Letter to the Editor

Coexistent Dowling-Degos Disease and Reticulate Acropigmentation of Kitamura With Progressive Seborrheic Keratosis

To the Editor:

Reticular pigment anomaly of flexures, also called Dowling-Degos disease (DDD), is an autosomal-dominant genodermatosis characterized by reticulate pigmentation of the flexures. We report a case of DDD concomitant with unusual clinical features. In addition to the typical flexural pigmented reticulate macules, progressive seborrheic keratosis–like lesions on the affected flexural areas and reticulate acropigmentation of Kitamura (RAK) also were present.

A 55-year-old woman presented with symptomatic reticulate hyperpigmentation on the neck, axillae, inframammary region, inguinal areas, extremities, and the dorsa of the hands and feet (Figure 1). Head hair and nails were normal, but axillary hairs appeared scarce. The eruptions began to develop in childhood and became more extensive throughout adulthood. She did not report pruritic or painful sensations on the affected flexural areas, but she experienced pigmentation worsening after sun exposure. In the last 15 years, a large number of seborrheic keratosis–like lesions gradually developed, predominantly in the flexural pigmented areas (Figure 1B). She also presented with pitted perioral scars. Her family history was positive for genodermatosis and some family members had similar presentations. Histopathologically, thin branching and elongation of rete ridges with basal hyperpigmentation were observed in the right inguinal area. Neither perivascular lymphohistiocytic infiltration nor acantholysis were found (Figure 2). A biopsy specimen from one of the seborrheic papules showed typical histopathologic features of the adenoid type (or reticulated type) of seborrheic keratosis including epidermal thickening consisting of basaloid cells and multiple keratin-filled cysts (Figure 3). Blood, urine, and stool routine examinations; liver and renal function tests; and blood pressure, were all within reference range. Chest radiograph, abdomen ultrasonography studies, and breast examinations were unremarkable. Tumor makers including carcinoembryonic antigen, cancer antigen (CA)–19-9, CA-242, α-fetoprotein, prostate-specific antigen, human growth hormone, neuron-specific enolaser, β-human chorionic gonadotropin, free prostate-specific antigen, CA-125, and CA-15-3 were found far below the upper limit of the reference range. Nevertheless, the ferritin blood level was

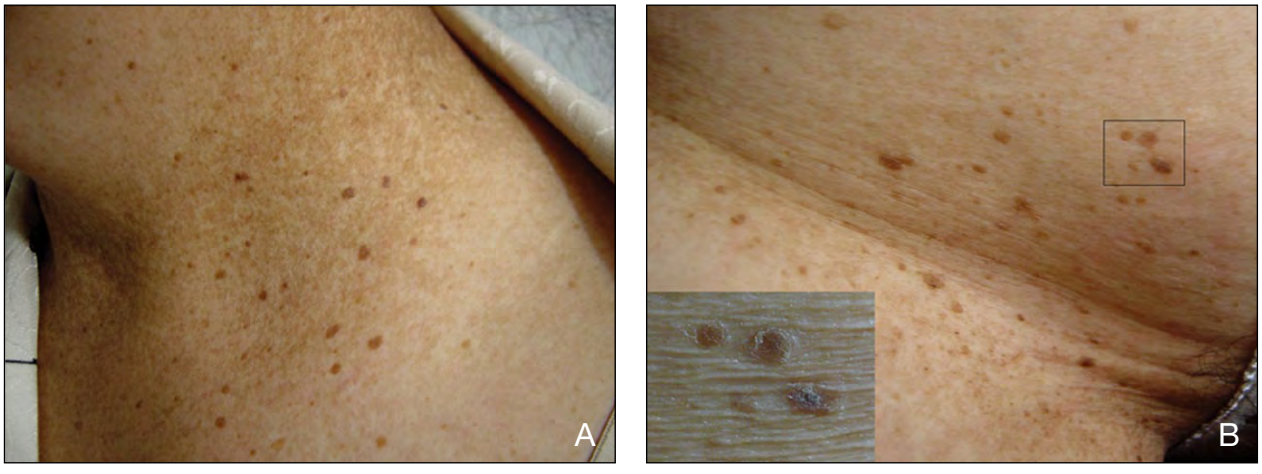

Figure 1. Clinical features of the reticulate pigmented eruption, including symptomatic reticulate hyperpigmentation on the axilla (A) and inguinal area (B). Seborrheic keratosis–like lesions gradually developed (inset)(B).

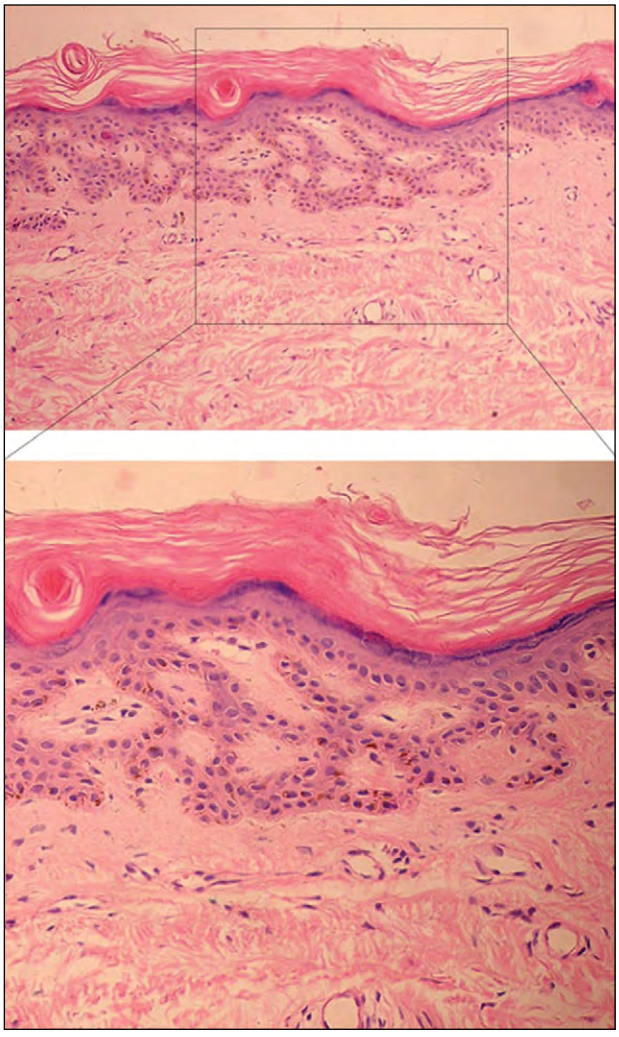

Figure 2. Light microscopy revealed typical thin branching of epidermal downgrowths from one of the lesions of reticulate hyperpigmentation (H&E, original magnification ×100 [inset magnification ×400]).

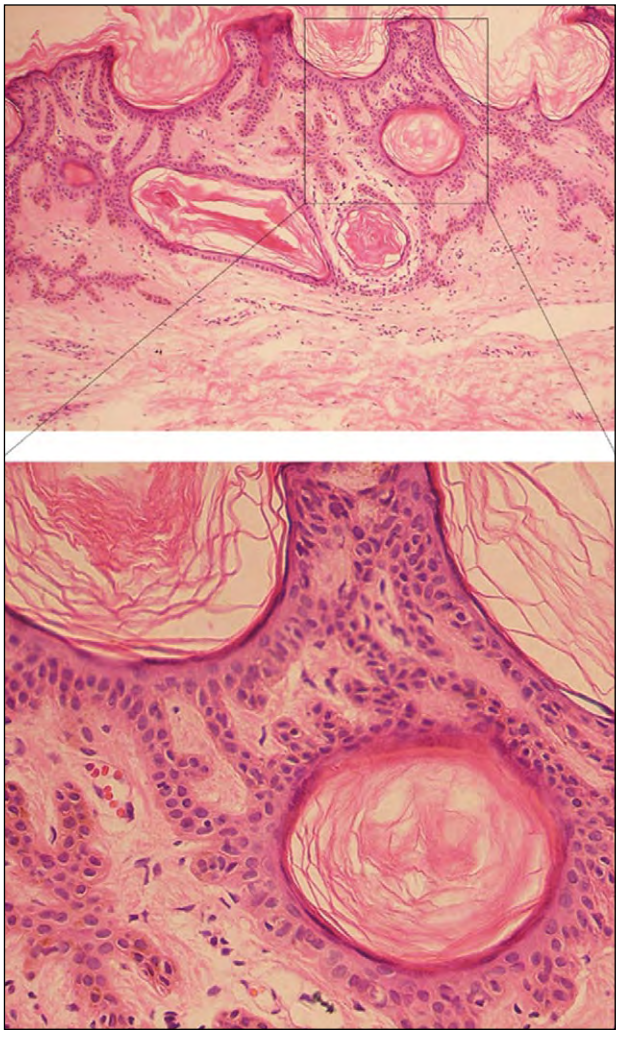

Figure 3. Light microscopy revealed many keratin-filled cysts from a seborrheic papule (H&E, original magnification ×100 [inset magnification ×400]).

250 ng/mL (reference range, 15–200 ng/mL). The overlap of DDD and RAK concomitant with progressive seborrheic keratosis was diagnosed on the basis of the clinical and histopathologic features.

Although DDD and RAK are benign conditions, they usually cause aesthetically annoying consequences.[1] They are subsumed under the heading of reticulate hyperpigmentation and are inherited as a rare autosomal-dominant trait with variable penetrance.[2] The association of these 2 conditions has suggested the existence of a clinical spectrum between the DDD and RAK poles[3] that could represent different features of the same disease. Findings from our case strengthen this hypothesis.

Some disorders such as multiple epidermal cysts, hidradenitis suppurativa, and squamous cell carcinomas have been reported in association with DDD-pigmented areas.[4] The coexistence between DDD and seborrheic keratosis does not seem to be coincidental in this patient because several members of her family were affected by both conditions. Seborrheic keratosis is one of the more common benign epidermal neoplasms; however, the association with malignancy can be useful as a cutaneous clue for diagnosis of occult internal tumors.[5] Therefore, one must emphasize the role of correctly identifying its variants. The distinct clinical presentations in this patient concomitant with progressive seborrheic keratosis may represent an unusual variant of DDD associated with RAK and scarce axillary hairs. These clinical features may reflect simultaneous underlying defects of follicular proliferation. Furthermore, the comparison

between the classic and variant forms may contribute to DDD gene mapping in special keratin genes.[6] Characterization of the DDD gene will provide important clues to better understand the molecular mechanisms of pigmentation and would increase the hope for effective interventional strategies in clinical practice.

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The authors report no conflict of interest.

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