Coexistence of reticulate acropigmentation of Kitamura and Dowling-Degos disease

Ana Rita Cabral, Felicidade Santiago, José Pedro Reis
Department of Dermatology, Coimbra University Hospital, Coimbra, Portugal

Abstract

Reticulate acropigmentation of Kitamura (RAK) and Dowling-Degos Disease (DDD) are rare genodermatoses inherited as an autosomal dominant trait with variable penetrance. They are part of a spectrum of diseases with hyperpigmented macules coalescing in a reticular pattern, facial and palmoplantar pits, breaks in dermatoglyphics, comedo-like lesions and epidermoid cysts, and a unique histological picture of hyperpigmented digitate epidermal downgrowths. The authors describe a case of a 45-year-old female with reticulate acropigmentation of the dorsa of the hands and feet, hyperpigmented macules on the axilla and around the mouth, and palmar pitting. Clinical and histological findings, together with a relevant family history, allowed the authors to consider this case an example of the rare event of an overlap RAK-DDD.

Case Report

An otherwise healthy 45-year-old female presented since early childhood, with a progressive and asymptomatic reticulated acropigmentation with brown pigmented macules on the dorsa of the hands, forearms and feet (Figures 1, 2). Hyperpigmented macules, ephelide-like, on both axillae, buttocks and perioral area, also appeared in adulthood (Figure 3). The physical examination revealed several breaks in dermatoglyphics, comedo-like papules or alterations in mucous membranes, hair, teeth and nails. The patient reported similar lesions (mainly the reticulated acropigmentation) on two relatives (mother and aunt) apparently in an autosomal dominant pattern. He refused a genetic study proposed by the authors.

The skin biopsy of a pigmented macule of the flexor aspect of the forearm revealed elongated rete ridges, with increased pigmentation of the basal layer and an increment in the number of melanocytes, characteristic of RAK (Figure 5).

Based on the history, clinical and pathologic findings, the authors believe that this case is one more example of the rare event of overlap RAK-DDD, among the few non-asian cases described in literature.

Discussion

Rebora and Crovato first suggested in 1983 that the two entities, RAK and DDD were different phenotypic expressions of the same genodermatosis. In similarity to the other RAK-DDD overlap cases described in the literature, our patient had typical characteristics of both variants. Concerning RAK, it was possible to perceive the reticulated hyperpigmentation with acral distribution and palmar pits and, in addition, the hyperpigmented lesions on the flexures were typical of DDD. The transmission was autosomal dominant, with other members of the family affected.

The true knowledge of the relationship between RAK and DDD will be possibly achieved, through the clarification of the genetic background of both diseases.
References


Case Report

Figure 3. Hyperpigmented, ephelide-like macules around the mouth.

Figure 4. Palmar pits.

Figure 5. Histopathology of an hyperpigmented lesion of the dorso of the hand.