A Case of Darier's Disease Complicated with Kaposi's Varicelliform Eruption

Kaposi'nin Variselliform Erüpsiyonu ile Komplike Olmuş Bir Darier Hastalığı Olgusu

Nuran ALLI,[a,b]
Ahu YORULMAZ,[a]
Evşen ATICI,[a]
Düriye ÖZER TÜRKAY[c]

Clinics of
[a]Dermatology,
[c]Pathology,
Ankara Numune Training and Research Hospital,
Ankara
[b]Department of Dermatology,
Kafkas University Faculty of Medicine,
Kars

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Yazışma Adresi/*Correspondence:*
Ahu YORULMAZ
Ankara Numune Research and Training Hospital,
Clinic of Dermatology, Ankara,
TÜRKİYE/TURKEY
ahuyor@gmail.com

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ABSTRACT Darier's disease (DD) is a rare genodermatosis of autosomal-dominant inheritance which is characterized by flesh-colored to brownish hyperkeratotic papules and plaques with a greasy and warty texture. DD runs a life-long course with exacerbations and remissions. Despite the assumed benign nature of the disease, DD has been linked to a variety of other diseases and complications. Kaposi's varicelliform eruption (KVE) is a disseminated cutaneous infection which is generally caused by herpes simplex virus in the setting of underlying primary dermatosis such as atopic dermatitis, pemphigus vulgaris, psoriasis, contact dermatitis, and DD. Here we report a case of DD which is complicated with KVE.

Key Words: Darier disease; kaposi varicelliform eruption; herpes simplex

ÖZET Darier hastalığı (DH), deri rengi ile kahverengimsi renk arasında değişen, yağlı ve verrüköz görünümde hiperkeratotik papül ve plaklarla karakterize nadir rastlanan, otozomal dominant kalıtım gösteren bir genodermatozdur. DH, relaps ve remisyonlarla karakterize hayat boyu devam eden bir seyir gösterir. DH, benign doğasına karşın çok sayıda hastalık ve komplikasyonla ilişkilendirilmiştir. Kaposi'nin variselliform erüpsiyonu (KVE), atopik dermatit, pemfigus vulgaris, psöriazis, kontakt dermatit ve DH gibi altta yatan primer bir dermatoz zemininde, genellikle de herpes simpleks virüs nedeniyle ortaya çıkan dissemine kutanöz bir enfeksiyondur. Biz de burada KVE ile komplike olmuş bir DH olgusunu sunmak istiyoruz.

Anahtar Kelimeler: Darier hastalığı; kaposi variselliform döküntü; herpes simpleks

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Darier's disease (DD; keratosis follicularis) is a rare genodermatosis characterized by greasy hyperkeratotic papules and plaques mostly on seborrheic areas. It is a chronic disease with exacerbations and remissions and a typical symptomatology of itching, superinfections and malodour.[1,2] Kaposi's varicelliform eruption (KVE) is a widespread cutaneous infection generally caused by herpes simplex virus (HSV) arising upon pre-existing lesions of atopic dermatitis, Sezary syndrome, pityriasis rubra pilaris, pemphigus foliaceus, Hailey-Hailey disease, and DD.[3] Here we report a case of DD complicated with KVE in a 67-year-old woman.

CASE REPORT

A 67-year-old woman presented with 10 days history of fever, malaise, nausea, vomiting, itchy and painful eruption over trunk and extremities. She told that she had been followed for a diagnosis of DD for about 40 years without any complication. On trunk and extremities especially on seborrheic areas, flanks and sacrum there were numerous 0,2-1 cm erythematous to yellow-brown greasy papules, some of which were crusted. It was also noticed that some of the papules were umbilicated, and randomly admixed with occasional umbilicated pustules with erythematous base (Figures 1, 2). Examination also disclosed umbilicated vesicles merging to form plaques on bilateral inframammarial areas and hemorrhagic vesicles on intermammarial area (Figures 1, 3). The body temperature of the patient was above 38ºC and abnormal laboratory findings were as follows: erythrocyte sedimentation rate (ESR): 55 mm/h (0-20), C-reactive protein (CRP): 58 (0,2-5 mg/L), blood eosinophil percentage 21,9% (0,9-4), alanine aminotransferase (ALT) 208 IU/L (3-50), aspartate aminotransferase (AST) 127 IU/L (4-50), alkaline phosphatase 252 IU/L (30-120), gamma-glutamyl transpeptidase (GGT) 480 IU/L (6-55). As we suspected a diagnosis of KVE in our patient, we performed serological tests for HSV type 1 and 2. Accordingly, HSV-1 and HSV-2 IgG screens were positive, HSV-1 and HSV-2 IgM screens were negative. Her blood bacterial culture was negative. The patient was consulted to gastroenterology department because of elevated liver enzyme levels. The consultant doctor thought that the reason for elevated liver enzyme levels was atorvastatin which she had been recently prescribed for hyperlipidemia. Accordingly, atorvastatin treatment was stopped and the patient was given only a low cholesterol diet. Histopathological examination of the skin biopsy from hyperkeratotic papules revealed papillomatosis, parakeratosis, suprabasal acantholysis and superficial perivascular lymphocytic infiltrate with occasional eosinophils (Figure 4). Consequently, a diagnosis of DD complicated with KVE was made and the patient was given intravenous acyclovir (10 mg/kg every 8 hours). After 3 days as the symptoms of the patient had subsided,

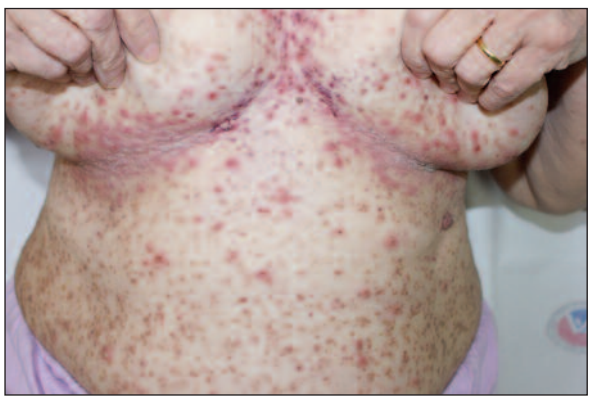

FIGURE 1: Widespread erythematous to yellow-brown warty papules with a greasy texture, a few number of pustules and umbilicated vesicles, some of which were hemorrhagic in nature coalescing to form plaques on inframammarial areas.
(See color figure at http://www.turkiyeklinikleri.com/journal/dermatoloji-dergisi/1300-0330/)

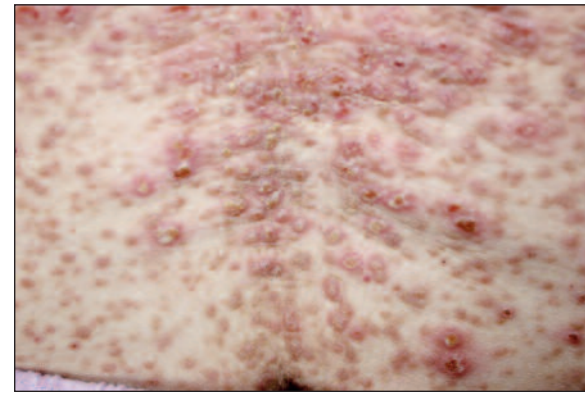

FIGURE 2: Crusted brownish papules and umbilicated erythematous vesicles and a few number of pustules on sacral area with a tendency to confluence.
(See color figure at http://www.turkiyeklinikleri.com/journal/dermatoloji-dergisi/1300-0330/)

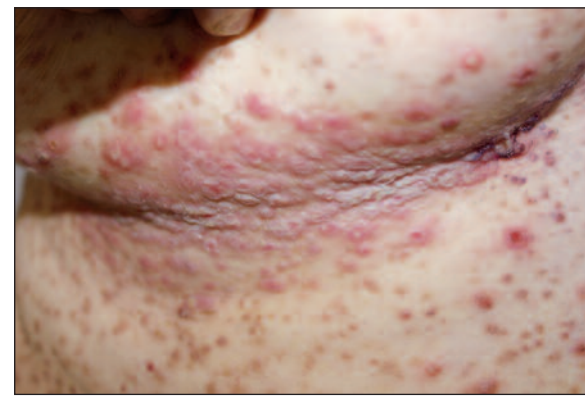

FIGURE 3: Cluster of umbilicated vesicles on left inframammarial area
(See color figure at http://www.turkiyeklinikleri.com/journal/dermatoloji-dergisi/1300-0330/)

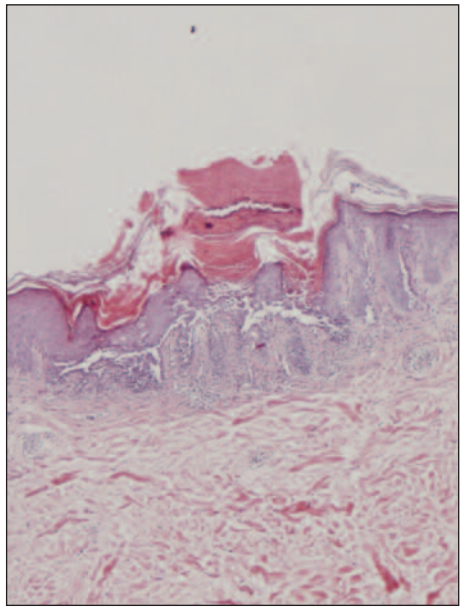

FIGURE 4: Papillomatosis, parakeratosis, suprabasal acantholysis and superficial perivascular lymphocytic infiltrate with occasional eosinophils (HE, x40).
(See color figure at http://www.turkiyeklinikleri.com/journal/dermatoloji-dergisi/1300-0330/)

oral regimen was established instead of intravenous route. At the 10 days follow up visit, the symptoms and the liver function tests of the patient were significantly improved.

DISCUSSION

DD, which is also known as keratosis follicularis, is a rare autosomal-dominant genodermatosis with high penetrance but variable expression.[1,2] Although clinical manifestations generally become apparent between the ages of 10 and 20 years, onset of the disease may be delayed.[2,4] DD is caused by mutations in the ATP2A2 gene, which encodes the sarco/endoplasmic reticulum calcium ATPase type 2 (SERCA2). SERCA2 is a calcium ATPase pump that has a central role in calcium transport and homeostasis.[2] DD is typically characterized by greasy, hyperkeratotic, flesh coloured to yellow or yellow-brown papules especially over seborrhoeic areas such as nasolabial folds, cheeks, forehead, hair margins, front and back of the chest, and flexures. In flexural sites the papules may coalesce into large, exophytic plaques forming papillomatous masses. On the other hand DD also has other distinctive clinical features like punctate hyperkeratotic papules on palms and soles, wart-like papules on dorsal aspects of hands and feet, nail involvement with longitudinal white or red streaks, V-shaped notch, subungual hyperkeratosis, and oral involvement with cobblestone appearing papules on palate, tongue or buccal mucosa.[1,2]

Secondary bacterial and viral infections are common complications of DD.[1,2] KVE, also known as eczema herpeticum, is a cutaneous eruption caused by HSV and other viruses such as coxsackie A16 and vaccinia. Although KVE most commonly occurs in patients with atopic dermatitis, it has also been described in various dermatoses like DD, pemphigus vulgaris, pemphigus foliaceus, toxic epidermal necrolysis, contact dermatitis, psoriasis, and lichenoid drug eruption.[3,5,6] The association between DD and KVE has been a topic of interest.[7] Even though it has not been fully described yet, an impaired barrier function of the epidermis and defect in cell-mediated immunity is suggested to be the stimulator of HSV replication.[4,8] Further studies are needed to define the exact pathogenesis of KVE. Here we present a case of DD complicated with KVE which is a serious association of DD.

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