Bilateral idiopathic hyperkeratosis of the nipple and areola

H. Akkari, L. Boussofara, W. Saidi, N. Ghariani, B. Sriha, C. Belajouza, M. Denguezli, R. Nouira

KEY WORDS

hyperkeratosis of nipple and areola, nevoid hyperkeratosis, tretinoin

ABSTRACT

Hyperkeratosis of the nipple and areola (HNA) is an unusual dermatosis that Levy-Franckel classified into three variants. This rare condition occurs primarily in young women and represents a cosmetic problem. Furthermore, its management is a therapeutic challenge. We report a rare case of a 32-year-old woman with idiopathic bilateral HNA, which belongs to the third Levy-Franckel classification.

Introduction

Hyperkeratosis of the nipple and areola (HNA) is an unusual dermatosis that occurs primarily in young woman and represents a cosmetic problem. Furthermore, its management is a therapeutic challenge. We describe a case of this rare breast dermatosis and discuss its clinical features, histology, and treatment options.

Case report

A 32-year-old unmarried woman presented with a 3-year history of bilateral thickening of the nipple and areola. Her family history and prior medical history were unremarkable. There was no history of pruritus, pain, or bleeding from the lesions and she had not been taking any medications. Physical examination revealed papillomatous thickening and a papular warty surface of the nipple and areola (Figs. 1a, 1b). There was no discharge from the nipple or tenderness on breast palpation. There were no other skin lesions such as warts, epidermal nevus, ichthyosis, or acanthosis nigricans. All baseline laboratory investigations were unremarkable. The mammogram and sonogram showed no abnormalities. The skin biopsy of the lesion revealed papillomatosis, acanthosis, and moderate hyperkeratosis of the epidermis. There was a mild lymphocytic infiltrate in the dermis. There was no evidence of premalignant or malignant changes (Figs. 2, 3).

Based on clinical and histological findings, the diagnosis of idiopathic HNA was made. The patient was treated with topical tretinoin 0.05% cream, showing no response after 2 months. Thereafter, acitretin was given at a dose of 25 mg/day with no improvement. Subsequently, surgical excision was recommended.

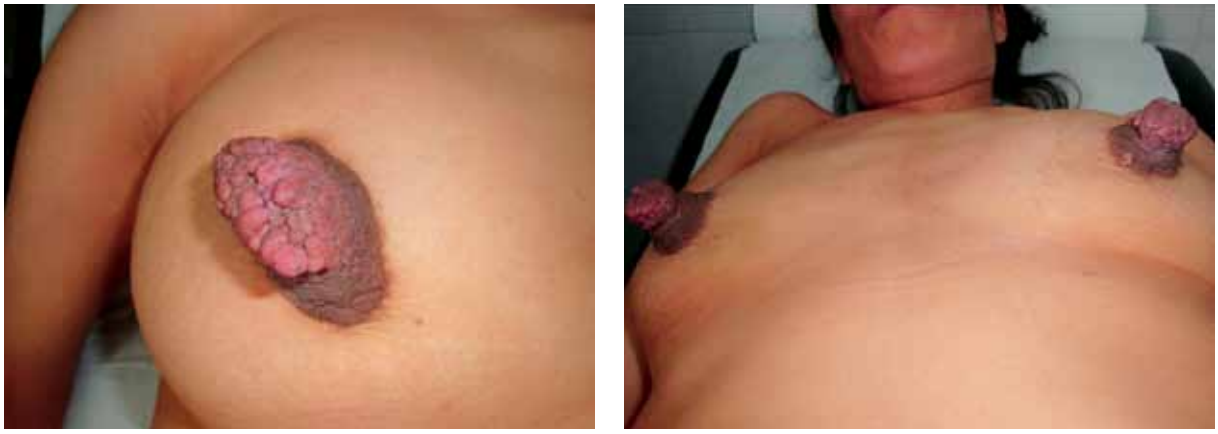

Figure 1 (a–b). Verrucous thickening and papular warty excrescences of the nipple and areola.

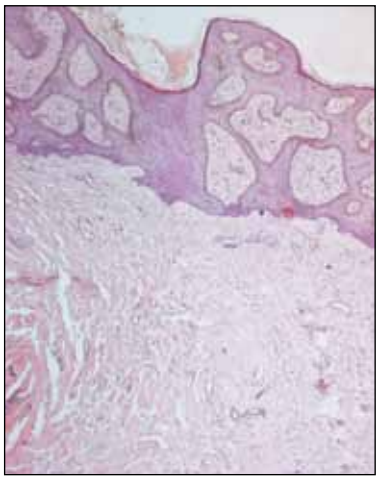

Figure 2. Papillomatosis, acanthosis, and moderate hyperkeratosis of the epidermis.

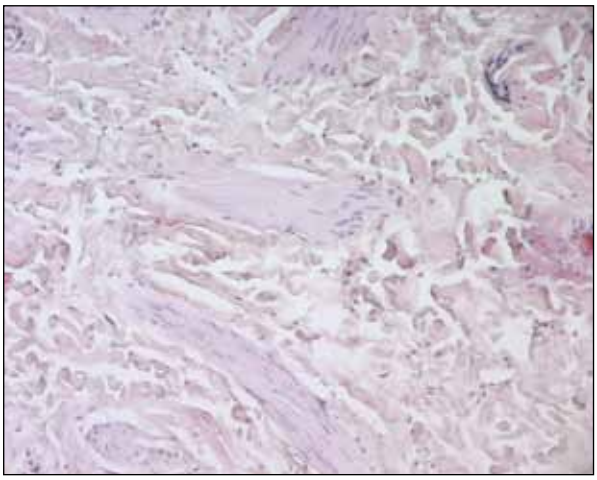

Figure 3. Absence of abnormalities of the deeper breast parenchyma.

Discussion

HNA is an uncommon dermatosis that has been divided by Levy-Franckel into 3 categories (1, 2). The first type is due to the extension of an epidermal nevus. The second group is associated with other dermatoses: chronic eczema, ichthyosis, acanthosis nigricans, or lymphoma. The third (previously called "nevoid form") is the idiopathic variant. In a report by Pérez-Izquierdo et al., HNA was classified into two variants, idiopathic or nevoid, and associated with other dermatoses or systemic diseases (3).

Idiopathic HNA occurs predominantly in woman in the second or third decade of life (1, 4). It is usually bilateral as in our case, but it may be unilateral. The etiology and pathogenesis of this condition are still unknown (2, 4). It has been thought that it may be correlated with endocrine factors because the condition may worsen in pregnancy and it has been associated with estrogen therapy (5). Clinically, lesions are typically characterized by chronic verrucous thickening of the nipple and/or areola. They may also appear as hyperkeratotic plaques (2). Clinical differential diagnosis is essential with consideration of type 1 and type 2 HNA, acanthosis nigricans, warts, and Paget's disease (1, 2). Our patient developed these lesions after puberty and had no association with epidermal nevi or other dermatoses. She was also free of symptoms associated with endocrinopathy or systemic diseases. It should be noted that the papillomatous appearance in our case posed the problem of differential diagnosis with florid papillomatosis. This was ruled out, however, because of the bilateral involvement of the nipple and areola, the absence of bleeding, excoriation, or swelling, and the histopathology results.

Histopathological features of HNA include orthokeratotic hyperkeratosis, papillomatosis, and mild acanthosis in the epidermis and perivascular lympho-

cytic infiltrate in the dermis (2). These findings can mimic epidermal nevus or acanthosis nigricans. This is why the diagnosis of HNA must be made based on both clinical and histological features.

Although the lesion is usually asymptomatic, its undesirable appearance may create a real psychological problem, especially for young woman. Moreover, treatment is usually unsatisfactory. Various therapeutic agents have been used including topical keratolytics, emollients, corticosteroids, and carbon dioxide laser and surgical excision, all with varying results (2, 4). It has been reported that systemic etretinate was ineffective in one case (6) but Okan et al. reported good cosmetic results with 1 to 2 months of continuous and then intermittent topical tretinoin (7). In our case no change was observed after both topical tretinoin and systemic retinoid therapy.

Idiopathic HNA is a benign, unusual condition. Because of the rarity of this dermatosis, there is a lack of awareness regarding it among many doctors. Similar cases need to be studied in order to better clarify the pathogenesis and therapy of this disorder.

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AUTHORS' ADDRESSES

Hayet Akkari, M.D;Department of Dermatology, Farhat Hached Academic Hospital, Avenue Ibn Jazzar 4000 Sousse, Tunisia

Lobna Boussofara, M.D; same address, corresponding author, e-mail: boussofara_l@yahoo.fr

Wafa Saidi, M.D; same address

Najet Ghariani,"Pr; same address

Badreddine Sriha, Pr; Department of Anatomocytology Farhat Hached Academic Hospital, Avenue Ibn Jazzar 4000 Sousse, Tunisia

Colandane Belajouza, Pr; Department of Dermatology, Farhat Hached Academic Hospital, Avenue Ibn Jazzar 4000 Sousse, Tunisia

Mohamed Denguezli, Pr; same adress

Rafiaa Nouira Pr; Head of the Department of Dermatology; same address