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DARIER'S DISEASE ASSOCIATED WITH BASAL CELL CARCINOMA

DARİER HASTALIĞIYLA BAZAL HÜCRELİ KARSİNOM BİRLİKTELİĞİ

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ABSTRACT

Darier's disease (DD) is a rare, autosomal dominantly inherited genodermatosis characterized by keratotic papules predominantly on the face, forehead, scalp, chest and the back.

The disease does not seem to predispose to cutaneous malignancies. In an extensive literature review, only four patients were reported with Darier s disease and basal cell carcinomas (BCC). A relatively young patient with Darier s disease who developed basal cell carcinoma on the left cheek and treated by wide total excision is presented.

Key words: Darer's disease, basal cell carcinoma, surgical treatment.

ÖZET

Darier hastalığı yüz, alın, saçlı deri, göğüs ve sırttaki keratotik papüllerle karekterize nadir, otozomal dominant bir hastalıktır. Darier hastalığı kutanöz tümörlerin oluşumuna predispozan olarak görülmemektedir. Yapılan literatür araştırmalarında, Darier hastalığı ile bazal hücreli karsinomun birlikteliğinin sadece 4 olguda var olduğu saptanmıştır. Bu sunumda, yaşı nispeten genç olan ve Darier hastalığı ile sol yanağında bazal hücreli karsinomu olan, geniş eksizyonla tedavi edilen bir bayan hasta sunulmaktadır.

Anahtar kelimeler: Darier hastalığı, bazal hücreli karsinom, cerrahi tedavi.

INTRODUCTION

Darier s disease, also known as keratosis follicularis, is characterized by greasy hyperkeratotic papules in seborrheic regions, nail abnormalities, and mucous membrane changes. Abnormal keratinocyte-keratinocyte adhesion and aberrant epidermal keratinization are the primary histologic features of DD (1-3).

Multiple discrete, scaling, rough, crusted, pruritic skin papules that are frequently malodorous and disfiguring characterize DD. The diagnosis is based on the typical clinical appearance and histology showing parakeratosis, dyskeratotic cells known as corps ronds and grains and suprabasal acantholysis (3).

Although DD has been reported in association with other medical problems such as neuropsychiatric disorders, salivary gland obstruction, renal and testicular agenesis, bone cysts, it has not been found to be associated with cutaneous malignancies except for 4 reported cases of BCC (3,4,9). We report the fifth case of DD associated with BCC.

CASE

A 36 years-old female patient admitted to our hospital with the complaint of a tumoral mass on her left cheek for two years (Figure 1).

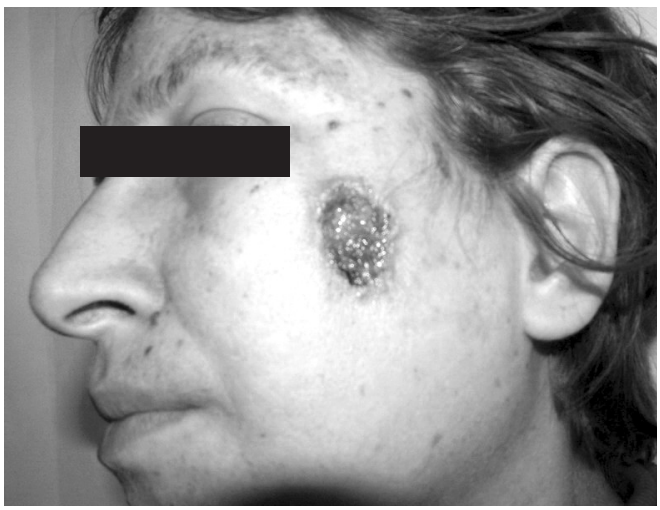

Figure 1. Preoperative appearance of the patient

Her past medical history was significant for multiple skin colored papules on her forehead, chin, scalp, neck, hands and forearms for 11 years (Figure 2). She had given no therapy for the papular lesions. She denied any past history of previous

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skin cancer, excessive sun exposure, radiation treatment, or arsenic exposure. There was no familial history for any similar disease or similar affection in parents/siblings.

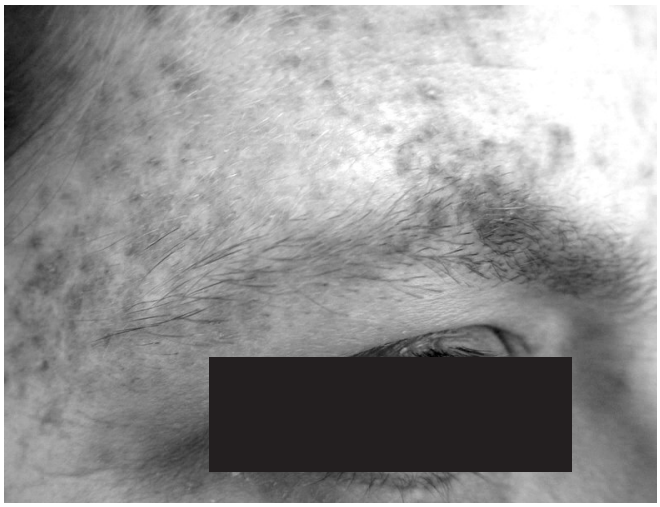

Figure 2. Note the skin-colored papules on the forehead

Dermatological examination revealed discrete, warty, greasy, skin colored papules with a brownish hue on her forehead, chin, scalp and neck. She also had similar lesions on her hands and extensor surfaces of distal 1/3 parts of forearms. There was a large, 3x2 cm ulcerated tumor with elevated borders on her left cheek. There were also several longitudinal white and red bands and onychoschizia on her hand nails. Pebbly white plaques (cobble stoning) and whitish papules with central umblications were evident on the gingiva and hard palate.
All routine laboratory investigations were normal.
The histopathologic examination of the tumor revealed masses of basaloid cells with pallisading of nuclei and the diagnosis was nodular basal cell carcinoma (Figure 3).

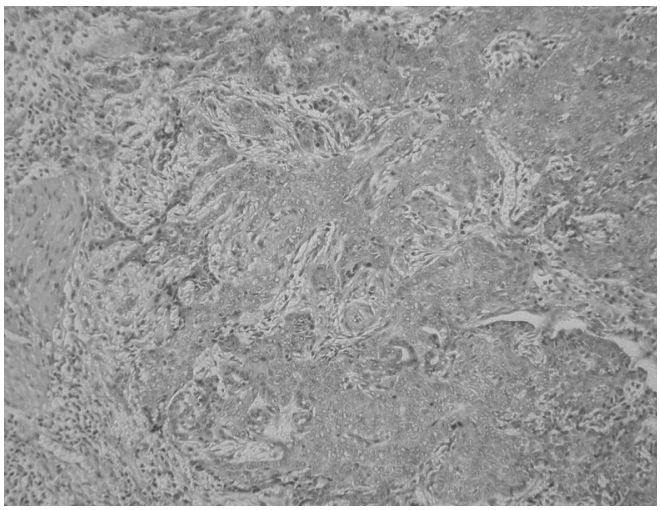

Figure 3. The tumoral area. The ulcerated tumor is composed of 'basaloid' epithelial cells, forming nests with peripheral palisading (HE x100)

Multiple foci showing typical features of Darier's disease found in the peritumoral skin were also reported (Figure 4).

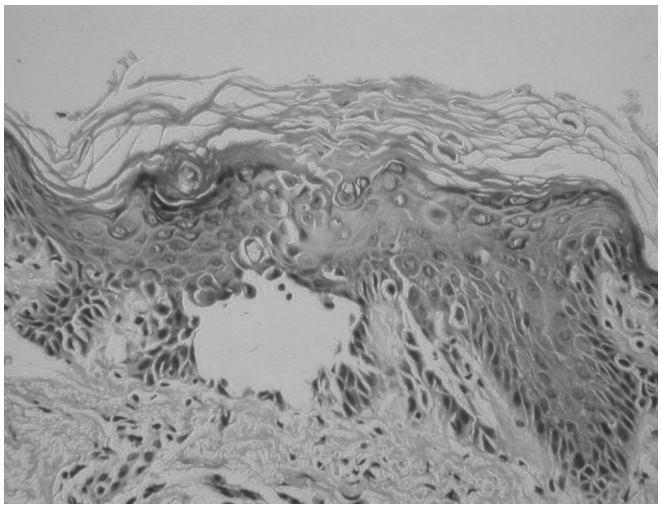

Figure 4. Foci of changes compatible with Darier's disease on the same excision. Parakeratosis, suprabasal acatholysis, 'corps ronds' and 'grains' (HE x200)

The patient was treated by wide total excision of the BCC on her left cheek, and discharged with systemic acitretin therapy for Darier's disease.

DISCUSSION

Genodermatoses are inherited disorders with dermatologic manifestations. Many of these disorders are autosomal dominant, and most are quite rare such as Darier's disease. Although some are associated with subsequent disease-related malignancies, Darier's disease is not associated with malignancy. Burge and Wilkinson reviewed the clinical features in 163 patients with Darier's disease and they had found no relationship between skin cancers and the dermatosis (1). In 1981, a patient with Darier's disease who developed several basal cell carcinomas was reported by Latour et al. (6). It was also documented that the patient had received superficial radiotherapy and grenz-ray therapy. Rapini and Koranda (8) reported in 1982 a patient with Darier's disease and skin cancer. The patient developed two basal cell carcinomas and there was no predisposing factors such as radiation therapy. In 1991, Hamadah and Grande (4) reported a patient with Darier's disease who had been treated previously with grenz-ray therapy. The patient had multiple basal cell carcinomas. The fourth case of Darier's disease with skin cancer was reported by Russo, Perez-Bernal and Camacho (9) in 1995, in which the patient had developed basal cell carcinomas that arose in skin lesions.
Though not clear, excessive sun exposure is unlikely to play an important role in associated skin cancer development, because patients with Darier's disease tend to avoid sun exposure due to the aggravating effect on their lesions (3,9). Alternatively the potentially carcinogenic effects of topically applied agents like retinoic acid or irradiation might be considered

(3,5,9). It was also documented that the immune system function might be deranged in Darier's disease. This disturbance can possibly play a role in cutaneous malignancy development (7).

Our patient had Darier's disease and a basal cell carcinoma without a history of potentially tumorigenic therapies and she had intact humoral and cell mediated immunity.

Since oral retinoids are effective in reducing papules of Darier's disease, they can also be considered beneficial for earlier detection of possible basal cell carcinomas which are hidden in the keratotic papules. Therefore, early excision of basal cell carcinomas can be achieved (9).

Although it is difficult to conclude to a cause to effect relation between Darier's disease and the basal cell carcinoma in that specific patient, the relatively young age points toward the necessity of close clinical surveillance for early tumor development. Because of the life-long course of Darier's disease and the rare occurrence of associated BCC, close follow-up and systemic retinoic acid treatment may be useful to control the disease and may serve to the early identification of associated cutaneous malignancies.

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