

Hyperkeratosis of the Nipple and Areola

Report of 3 Cases

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Background: To date, approximately 43 cases of hyperkeratosis of the nipple and areola have been reported, most of which have been sporadic.

Observation: We describe 3 patients with this dermatosis who were encountered in the outpatient clinic of the Department of Dermatology at the American Uni-

versity of Beirut Medical Center, Beirut, Lebanon, within a 1-year period.

Conclusion: Hyperkeratosis of the nipple and/or areola may be more common than what has been reported in the literature. We propose a revised classification for the condition.

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APPROXIMATELY 43 cases of hyperkeratosis of the nipple and areola have been reported in the literature.¹ Most cases have been sporadic. Recently, within a 1-year period between June 1999 and May 2000, 3 patients with this dermatosis were encountered in the outpatient clinic of the Department of Dermatology at the American University of Beirut Medical Center, Beirut, Lebanon. This observation may indicate that hyperkeratosis of the nipple and/or areola may be more common than previously thought.

REPORT OF CASES

CASE 1

A healthy 32-year-old woman presented with asymptomatic progressive thickening of the areola of the left breast occurring over a period of 9 years. On physical examination, a verrucous keratotic plaque was found involving the areola of the left breast (**Figure 1 A**). The left nipple as well as the areola and nipple of the right breast were normal. There was no evidence of warts, ichthyosis, or acanthosis nigricans. No family history of similar lesions was reported. Findings from histopathological examination showed profound compact hyperkeratosis and plugging with a slight to moderate degree of retiform hyperplasia of the epidermis (**Figure 1B**).

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CASE 2

A healthy 20-year-old woman presented with asymptomatic, progressive changes involving both nipples and areolae occurring over an 8-year period. On physical examination, thick hyperkeratotic, verrucous and darkly pigmented plaques were found involving both areolae and nipples (**Figure 2**). The patient was otherwise healthy and had no other skin lesions. On histopathological examination, hyperkeratosis, papillomatosis, retiform hyperplasia, and a relative increase in pigmentation of the basal cell layer of the epidermis were revealed.

CASE 3

A healthy 22-year-old woman complained of asymptomatic verrucous nipples of a 4-year duration. On physical examination, both nipples appeared hyperpigmented and keratotic. The areolae were normal. Warts, ichthyosis, or acanthosis nigricans were not present. A review of her family history for similar lesions was normal. Findings from histopathological examination showed areas of compact and basket-woven hyperkeratosis, focal parakeratosis, and mild papillomatosis of the epidermis.

COMMENT

The fact that 3 cases of hyperkeratosis of the nipple and areola have been encoun-

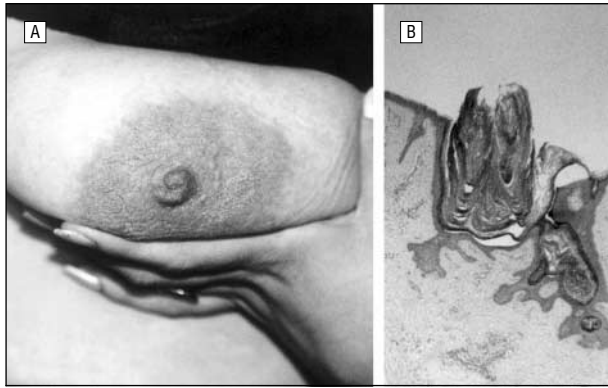


Figure 1. A, Verrucous keratotic plaque of the left areola. B, Keratotic plugs with hyperplasia of their lining.

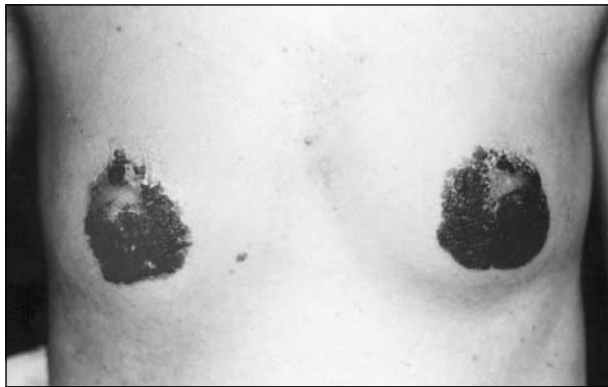


Figure 2. Involvement of nipples and areolae.

tered within a 1-year period indicates that this condition is not as rare as has been reported. Alternatively, it may represent a selection bias related to the tertiary level of care rendered at the American University of Beirut Medical Center, to which unusual cases are referred.

Several case reports in the literature²⁻⁴ refer to the classification of hyperkeratosis of the nipple and areola suggested by Levy-Franckel⁵ in 1938, who described the following 3 types: type 1, hyperkeratosis of the nipple and areola representing an extension of a verrucous (epidermal) nevus (this type is usually unilateral); type 2, hyperkeratosis of the nipple and areola associated with other dermatoses such as ichthyosis, acanthosis nigricans, Darier disease, and lymphoma (this type may be bilateral); and type 3, also known as idiopathic or nevoid hyperkeratosis of the nipple and areola, an unusual variant that appears predominantly in women in the second or third decade of life (this type is usually bilateral and according to some authors may appear at puberty,⁶ during pregnancy,^{7,8} and in men with prostate cancer treated with diethylstilbestrol⁹).

Considering the description of type 1 above, it is apparent that this is an epidermal nevus that happens to involve the nipple and/or the areola, and in this respect it should not be considered hyperkeratosis of the nipple and areola. Thus, the diagnosis of hyperkeratosis of the

nipple and areola would be reserved to the dermatosis when it occurs alone or is associated with ichthyosis, acanthosis nigricans, or occurs secondary to hormonal changes,⁶ an internal malignancy,⁹ or a lymphoma.^{4,10}

Although several articles²⁻⁴ refer to the condition as “nevoid hyperkeratosis of the nipple and areola,” one wonders whether the term *nevoid* is truly applicable. Consequently, we suggest that *nevoid* be deleted. As a matter of fact, *nevoid* has been used only in a few articles in the literature.^{6,11} Therefore, we propose the following alternative classifications: (1) primary hyperkeratosis of the nipple and areola (occurring coincidentally with other skin diseases, namely, disorders of keratinization such as ichthyosis, acanthosis nigricans, and Darier disease); (2) secondary hyperkeratosis of the nipple and areola (occurring secondary to hormonal changes⁶ or in the setting of an internal malignancy⁹ or lymphoma^{4,10}); and (3) idiopathic hyperkeratosis of the nipple and areola (occurring predominantly in women in the second or third decade of life, for which no obvious cause can be detected). Our 3 cases presumably belong to the last classification.

Response to topical treatments for hyperkeratosis of the nipple and areola, such as keratolytic therapy, cryotherapy, and retinoid therapy, is variable. Recently, carbon dioxide laser treatment has been reported to be effective.¹¹

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