CASE REPORT

Nevoid hyperkeratosis of the nipple and areola

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ABSTRACT

Nevoid hyperkeratosis of the nipple and areola (NHNA) is a rare condition. It is characterized by hyperpigmented verrucous plaques on the nipple and/or areola. Different opinions exist regarding its nomenclature, etiopathogenesis, treatment and whether it is a distinct entity. We report a 33-year-old female with unilateral NHNA and review the above mentioned aspects of the condition.

KEY WORDS: Nevoid hyperkeratosis of the nipple and areola, hyperkeratosis, nipple

INTRODUCTION

Nevoid hyperkeratosis of the nipple and areola (NHNA) is a rare, benign condition of unknown etiology characterized by verrucous hyperpigmented plaques on the nipple and/or areola that was first described in 1923. Since then many cases have been reported in the literature and some have argued whether it is a distinct entity. Since the pathogenesis is unknown and it is a rare dermatosis, specific treatment guidelines does not exist for this condition.

CASE REPORT

A 33-year-old healthy Kuwaiti female presented with a two-year history of hyperkeratotic and hyperpigmented lesions over her right nipple and areola. There was no history of pruritus, pain or discharge. She had recently delivered and was lactating but the lesions existed before pregnancy. She denied being on any medications. Past medical and family history was not significant. There was no history of atopy, warts, epidermal nevi, or ichthyosis. Basic laboratory and endocrinologic findings were normal.

Physical examination revealed brown papillomatous lesions on the right nipple as well as hyperpigmented verrucous plaques with black dots on the areola (Fig. 1). Punch biopsy specimen was taken from the areola which showed hyperkeratosis, filiform acanthosis, papillomatosis, keratinous plugs and keratin cysts (Fig. 2). The papillary dermis was slightly fibrotic. Therefore the diagnosis of NHNA was made. Liquid nitrogen cryospray was applied with partial improvement.

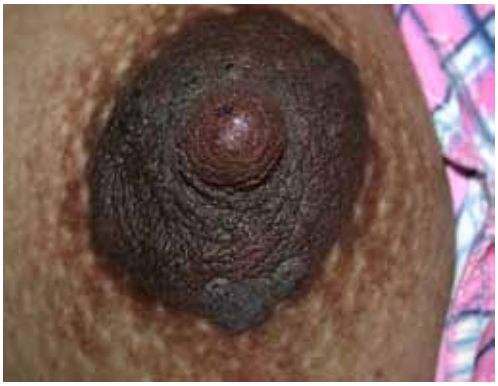

Fig. 1 Hyperpigmented verrucous lesions with diffuse involvement of the nipple and areola.

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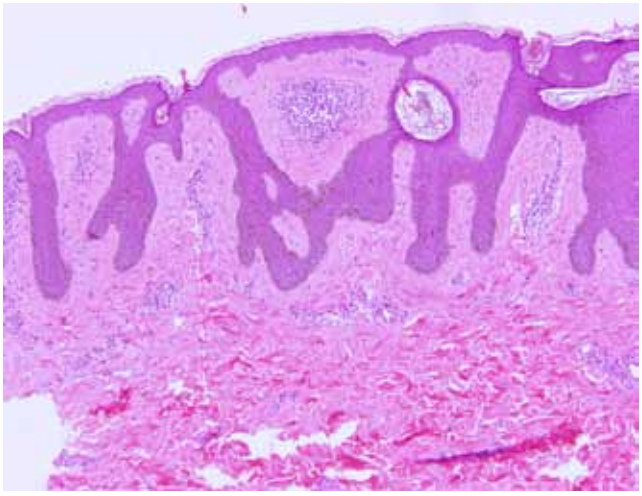

Fig. 2 Filiform acanthosis with extensive and marked elongation of the rete ridges and papillary dermis with mild papillary fibrosis and keratinous plugs.

DISCUSSION

Tauber first reported NHNA in 1923.[1] In 1938, Levy-Frankel described three distinct types of hyperkeratosis of the nipple and/or areola. Type 1 is hyperkeratosis as an extension of epidermal nevus. Type 2 is hyperkeratosis associated with other dermatoses such as ichthyosis, eczema, lymphoma, seborrheic keratosis, acanthsosis nigricans and Darier's disease. Type 3 is isolated nevoid form (NHNA).[2] The third type to which our patient also belongs occurs mainly in young women of childbearing age.[3] It is mainly bilateral but many unilateral cases like ours have been reported.[4,5]

The etiology of NHNA is not known. Pathogenic mechanisms such as a change in estrogen level have been proposed but not fully substantiated.[6] Clinically, it presents as hyperkeratotic or verrucous, hyperpigmented plaques of the nipple, areola or both. The histopathologic findings include hyperkeratosis, papillomatosis, filiform acanthosis, keratinous plugging and slightly fibrotic papillary dermis. Baykal et al reviewed the clinical and histopathologic features of 7 cases with NHNA and concluded that it is a distinct entity with distinct clinical and pathologic features. They also proposed criteria to distinguish it from seborrheic keratosis (SK), mainly that NHNA diffusely involves the nipple and areola but SK is characterized by sharply demarcated papules.[3] Others argue that it is not a distinct entity, but rather epidermal nevus or SK. We believe that according to Levy-Frankel classification those cases which are associated with other dermatoses such as epidermal nevi or SK are type 2 hyperkeratosis of the nipple and areola (HNA), but the nevoid form (NHNA) is a distinct entity and distinct from SK, etc. So, in this regard we agree with those who say that the diagnosis of the NHNA is a diagnosis of exclusion.[7]

The unknown etiology along with rarity of the disorder makes formulating specific treatment guidelines very difficult. However various treatment modalities have been reported in the literature. These include topical steroids,[8] tretinoin,[9] 6% salicylic acid gel,[10] other keratolytics,[11] calcipotriol,[12] oral etretinate,[13] cryotherapy,[14] radiofrequency,[15] carbon dioxide laser,[16] shave excision,[17] and surgery.[18] We treated our patient with cryotherapy with partial response.

The disorder is a benign one with good prognosis, but without treatment it persists indifintely.[19]

We report this case of NHNA for the rarity of the condition which we believe is under-reported because of lack of awareness.

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