A patient with reticular pigmentation

Iran J Dermatol 2013; 16: 42-44

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Conflict of interest: none to declare

Received: 22 April 2012
Accepted: 10 August 2012

A 38- year-old man in good general health and normal intellectual functioning referred to the dermatologic clinic with numerous pruritic reticulate pigmentations on his neck, chest and upper back since 10 years ago. We noticed similar lesions on his axillae, groin and infra mammary area with no family background. Upon examination, symmetrical multiple small, rounds, dark brown freckle -like macules on his flexural surfaces and pruritic papules with a smooth surface on his chest were observed (Figure 1,2). Hair and nail were normal. Potassium hydroxide smear was negative. The patient was previously treated with topical hydroqinone, tretinoin, adapalene and corticosteroids with no response. A skin biopsy was preformed.

What is the diagnosis?

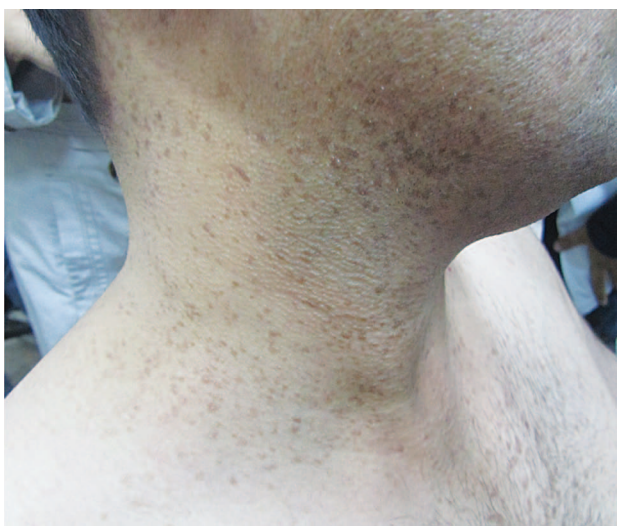

Figure 1. Pigmented and erythematous macules and papules in the neck region

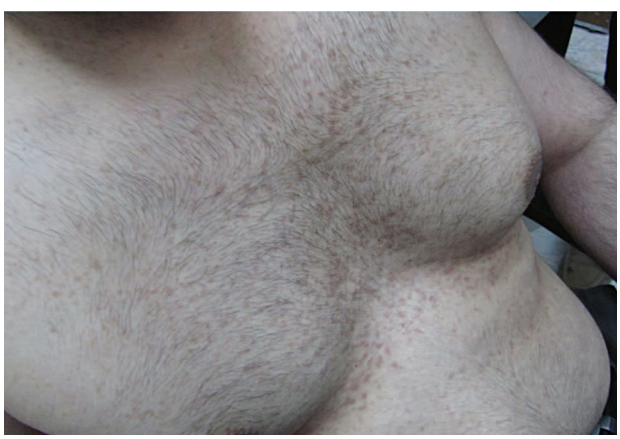

Figure 2. Pigmented and erythematous macules and papules on the chest.

Diagnosis

Dowling-Degos Disease

Microscopic findings

The epidermis showed elongated rete ridge (digitated) toward the dermis, hyperpigmentation of the basal layer with a perivascular lymphohistiocytic infiltrate in the dermis, and occasionally pseudo-horn cysts (Figure 3).

DISCUSSION

Reticulate pigmented anomalies include entities such as Dowling- Degos Disease (DDD), Galli Galli disease (the acantholytic variant of DDD), Habers syndrome (rosacea like facial eruption plus the clinical feature of DDD), Reticulated acropigmentation of Kitamura (atrophic acral lentigo-like lesions, palmoplantar and dorsal phalangeal pitting), acanthosis nigricans and lentigines in neurofibromatosis type 1 regarding to clinical or histological features [1,2].

Dowling-Degos disease was described by Wilson-Jones and Grice in 1978 as a reticulate pigmented anomaly of the flexures [3]. Dowling- Degos disease (DDD) is a pigmentary disorder usually due to loss of function mutations in Keratin 5 gene 1 [4]. The onset of this rare genodermatose is typically during the third to fourth decade of life with multiple small, round pigmented macules that resemble freckles. Pigmentation is slowly progressive and initially affects the axillae and groin and later involves the intergluteal and infra mammary folds, neck and inner aspects of the arms and thighs; other body folds may also become involved [5]. Occasionally, the pigmentary change can be palpable and some patients are described as having seborrheic keratosis, wart-like or erythematous papules [5]. Nails and hair are normal. Other features such as perioral pitted scars and comedone-like lesions on the upper eyelids, back and neck are described in about one-third of the patients. Involvement of the genitalia, particularly pigmented lesions of the vulva, has been described [5,6]. Dowling-Degos disease may be associated with hidradenitis suppurativa [7].

Genders are equally affected and there is no racial predilection as it has been reported worldwide. The histology is diagnostic, with a distinctive form of acanthosis, characterized by an irregular elongation of thin branching rete ridges, with concentration of melanin at the tips. The condition involves the follicular infundibulum, and there is follicular plugging in some cases. The melanocyte count is normal [8,9].

In differential diagnosis, Galli-Galli disease has the same clinical and histologic features as seen in DDD, with the exception of the presence of suprabasal non-dyskeratotic acantholysis in biopsy specimens of the lesional skin. Haber's syndrome is characterized by similar skin lesions and a photosensitive rosacea-like facial eruption that develops during adolescence. In reticulate acropigmentation of Kitamura, there are an increased number of melanocytes and pigment

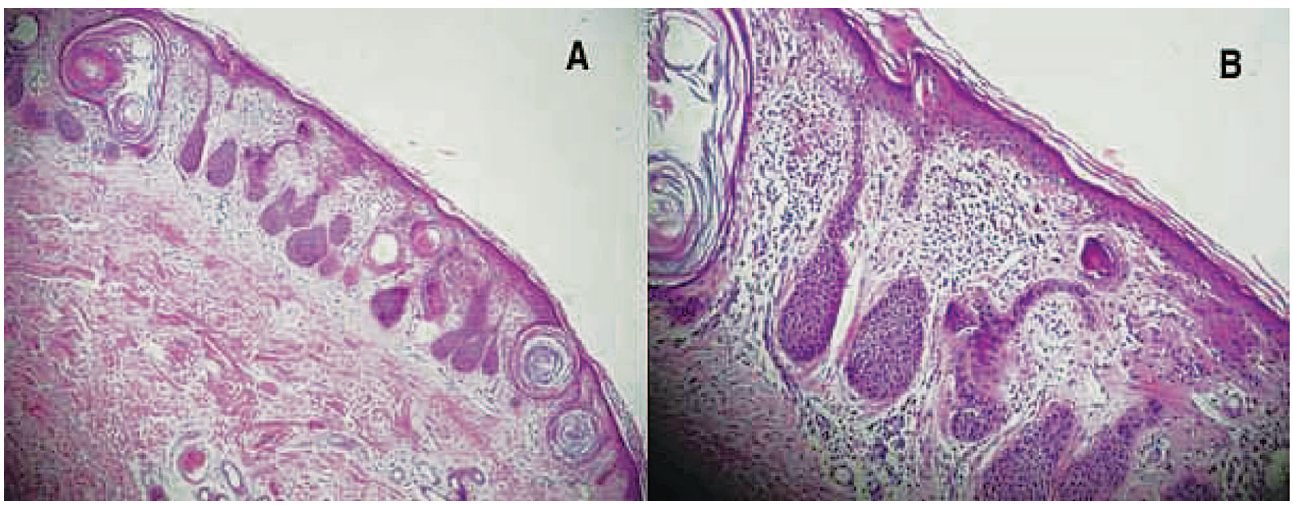

Figure 3. Histopatological features. A, B. Finger like rete ridges with increased pigmentation of the basal layer and thinning of the suprapapillary epidermis associated with tiny cysts or dilated follicles (H&E 40 and 100 respectively).

change is found on the hands and feet [1-6]. Acanthosis nigricans, which might be confused with DDD very early in the disease process, must be distinguished clinically by velvety plaques [10]. Patients with neurofibromatosis 1 develop lentigines in the axillae and groin which makes it distinguishable from DDD. Topical hydroquinone, tretinoin, adapalene, and corticosteroids have been used with varying degrees of success. Successful treatment with the erbium:YAG laser has been reported [1].

Our case showed reticulate hyperpigmentation of the flexures without other characteristic findings such as perioral scars and comedo-like lesions. We recommended laser treatment but he did not accept the suggested treatment.

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