Nevoid hyperkeratosis of the nipple mimicking a pigmented basal cell carcinoma

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INTRODUCTION

Nevoid hyperkeratosis of the nipple and/or areola (NHNA) is an uncommon skin disease without well-defined etiology, reported for the first time in 1923.[1] The disease is characterized by slowly growing verrucous thickening and brown pigmentation of the areola or nipple.[2] Here we report a lesion on the nipple clinically and dermoscopically identical to a small-size pigmented basal cell carcinoma. Nevertheless, histopathologic examination suggested the diagnosis of NHNA in a very early stage.

Abbreviation used:

NHNA: nevoid hyperkeratosis of the nipple and/or areola

CASE REPORT

A 30-year-old white woman presented with an asymptomatic, brownish-to-blue grayish lesion on her right nipple of 4 years' duration. The lesion measured 8 mm on the major axis and presented as an irregularly ovoid shape positioned around the base of the nipple (Fig 1). Physical examination did not find similar lesions elsewhere, and findings from a general objective examination were normal. Dermatologic history included a basal cell carcinoma in the lumbar region surgically removed 5 years before. Both dermoscopy and biopsy were performed. Dermoscopic examination found multiple blue-gray globules and leaflike areas (Fig 2) mimicking a pigmented basal cell carcinoma. Histopathology examination found a skin fragment covered by markedly thickened epidermis with mild papillomatosis; the basal layer showed hyperpigmentation without melanocyte proliferation, and mild fibrosis of the upper dermis. Remarkable proliferation of basal cells was observed (Fig 3), confirming a histopathologic diagnosis of NHNA.

Fig 1. Brownish, blue-grayish lesion around the base of the nipple.

DISCUSSION

NHNA is a rare and benign skin disease, which occurs predominantly in women of child-bearing age, especially during the second and the third decades of life.[3] Studies reported an associated worsening during pregnancy, supporting the hypothesis that NHNA might be a hormonal hyperkeratosis.[4] Moreover, to further support this

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Fig 2. Dermoscopy: multiple blue-gray globules **(a)** and leaflikes areas **(b)**.

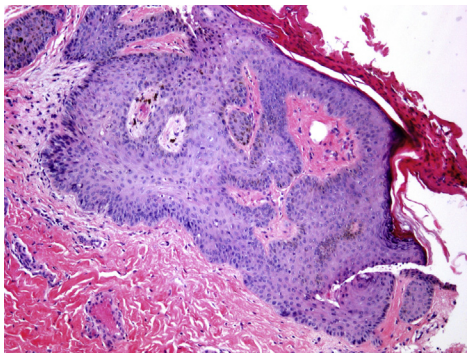

Fig 3. A fragment of skin covered with epidermis markedly thickened with mild papillomatosis. The basal layer shows hyperpigmentation without melanocyte proliferation. Mild fibrosis of the upper dermis and remarkable proliferation of basal cells are observed. (Hematoxylin-eosin stain; original magnification: ×10.)

hypothesis, men undergoing hormonal therapy can suffer from this same disease.[5]

The literature reports fewer than 50 cases and the etiology remains poorly understood. There are no consensus guidelines about the treatment; topical agents such as keratolytics, steroid, retinoids, or calcipotriol and ablative modalities like cryotherapy, carbon dioxide laser, radiofrequency, or shave excision are all potential treatment options.[6-8]

NHNA is usually characterized by brownish and verrucous thickening of the nipple or areola unilaterally or bilaterally, which has been classified into 3 types by Levy-Frenckel in 1938[9]:

- Type 1: associated with an epidermal nevus
- Type 2: associated with various dermatoses such as acanthosis nigricans, Darier disease, chronic eczema, and cutaneous T-cell lymphoma
- Type 3: isolated form with unknown etiology

The differential diagnosis of NHNA includes Paget's disease, basal cell carcinoma, seborrheic keratosis, melanoma, erosive adenomatosis, and hyperkeratosis secondary to prolonged friction.[10]

Considering clinical and histopathologic findings, our patient's disease is most consistent with NHNA type 3, although the clinical diagnosis was made very difficult by some aspects: (1) the lesion was in a very early stage when the clinical features typical of the disease were not yet evident and (2) dermatoscopic features of NHNA have not been previously described. NHNA observed in early stages can also show dermoscopic features mimicking a pigmented basal cell carcinoma, such as multiple blue-gray globules and leaflikes areas.

Some similarities between early-stage NHNA and pigmented basal cell carcinomas make differential diagnosis very challenging and can induce potential pitfalls. A definitive diagnosis can only be achieved through histopathology.

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