

## CASE REPORTS

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### DARIER'S DISEASE WITH WARTY DYSKERATOMA AND BASAL CELL EPITHELIOMA

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A 48-year-old man with warty dirty papules over the seborrhoeic areas, also had a nodule with a central keratotic crater over the right cheek. Biopsy of this nodule revealed features of Darier's disease, warty dyskeratoma and basal cell epithelioma. Even though Darier's disease and warty dyskeratoma are considered as distinct entities, the presence in the same lesion emphasizes the need for further studies on this association.

**Key Words :** Darier's disease, Warty dyskeratoma, Basal cell epithelioma

#### Introduction

Darier's disease is an autosomal dominant disorder of keratinisation.<sup>1</sup> Warty dyskeratoma is considered to be a benign tumour.<sup>2</sup> We are reporting the co-existence of these disorders along with basal cell epithelioma from the same lesion.

#### Case Report

A 48-year-old man presented with skin lesions of 6 years duration, consisting of multiple brown-black, warty, greasy, follicular and non-follicular papules, some of which also showed keratin plugs, mainly over the seborrhoeic areas. Few papules were also present over the dorsa of fingers, hands, forearms and the feet. Erythematous papules were present over the hard palate in a cobble-stoned distribution. Palms and soles showed keratotic papules and pitting. Red and white bands, and V shaped nicking at the free margin were noted on the finger nails.

There was a painless mobile, nontender nodule with a keratotic crater just above the upper portion of the right nasolabial groove. It had an erythematous, raised and rolled border.

Routine investigations were within normal limits. Exision biopsy of the nodule in the right nasolabial fold showed hyperkeratosis, suprabasal lacunae, corps ronds and grains in one area suggestive of Darier's disease and hyperkeratotic invagination filled with keratinous material and acantholytic cells. In the lower part of the invagination, numerous slender villi lined with a single layer of basal cells projecting inward into the invagination was seen in another area, suggestive of warty dyskeratoma. Features of basal cell epithelioma of the undifferentiated, well circumscribed type were also seen in the same section.

#### Discussion

Co-existence of Darier's disease and warty dyskeratoma has been previously reported.<sup>1</sup> Each of these diseases is believed to represent a distinct clinical entity in spite of their similarities histopathologically.<sup>2</sup> The

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occurrence of basal cell epithelioma with either of these two conditions has not hitherto been reported.

El-Gothamy and Kamel<sup>3</sup> have reported various ultrastructural changes in Darier-White disease. Lesional skin of these patients shows acantholysis, formation of vacuoles in the cytoplasm, breakdown of desmosome keratinocyte complexes and abnormal intracellular aggregation of keratin filaments.<sup>4,5</sup> These changes suggest that Darier's disease is a disorder of keratinocyte differentiation. However Burge et al<sup>6</sup> hypothesised that the ultrastructural changes may be secondary to damage by proteases like plasmin. The exact cause of cell dyshesion in this disease is yet to be elucidated. Many attempts have been made to correlate warty dyskeratoma with Darier's disease using ultrastructural and immunohistological findings. These studies have failed to demonstrate similarities between the two diseases. But in our case the changes were seen in a single lesion.

Szymanski in 1957, described warty dyskeratoma as "a benign cutaneous tumour that resembles Darier's disease microscopically."<sup>2</sup> Since both histological changes of Darier's disease and warty dyskeratoma occurred in the same lesion, we think these two conditions may be interrelated and the coexistence may not be a chance occurrence.

Basal cell epithelioma occurring either in Darier's disease or warty dyskeratoma has not been reported to date. Ackerman<sup>7</sup> in 1972 has described the presence of suprabasal cleft with acantholytic cells in the epidermis overlying basal cell epithelioma, whether dyskeratosis predisposes to the development of warty dyskeratoma and basal cell epithelioma or is a fortuitous common finding in these conditions, is conjectural.

The unique occurrence of the triad of Darier's disease, warty dyskeratoma and basal cell epithelioma has not been reported in earlier literature.

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