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Nevoid Hyperkeratosis of the Nipple and/or Areola – A Report of 2 Cases

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Abstract: ***Nevoid hyperkeratosis of the nipple and/or areola (NHNA) is a rare but benign skin disease, which occurs predominantly in females during the 2nd and 3rd decade of life. Usually bilateral although unilateral has also been described. Naevoid or idiopathic, this is a distinct entity not secondary to any dermatoses. Though occurrence and aggravation around puberty and pregnancy support the hypothesis that the etiology is hormonal. Here we report two cases of NHNA without any of the hormonal predisposing factors thus making it a rare presentation of the condition.***

Keywords: Naevoid, hyperkeratosis, nipple, benign

Key Message: The eruption of such a malignant looking lesion over areola and/or nipple causes excessive concern in patients, thus its effective management with a decent cosmetic outcome is mandatory

1. Introduction

Nevoid hyperkeratosis of the nipple and/or areola (NHNA) is a rare skin disorder with an unknown etiology[1].It has been put under type 3 of hyperkeratosis of nipple and/or areola(HNA) according to the classical Lewy Franckel classification. Usually bilateral, though unilateral involvement has been described. The eruption of such a malignant looking lesion over areola and/or nipple causes excessive concern in patients, thus its effective management with a decent cosmetic outcome is mandatory[1].

We herein report bilateral NHNA in a 22-year-old female and unilateral in a 20 year old male.

2. Case Presentation

Case report 1: A 22-year-old non obese unmarried female presented to us with complaints of asymptomatic discoloration and thickening of the both areolas since two years [Figure 1]. It initiated as pink, scaling over the areola which gradually became darker.

On examination both nipples were dark diffusely thickened with a verrucous surface (right more than left). No abnormality (like nipple discharge, palpable mass, lymphadenopathy) was noted on examination.

Routine laboratory tests were within normal limits.

The skin biopsy specimen showed hyperplasia of epidermal lining with hyperkeratosis, acanthosis and marked elongation of rete ridges. Follicular plugging and keratin filled ostia noted. Superficial dermis shows mild perivascular lymphocytic infiltration. [Figure 2].

She was injected with triamcinolone on the left and triamcinolone plus 5-FU on the right. She is being called every 21 days for the same.

Case report 2: A 20 year old non obese male presented with complaints of thickening and discolouration of the right areola since 1 month gradually progressive in nature. It was not associated with any itching or pain. [Figure 3].

On examination the right nipple and areola were hyperpigmented and had a hyperkeratotic verrucous surface. There were no other abnormal findings similar to the first case. The left breast was normal on examination.

Routine laboratory tests were within normal limits.

Histopathologically, it showed hyperkeratosis, filiform acanthosis, papillomatosis and keratin plugging with mild lymphocytic infiltration in dermis.

He was suggested topical application of mometasone in the morning and retinoic acid at bedtime. Post one month of therapy improvement was noted but the patient has been lost to follow up.

After clinical and histopathological evaluation both our patients can be labelled as NHNA.

3. Discussion

NHNA is a rare but benign skin disease, which occurs predominantly in females during the 2nd and 3rd decade of life. It has an uncertain etiology and hence its treatment is challenging with a variable outcome. It is characterized by hyperpigmented verrucous thickening of the nipple and/or areola present unilaterally or bilaterally.

HNA was classified into 3 types by Levy-Frenckel in 1938:[2, 3]

- Type 1: Associated with epidermal nevus.
- Type 2: Associated with various dermatoses for eg acanthosis nigricans, , chronic eczema, cutaneous T-cell lymphoma, Darier's disease,
- Type 3: Isolated form with an unknown etiology.

Pérez-Izquierdo *et al.* [4] suggested an alternative classification of two types:
1 idiopathic or naevoid (unilateral or bilateral);

2 secondary, divided into:

i local (unilateral or bilateral): acanthosis nigricans, verrucous naevus, or seborrhoeic keratosis
ii systemic (bilateral): dermatosis, ichthyosis, malignant lymphomas, Darier's disease, chronic eczemas
iii drug related: diethylstilboestrol, spironolactone.
Another classification proposed by Mehanna *et al.* [5] includes a suggestion that the term 'naevoid' should be replaced by'idiopathic'.

Naevoid or idiopathic, this is a distinct entity characterized by verrucous thickening and brownish discolouration occurring over nipples and areolae not secondary to any other dermatoses[2]. Though occurrence and aggravation around puberty and pregnancy support the hypothesis that the etiology is hormonal.[1]

Hyperkeratosis of the nipples may coexist with ichthyosis, ichthyosiform erythroderma, acanthosis nigricans, Darier's disease (in association with other skin lesions, but also described as an isolated presenting phenomenon) [6], T-cell lymphoma[7, 8], and chronic graft-versus-host disease [9]. It has also been described in men suffering from prostate carcinoma on hormonal treatment [10].An appearance resembling verrucous naevi around both areolae has been described as a manifestation of inadequate hygiene.

In both our patients NHNA was seen without any of the hormonal predisposing factors thus making it a rare presentation of the condition.

Differential diagnosis of NHNA are :Acanthosis Nigricans, in which involvement of flexoral areas, coexisting endocrinopathies or obesity help in ruling out NHNA; epidermal nevus may be differentiated by its early age of onset; seborrheic dermatitis by its well demarcated velvety plaques over the areola instead of diffuse pattern of involvement. NHNA on biopsy may be mimicked by the above-mentioned conditions, histopathological differentiation can be made with the help of diagnostic clues such as filiform downward acanthosis and anastomizing rete ridges seen in NHNA.[1]

Management options for naevoid hyperkeratosis include topical retinoids[4], topical steroids[11], topical calcipotriol (particulary effective if associated with acanthosis nigricans, cryotherapy, carbon dioxide laser, shave excision, tangential excision using radiofrequency, and areola reconstruction with a skin graft.

History, physical examination, and the skin biopsy helped us rule out malignant conditions such as pagets disease. We counselled our patients about the benign nature of the disease and explained that the disorder is not half as threatening as it looks.[1]

Mainly what bothers patients is the cosmetic appearance of the nipple and areola. Patients should be told to be alert for any changes in the breasts. The course and progression must be explained as lesions may show recurrence and later resistance to treatment.

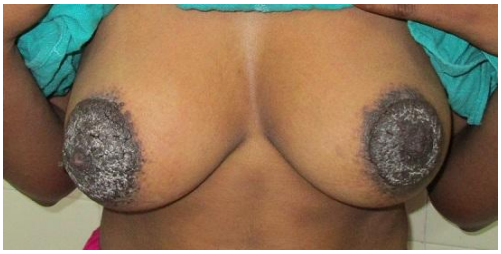

Figure 1: Both nipples were dark diffusely thickened with a verrucous surface (right more than left)

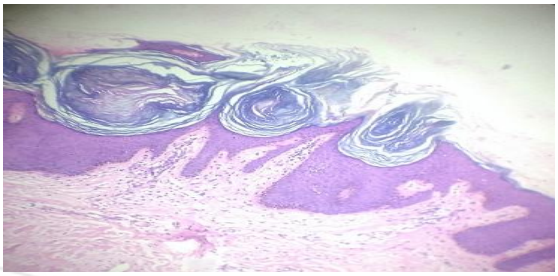

Figure 2: Histopathological picture showing hyperplasia of epidermal lining with hyperkeratosis, acanthosis and marked elongation of rete ridges. Follicular plugging and keratin filled ostia noted.

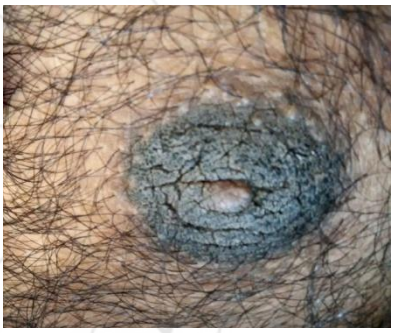

Figure 3: The right nipple and areola hyperpigmented with a verrucous surface.

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