



Fatal Herpes Simplex Virus Infection in Darier's Disease

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Abstract

Darier disease is an autosomal dominant disorder characterized by small, hyperkeratotic papules localised on the trunk especially in the seborrheic regions. Vegetation and infections in the intertriginous areas are common in this disease while treatment is not usually satisfying. While the urea or lactic acid containing moisturizers, topical steroids, and topical retinoids are used in mild cases, systemic retinoids are preferred in severe cases. Kaposi varicelliform eruption caused by herpes simplex virus is rare but a well-defined complication. Clinical findings of this viral infection, which are usually uncommon, may delay diagnosis and treatment. Here, we report the case of a patient with Darier's disease treated with corticosteroids who later developed fatal herpes simplex virus infection despite the aciclovir therapy.

Key Words: Darier's Disease; Herpes Simplex Infection; Fatal Herpes Simplex Infection.

Darier Hastalığında Fatal Herpes Simpleks Virüs Enfeksiyonu

Özet

Darier Hastalığı, otozomal dominant kalıtımla geçen, klinik olarak gövdede ve özellikle seboreik alanlarda lokalize, küçük, hiperkeratotik papüllerle karakterize bir hastalıktır. Hastalıkta intertriginöz alanlarda vejetasyon ve enfeksiyonlar sıklıkla görülür. Tedavisi genellikle tatmin edici değildir. Hafif olgularda üre veya laktik asit içeren nemlendiriciler, topikal steroidler, topikal retinoidler kullanılırken, şiddetli olgularda sistemik retinoidler tercih edilir. Herpes simpleks virüsünün neden olduğu Kaposinin variselliform erüpsiyonu hastalığın nadir görülen fakat iyi tanımlanmış bir komplikasyonudur. Bu viral enfeksiyonun alışılmışın dışındaki klinik bulguları, sıklıkla tanıda ve tedavinin verilmesinde gecikmelere neden olabilir. Burada kortikosteroid tedavisi alan bir Darier hastasında takipleri esnasında gelişen ve asiklovir tedavisine rağmen fatal seyreden, herpes simpleks virüs enfeksiyonu sunulmaktadır.

Anahtar Kelimeler: Darier Hastalığı; Herpes Simpleks Enfeksiyonu; Fatal Herpes Simpleks Enfeksiyonu.

INTRODUCTION

Also known as keratosis follicularis or Darier-White disease, Darier's disease is an inherited disease characterised by keratinisation disorders with skin, nails, and mucous membrane involvement (1). Clinically, it manifests itself especially on the seborrheic areas of the skin like the forehead, scalp, nasolabial and retroauricular folds, chest, and back in skin colour or as yellow-brown, oily, wart-like papules. Changes in the nails provide important clues in the diagnosis. Longitudinal white and red lines, longitudinal ridges and grooves on the nails along with subungual hyperkeratosis are common signs practitioners often come across. The most pathognomonic indication is V-shaped grooves on the free edge of the nails. In addition to that, kaposi varicelliform eruption (KVE) caused by herpes simplex virus is a rare but well-recognized complication (2-4).

The unusual clinical manifestations of this viral infection cause delays in the diagnosis and treatment processes (2). This study presents a fatal case of herpes simplex virus infection in a Darier's disease patient taking corticosteroid therapy.

CASE REPORT

A 55-year-old female patient, who was also a Darier's disease patient for 30 years with a history of routine follow-ups, was admitted to our clinic with extensive lesions all over the body. The dermatological examination showed expansive erythematous, hyperkeratotic, and some of them verrucous, papules on the face, trunk, and extremities as well as eroded and extensively macerated plaques under the abdomen and around the groin. There were brown longitudinal ridges on the fingernails (Figures 1, 2, and 3).



Figure 1. Lesions on the trunk



Figure 2. Lesions on the nails



Figure 3. Lesions beneath the breasts.

The patient had been using 25 mg acitretin for the last 6 months. We learnt that she had not been on any other drugs. The patient reported that she had been given triamcinolone acetonide bulbs twice in the last two months due to the increase in her complaints although she added that the treatment did not work. The biopsy obtained from the pustular area on the back of the hand showed hyperkeratosis in the epidermis and dyskeratotic cells undergoing suprabasal decomposition. The results were consistent with Darier's disease. The routine laboratory tests did not show any further issues.

Upon observing widespread eczematous skin lesions, we started a systemic methylprednisolone 60 mg/day treatment. The samples obtained from the sharp-edged plaques with activation on the edges and the native preparation obtained from the tongue were considered positive, which urged us to start systemic and topical antifungal medication. In the follow-up sessions, we observed that the patient developed umbilicated vesicular lesions that could only be measured in millimetres. Apart from this, the Tzanck test was also positive. Therefore, we urgently applied 3x250 systemic acyclovir parenterally. The PCR results of the samples from the vesicular lesions on the trunk showed that HSV-1 was positive. The patient developed respiratory distress thus we started antibiotic treatment (imipenem + cilastatin 4x500mg, teicoplanin 1x400mg). Despite all the treatments we applied, the patient developed

systemic dissemination and we lost the patient due to acute respiratory distress syndrome.

DISCUSSION

Darier-White disease is an autosomal dominant inherited disease that is characterised by keratinisation changes in the skin and mucous membranes only. Especially hairline, temples, ears, and hairy scalp on the face and seborrheic areas like the chest, back, and flexures disclose skin coloured follicular or perifollicular, dirty yellow, reddish, brown hard papules which eventually turn to yellowish-brown extensive squams. Vegetation and infections are common around the intertriginous areas (5). In our case, too, the patient had superficial fungal infection on the intertriginous areas. Herpes simplex skin infections tend to be more common in Darier's disease (3, 4). Herpes simplex infections are generally self-limiting infections. However it may lead to severe muco-cutaneous tables, systemic involvement, and fatal results in immunocompromised patients (4, 7, 8). In our case, we observed lesions while applying the systemic steroid therapy.

Disseminated zoster HIV infection gives way to cellular immune system damages like hematopoietic stem cell and solid organ transplantation (3, 9).

Herpes simplex infections in Darier's diseases is very rare. Nikkels A.F. et al.'s Darier's patient on corticosteroids who developed fatal herpes simplex infection is one of the few reports in the literature (4).

We think that our case, in which the patient developed fatal herpes simplex infection while receiving steroid therapy, is worth sharing due to its rarity.

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