INTRODUCTION
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: Darier's disease, basal cell carcinoma, surgical treatment.

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).

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
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.

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. Pebble white plaques (cobble stoning) and whitish papules with central umbilications 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 palisading of nuclei and the diagnosis was nodular basal cell carcinoma (Figure 3).

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

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.
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.

REFERENCES