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Quiz
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Papules on the Nipples

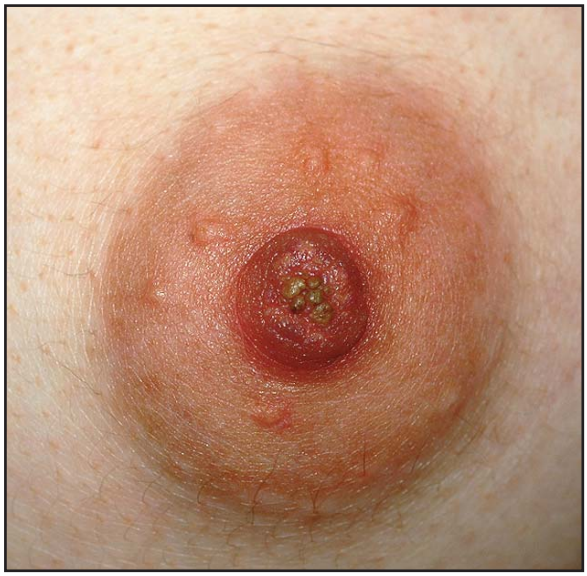

Figure 1a. Yellow-brown crust-like verrucous papules on nipple.

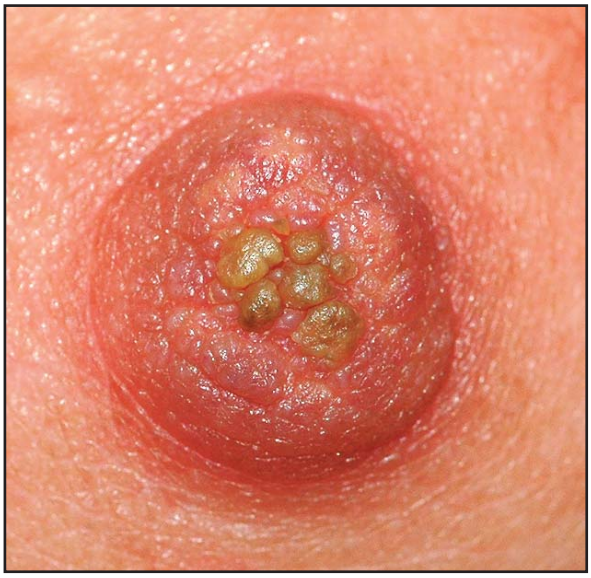

Figure 1b. Close-up view.

A 20-year-old woman presented with a 5-year history of irregular thickening of both nipples. On physical examination, both nipples were covered with yellowish-brown crust-like verrucous papules (**Figure 1a,b**). Lesions were usually asymptomatic but sometimes they cause slight discomfort due to friction of clothes. Her medical and familial history was unremarkable for breast malignancies and she had not been taken any medication or oral contraceptive.

What is the diagnosis?

Nevoid Hyperkeratosis of the Nipple and Areola

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Abstract

Observations: A 20-year-old woman presented with a 5-year history of irregular thickening of both nipples. On physical examination, both nipples were covered with yellowish-brown crust-like verrucous papules. Nevoid hyperkeratosis of the nipple and areola is characterized by irregular, verrucous thickening and yellow-brown hyperpigmentation of the nipple or/and areola. It may be unilateral or bilateral. It usually affects women in the second or third decade of life, especially during pregnancy.

Discussion

Nevoid hyperkeratosis of the nipple and areola (NHNA) is a rare condition. To the best of our knowledge, fewer than 70 cases have been reported in the literature after the first description was made by *Tauber* in 1938 [**1**]. NHNA is characterized by irregular, verrucous thickening and yellow-brown hyperpigmentation of the nipple or/and areola [**2**]. Involvement of nipple and areola may be unilateral or bilateral [**3**]. Due to its asymptomatic nature it is possible that some cases may be overlooked. It usually affects women in the second or third decade of life, especially during pregnancy [**4**]. It can also occur in males. The etiopathogenesis of NHNA is obscure. Endocrine factors have been proposed, because it may worsen in pregnancy and it has been associated with estrogen therapy [**5**].

Classification of NHNA has been made by *Levy-Franckel* in three categories: 1) an isolated or nevoid form (nevoid hyperkeratosis), 2) an epidermal nevus extension, 3) in association with ichthyosis, acanthosis nigricans, *Darier*'s disease, ichthyosiform erythroderma, T-cell lymphoma, chronic eczema [**6**]. The differential diagnosis of NHNA is a long list; epidermal nevus, *Paget*'s disease, acanthosis nigricans, seborrheic keratosis, chronic eczema, atopic eczema, *Darier*'s disease, basal cell carcinoma, dermatophytosis, and *Bowen*'s disease [**7**].

Main histopathologic characteristics of NHNA are prominent orthokeratotic hyperkeratosis, variable degrees of acanthosis, hyperpigmentation and filiform papillomatosis [**8**]. In some cases, sparse dermal inflammatory infiltrate may present. These features resemble those of epidermal nevus or acanthosis nigricans [**3**]. Our patient did not accept the biopsy procedure. But as in this case report, histopatological assessment is usually not necessary, since most of them are easily diagnosed with clinical presentation and personal history.

Management of NHNA is generally hard and unsatisfactory. Many therapeutic modalities, such as topical keratolytics (6% salicylic acid gel, 12% lactic acid lotion), topical corticosteroids, retinoid acid, calcipotriol, cryotherapy, surgery, shave exicision and carbon dioxide laser have been used, with varying results [**5, 6, 7, 8, 9, 10**].

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