPV 3 and HPV 10 are found both in EV patients as well as in common flat warts).[1] The genomes of EV-specific HPVs were found to have a variable degree of homology and on this basis have been classified into four main groups.[2]

1. HPVs 5, 8, 12, 14, 19, 20-23, 25, 36
2. HPVs 9, 15, 17, 37, 38
3. HPV 24
4. HPVs 3 and 10

From the Department of Dermatology, Central hospital and Faculty of Medicine, Al-Fateh University of Medical Sciences, Tripoli, Libya.

Address correspondence to : Dr M L Khatri, P.O. Box - 13457, Tripoli, Libya.

The susceptibility to the virus is inherited usually through an autosomal recessive gene, though autosomal dominant and X-linked dominant patterns have been reported.[3] A defect in cell-mediated immunity is thought to be a reason of life long HPV infection. Ultraviolet light plays a part in carcinogenesis, probably by diminishing the number of Langerhans cells.[2]

We describe a case of EV with unusual association of coeliac disease and porokeratosis of Mibelli.

Case Report

A 10-year-old Libyan male child presented with gradually developing generalized maculopapular lesions, since the age of 4 years. The lesions initially appeared over face and slowly became generalized in two years with maximum intensity over the face, dorsum of hands and trunk. The lesions were slightly hypopigmented with mild scaling in some and were asymptomatic. The size of the lesions varied from 2 to 4 mm. For the last two years he also developed a gradually enlarging well defined thin plaque with raised margins and atrophic centre on the right cheek which is now 5 cm in size (Fig. 1).

The child was diagnosed to have coeliac disease at the age of three years

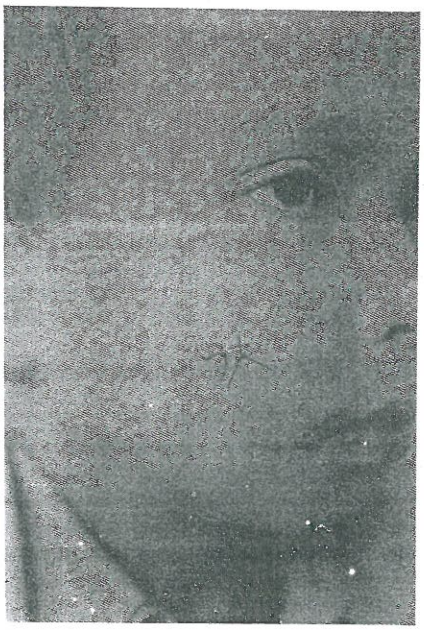

Fig. 1. Hypopigmented lesions of EV on face with lesion of porokeratosis of Mibelli on right cheek

and has been on gluten free diet. His physical growth is moderately retarded but his mental development is normal.

Routine investigations on blood, urine and stools did not reveal any abnormality except mild anaemia (Hb 11.3 gm% and RBC 3.9 million/ cu mm). Lipid profile revealed total lipids 370 mg% with cholesterol 88 mg% and triglycerides 60 mg%. Immunologic screening revealed a slightly decreased IgG 531 mg/ dl (normal 735 to 1685 mg/ dl), IgA and IgM were within normal limit of 118 mg/ dl (normal 70-312 mg/dl) and 221 mg/dl (normal 63-277 mg/dl), respectively.

Biopsy from the trunk lesion revealed basket weave appearance of stratum corneum with little parakeratosis at places. The cells in the stratum granulosum and spongiosum showed vacuolization and pyknotic nuclei at some places. Dermis showed some nonspecific inflammatory cells. The findings were characteristic of EV. Biopsy from the cheek lesion revealed hyperkeratosis, parakeratosis and hyperkeratotic plug having feather-like arrangement of parakeratotic nuclei. The granular cell layer was well marked. Dermis showed infiltration of chronic non-specific inflammatory cells mainly lymphocytes, plasma cells and histiocytes. The picture was suggestive of porokeratosis of Mibelli.

Comments

In reported cases of EV, premalignant lesions have been mostly in the form of actinic keratosis.[3] Our patient developed porokeratosis of Mibelli on the cheek at the age of 8 years. Such an association with EV has not been reported earlier. However development of porokeratosis of Mibelli has been reported in immunosuppressed patients.[4] Porokeratosis of Mibelli has been reported to have developed into squamous cell carcinoma in at least 20 patients.[5] Hence we can presume that development of porokeratosis of Mibelli in this case is a premalignant change.

Our patient also suffered from coeliac disease. Such an association has also not been recorded previously. Association of coeliac disease might have lowered the cell-mediated and humoral immunity of the patient which might be a contributing factor in the pathogenesis in this case. Cell-mediated immunity (CMI) studied by in-vitro and vivo tests, was found to be depressed in most of the EV patients.[2,6] We could not investigate the CMI status of our patient. Similar finding has been reported by Slawsky et al.[6] Although no defect of non-specific or specific humoral responses has been

substanciated, low antibody titers or an absence of antibodies in some patients in spite of life long infection fould contribute, at least in part, to spread of the viruses.[2]

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