

Association of Dowling-Degos disease and multiple seborrheic keratosis in a “Christmas tree pattern”

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Abstract

Dowling-Degos disease is a rare sporadic or autosomal dominant pigmentary entity, in which clusters of papules and reticulate macules slowly develop with predominance in flexural regions. This entity is due to mutations in the keratin 5 gene, and is related with other cutaneous disorders. We report the sporadic form of Dowling-Degos disease in an elderly man with multiple seborrheic keratosis in a “Christmas tree” pattern. Worthy of note in this case study is the lesions evolved for over than 30 years. The aim is to describe the association of these keratoses with Dowling-Degos disease in a healthy man.

Keywords: Dowling-Degos disease, Reticular pigment anomaly of flexures, Seborrheic keratosis.

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Introduction

Dowling-Degos disease or reticular pigment anomaly of flexures is a rare autosomal dominant genodermatosis clinically characterized by the gradually progressive development of symmetrical reticulate pigmentation of the flexures, perioral comedone-like lesions and pitted scars (1-7). Classical changes can appear in childhood, but adult onset is more common, usually in the third or fourth decade of life, affecting both genders with predominance of 2:1 in females (1,2,4,7). The “Christmas tree” pattern may be found in skin lesions of exanthematic psoriasis, Kaposi sarcoma, mycosis fungoides, pityriasis rosea, and secondary syphilis (8,9). The aim is to describe multiple seborrheic keratosis in a “Christmas tree” pattern affecting a healthy man with Dowling-Degos disease.

Case report

A 75-year-old afro descendent man was hospitalized with transient ischemic attack (TIA). Because of diabetes mellitus, arterial hypertension, and antecedent ischemic cerebrovascular events, he was taking captopril, metformin, indapamide, and simvastatin. Physical examination, showed perioral acneiform pits and comedones, and symmetric bilateral pigmented papules in the flexures (neck, axilla, and groin) and around the ears. There were thoracodorsal macules with a “Christmas tree” pattern, with no remarkable skin changes in extremities (Fig. 1). He was 45-years-old at the onset of lesions, which slowly appeared in the groins and axillae and after affected ears and neck. Over three decades, the pigmented changes had progressed and evolved asymptomatic, except for rare pru-

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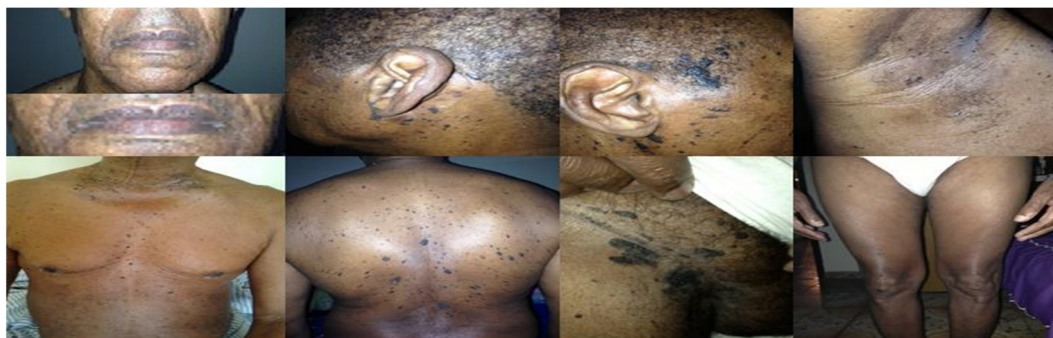


Fig. 1. Bilateral perioral acneiform comedones and pits, symmetric pigmented papules in the flexural regions (neck, axillae, and groins) and around the ears. There were thoracodorsal dark macules presenting a "Christmas tree" pattern, and no remarkable skin changes in extremities.

ritus. He wondered that the skin papules were associated with his natural ageing; and the pigmented macules over the trunk as a consequence of his afro descendent origin. Otherwise, he observed that the main development of hyperkeratotic papules occurred in areas of friction. He denied family history of similar lesions. During the last two years, he had been under clinical outpatient surveillance. Laboratory tests and imaging studies were done to discard the hypothesis of eventual unsuspected malignancy. Routine controls, tumor markers, gastrointestinal endoscopy, and chest

images were normal. The histopathology study of flexural lesions showed keratin plugs within the dilated follicular ostium of pseudocysts from seborrheic papules, increased melanin in the basal membrane, melanophages in the superficial dermis, melanocytes in the granular layer, and thin branches of epidermal downgrowths in the upper dermis (Fig. 2). The clinical features and microscopic findings were consistent with the diagnosis of Dowling-Degos disease coexistent with seborrheic keratosis. Taking in account the absence of any consistent finding of internal malignancy or HIV infection, he was discharged to home after improvement of the neurological manifestations related to TIA.

Discussion

Dowling (1938) and Degos (1954) were the first to describe the type of reticular pigmentation called dark dot disease, Dowling-Degos disease, or reticular pigment anomaly of flexures (2,5-7). This entity is due to mutations in the keratin 5 gene, and is related with other cutaneous disorders. Acropigmentation of Dohi, Dowling-Degos disease, Galli-Galli disease, and reticulate acropigmentation of Kitamura, can be overlapped and may be variants of the same entity (1,3-6). Associated conditions include epidermal cysts, hidradenitis suppurative, keratoacanthoma, pilonidal cysts, seborrheic keratosis and squamous cell carcinoma (2-4,6). These phenomena are indicative of the role played by simultaneous underlying defects of follicular prolifera-

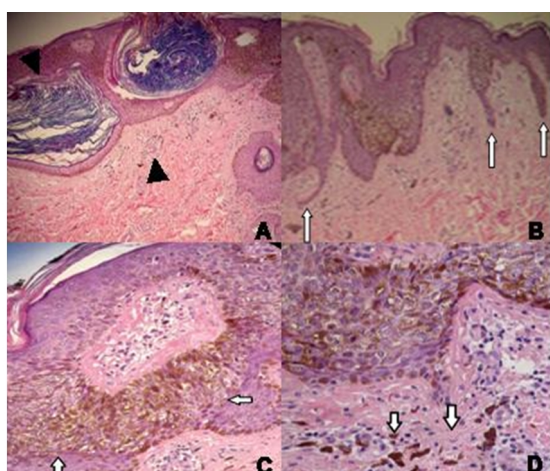


Fig. 2. (Histopathology features of specimens from the flexural lesions). A: Seborrheic keratosis associated with epidermal downward filiform elongations (arrows) to the upper dermis (HE, x20); B: Keratin plugs within dilated follicles (arrowheads) appearing like horny pseudocysts (HE, x20); C: Conspicuous deposits of melanin (arrows) in the cells of basal layer (HE, x40); and D: Increased number of melanophages (arrows) in the upper dermis (HE, x40).

tion (5). The pigmented changes typically follow a slow and usually asymptomatic course (1,2), with symmetrical growth of the lesions over several years, and worsening after sunlight exposition (2). Confluence of papules in areas of friction, and episodes of mild pruritus may be observed (2,4). The changes can appear with diverse tones of brown, black or blue discoloration, and in addition to flexures, the round to oval lesions may affect the face, chest, perineum, or extremities (1,2,4). Histopathology of Dowling-Degos disease include adenoid or reticulated type of seborrheic keratosis with epidermal thickening of basaloid cells, increased deposits of melanin in the basal layer, numerous melanophages in the papillary dermis, epidermal rete ridges with downward filiform elongation (“antler-like”), and dilated follicles appearing as keratin-filled cysts (1-7). This old Brazilian man presented a longstanding development of a reticular pigment anomaly of flexures coexistent with seborrheic keratoses, in addition to numerous hyperchromic macules on his back mimicking the “Christmas tree” pattern. An initial concern should be about the eventual paraneoplastic significance of this finding. Progressive seborrheic keratosis are common findings among patients of elderly groups, and may be found in individuals with the diagnosis of Dowling-Degos disease (2,3,5,6). Because of the esthetically consequences, patients frequently search for interventional strategies (5); although medical treatments are not effective to control their cutaneous changes, ablative laser therapy constitutes a successful option (1-4,6,7). The thoracodorsal changes with “Christmas tree” distribution could be due to a Wolf isotopic response, for example associated with an undiagnosed episode of

juvenile or early-adult pityriasis rosea (10). In conclusion, the elderly man herein reported had characteristic changes of sporadic Dowling-Degos disease, which coexisted for over than 30 years with hyperpigmented thoracodorsal lesions presenting a “Christmas tree” pattern. Although with inherent weaknesses of single case studies, the present report of concomitant uncommon conditions may contribute to better knowledge about their pathogeneses still unclear.

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