Case for diagnosis

Caso para diagnóstico

Clarissa Barlem Hohmann [1]
Renan Rangel Bonamigo [3]
Cristiane Almeida Soares Cattani [5]
Bruna Köche [2]
Sérgio Torres Dornelles [4]

HISTORY OF THE DISEASE

A 53-year old, previously healthy, male patient from Porto Alegre, Rio Grande do Sul, Brazil complained of pruritus over the previous six months in the armpits and bilaterally in the groin, together with a progressive darkening of the skin, principally in the flexures, a condition that initiated 20 years previously. He also complained of a painful lesion on the upper surface of his right foot. He reported that his maternal grandfather, mother, aunt and daughter all had similar dermatological conditions.

Dermatological examination revealed diffuse hyperpigmentation of the face, comedones on the face and trunk, pitted, punctiform scars on the nose (Figure 1) and over the bilateral malar regions, and a cystic lesion of approximately 1.5 cm on the left side of his jaw. Confluent hyperchromic macules were found over extensive areas, as well as the formation of velvety plaques in the armpits and groin (Figure 2) and a brownish plaque on the back. On the upper part of the right foot, a verrucous plaque with a tumoral appearance was found (Figure 3).

The skin lesions on the trunk and armpits tested negative for fungal infections. Histopathology of the skin lesion in the right armpit showed acanthosis, with hyperpigmentation of the basal layer and formation of keratin cysts (Figure 4). Histopathology performed on tissue from the dorsal foot lesion revealed a keratoacanthoma. Serology for hepatitis B and C and for HIV was nonreactive. Other routine laboratory tests were normal. Treatment was initiated with topical retinoids, applied over the entire area affected except for the lesion on the foot, which regressed spontaneously.

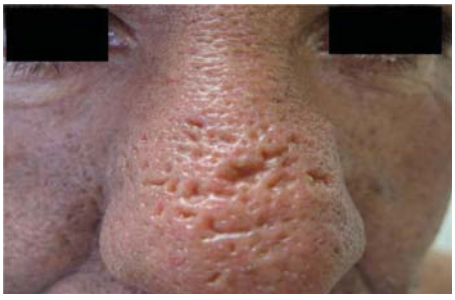

FIGURA 1: Cicatrizes deprimidas no dorso nasal

FIGURA 2: Placa hiperpigmentada "aveludada" na região inguinal

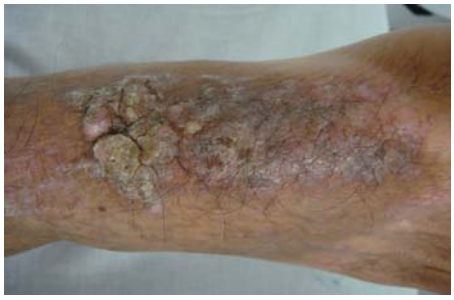

FIGURA 3: Placa verrucosa com aspecto tumoral na face dorsal do pé direito

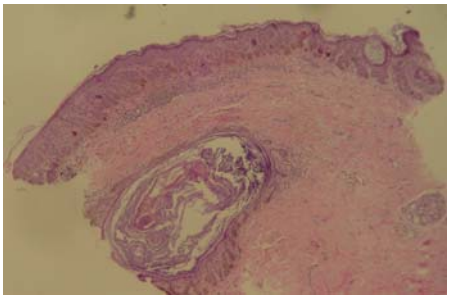

FIGURA 4: Acantopapilomatose, padrão digitiforme das papilas dérmicas, hiperpigmentação da camada basal e formação de cistos córneos (HE, 25x)

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[1] Resident Physician in Dermatology, Public Health Dermatology Outpatient Clinic, Porto Alegre, RS, Brazil.
[2] Resident Physician in Dermatology, Public Health Dermatology Outpatient Clinic, Porto Alegre, RS, Brazil.
[3] Dermatologist, Preceptor of the Residency Program in Dermatology, Public Health Dermatology Outpatient Clinic, Porto Alegre, RS, Brazil. Adjunct Professor of Dermatology and Professor of the Postgraduate Program in Pathology, Federal University of Health Sciences of Porto Alegre (UFCSPA), Porto Alegre, RS, Brazil.
[4] Dermatologist. Masters degree in Clinical Medicine from the Federal University of Rio Grande do Sul (UFRGS). Preceptor of the Residency Program in Dermatology, Public Health Dermatology Outpatient Clinic, Porto Alegre, RS, Brazil.
[5] Dermatologist. Preceptor of the Residency Program in Dermatology, Public Health Dermatology Outpatient Clinic, Porto Alegre, RS, Brazil.

COMMENTS

Dowling-Degos disease is a rare genodermatosis that is inherited in an autosomal dominant manner with varying degrees of penetration, in which there is a mutation in the keratin-5 gene. [1,2] It is characterized clinically by reticular hyperpigmentation in the flexures, such as in the neck, armpits, antecubital fossae, the area under the breasts and in the groin. Additional findings include pitted perioral scars in patients with no previous history of acne and hyperkeratotic comedone-like follicular papules in the neck and axillary regions. The most frequently found histopathological characteristics are acanthosis and papillomatosis, with dermal papillae in a digitiform pattern, keratin cysts in the epidermis and dilated keratinized hair follicles. Epidermal atrophy may also be found. Diffuse deposits of melanin are found in the basal layer and varying quantities of melanophages in the papillary dermis, as well as a superficial perivascular infiltrate of lymphocytes.

The disease is often associated with other dermatoses such as epidermal cysts, keratoacanthoma, epidermoid carcinoma, abscess, hidrosadenitis, seborrheic keratosis and pilonidal cysts, which in many cases constitute the principal reason for the patient to seek medical help. [3,4]

Despite the lack of any consensus regarding the classification of hereditary pigmentary dermatoses, it was recently agreed that Dowling-Degos disease is a clinicopathological entity that has several variants including the acantholytic form referred to as Galli-Galli disease and the acral form referred to as reticulate acropigmentation of Kitamura. [1,2,5]

The present paper reports the case of a patient with clinical and histopathological characteristics of Dowling-Degos disease. In addition, the appearance of a verrucous lesion on one of the patient's feet years after onset of the disease was particularly noteworthy. This lesion was diagnosed as a keratoacanthoma at histopathology. This tumor has been described in association with Dowling-Degos disease and the case in question corroborates this association. [3]

With respect to the treatment of Dowling-Degas disease, the therapeutic options are frustrating and results are poor. A partial response may be obtained in some patients with the use of topical retinoids or azelaic acid. [5] In addition, there have been reports in the literature on the topical use of adapalene and also on the use of the Er-YAG laser as possible alternative treatments. [6,7] ❑

Abstract: Dowling-Degos disease is a rare genodermatosis characterized principally by progressive reticulate hyperpigmentation of the flexures. Although the condition is benign, it may be associated with skin malignancies. Furthermore, the characteristic skin changes may be psychologically and socially detrimental to the patient as a result of the unsightliness of the condition. The present report describes a case in which this disease was associated with a keratoacanthoma, and summarizes current concepts on this skin disorder.
Keywords: Evolution; Hyperpigmentation; Keratoacanthoma

Resumo: A doença de Dowling-Degos é uma genodermatose rara, caracterizada principalmente por hiperpigmentação reticular progressiva de áreas flexurais. Apesar de apresentar evolução benigna, pode estar associada a neoplasias da pele. Além disso, as alterações cutâneas características potencialmente ocasionam prejuízo psicossocial, devido aos danos estéticos significativos. Os autores descrevem um caso dessa doença associado a ceratoacantoma e sintetizam os conceitos atuais sobre ela.
Palavras-chave: Ceratoacantoma; Evolução; Hiperpigmentação

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MAILING ADDRESS / ENDEREÇO PARA CORRESPONDÊNCIA:
Clarissa Barlem Hohmann
Travessa Jaguarão, 45/1007; Bairro São João.
90520 070 Porto Alegre, RS - Brasil.
E-mail: clabarlem@hotmail.com
Tel.: (51) 3273-3216 (51) 9806-9814.

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