A Case of Erythrokeratodermia Variabilis in Korean

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Dear Editor:

Erythrokeratodermia variabilis (EKV) is a rare, heterogeneous, autosomal dominant genodermatosis. It is characterized by two distinct features: transient erythematous patches of various size and morphology and fixed hyperkeratotic plaques. Here, we describe a rare case of EKV of a Korean male patient.

A 39-year-old male was presented with pruritic erythematous patches and keratotic plaques on the whole body, which had persisted since birth. The keratotic lesions of the knee, axilla, buttock, palm and sole were consistently maintained on the same location, but erythematous patches transiently showed depending on stress and weather. The elder brother had a history of similar skin lesions since birth and his father also had palmoplantar keratoderma.

Physical examinations revealed symmetrical keratotic plaques on axilla, inguinal and buttock area with mild erythematous patches on the trunk. It was accompanied by symmetric palmoplantar keratoderma. A histopathological examination of the axilla and flank demonstrated hyperkeratosis and moderate acanthosis in the epidermis with mild perivascular lymphocyte infiltrate in the upper dermis. The diagnosis therefore depends on the clinical features and family history. EKV generally responds well to retinoids which restores the deficient keratinosome. Emollients, topical retinoid acid, intralesional steroids have also been used. The clinical differential diagnosis of EKV includes plaque psoriasis, CHILD syndrome, progressive symmetric erythrokeratoderma, and familial pityriasis rubra pilaris. The main feature distinguishing EKV from others is the transient lesions and possibly lack of facial lesions in most cases.

EKV is an extremely rare disease and so far, only two cases have been reported in Korea. However, one case was reported in child, the other from a foreigner. This study is the first case of EKV in a Korean adult. It's also worth

Received December 5, 2018, Revised March 2, 2019, Accepted for publication March 25, 2019

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Fig. 1. (A~D) Well demarcated, symmetric erythematous keratotic plaques on both axilla, trunk and buttock, and thigh. (E, F) Thick keratoderma on palm and soles are shown. We received the patient’s consent form about publishing all photographic material.

Fig. 2. (A) Marked hyperkeratosis and mild acanthosis with mild inflammatory infiltration in upper dermis. Mild increase in number of eccrine glands is shown (left axilla: H&E, ×40). (B) Close up view, compact hyperkeratosis, normal-appearing granular layer and irregular acanthosis in the epidermis with mild perivascular lymphocyte infiltration in the papillary to upper dermis (left axilla: H&E, ×200).

noting that the patient has a familial history and was treated successfully with retinoid. Therefore, dermatologists should keep in mind that EKV occurs rarely in Koreans with a favorable outcome using acitretin.

CONFLICTS OF INTEREST

The authors have nothing to disclose.
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