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We hope that making available the relevant information on Pachyonychia Congenita will be a means of furthering research to find effective therapies and a cure for PC.
Pachyonychia congenita in the absence of other syndrome abnormalities

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Pachyonychia congenita was first described by Jadassohn and Lewandowski in 1906. Nail changes that affect all nails symmetrically are the hallmark of the disease. The base of the nails appears to be normal but the free edge is raised by a thick, subungal hyperkeratosis. The lateral borders often angulate toward the center. Associated symptoms include palmoplantar hyperkeratosis, follicular keratosis, xerodermia, palmoplantar hyperhidrosis, and blisters on the soles of the feet. Leukokeratosis of the tongue and buccal mucosa, corneal abnormalities, and malformations of the teeth have also been described. On the basis of the prevalence of associated symptoms, numerous subdivisions of pachyonychia congenita have been suggested. Both autosomal dominant and autosomal recessive forms have been described, reflecting heterogeneity.

We describe a Moroccan family with nail deformities in the absence of other syndrome abnormalities, which demonstrates that the coexistence of associated symptoms is not an absolute prerequisite for the diagnosis of pachyonychia congenita.

CASE REPORT

Several members of a family had nail deformities that had been present since the first months of life. Affected members were the grandmother, the father, and his four sons, ages 73, 51, 21, 19, 16, and 13 years, respectively. The mother and four other siblings were not affected (Fig. 1). There was no history of consanguinity. In the affected persons all nails of hands and feet showed symmetric thickening and hardening, with yellow-brown discoloration, subungal hyperkeratosis, and upward growth of the distal nail with hypercurvature (Fig. 2). On further examination, the affected persons had no deformities of the skin, hair, mucous membranes, eyes, or teeth. To exclude onychomycosis, nail cultures were examined for the presence of fungi. All cultures were negative.

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Fig. 1. Pedigree of family manifesting a monosymptomatic form of pachyonychia congenita. Circle, female; square, male; shaded symbols, affected; open symbols, unaffected.

Fig. 2. Fingernail shows thickened nail plate, subungal hyperkeratosis, and upward growth of the distal nail with hypercurvature.

No treatment was given because the patients' complaints were of a cosmetic nature only.

DISCUSSION

The nail changes of all five affected family members consisted of symmetric thickening, subungal hyperkeratosis, and upward growth of the distal nail with angulation of the lateral borders toward the center. All nails were equally affected. The nail changes had been present since the first months of life. The pedigree is consistent with an autosomal dominant mode of inheritance.
In the differential diagnosis, congenital onychogryphosis and pachyonychia congenita must be considered. Congenital onychogryphosis is a hereditary nail disorder with an autosomal dominant mode of inheritance. Associated features are lacking. Onychogryphosis appears in adolescence; affects predominantly the thumb and large toenails, and lacks massive subungual hyperkeratosis; the nails curve downward at the free margin. Pachyonychia congenita is characterized by nail changes similar to those of our patients; all nails are symmetrically affected, and the condition appears within the first months of life. Furthermore, the coexistence of associated features is characteristic of the disease.

The nail deformities present in our patients are characteristic of pachyonychia congenita, although the associated findings of the syndrome are lacking. Therefore the family appears to represent a monosymptomatic form of pachyonychia congenita. In these patients the same gene is probably affected as in other cases of autosomal dominant pachyonychia congenita; however, it may represent a different allele, resulting in a milder phenotype.

REFERENCES


Ambivalent response of lymphomatoid papulosis treated with 8-methoxypsoralen and UVA

Peter Wolf, MD, Philip R. Cohen, MD, and Madeleine Duvic, MD
Houston, Texas

Lymphomatoid papulosis is a T-cell clonal proliferation that histologically resembles T-cell lymphoma, except that the lesions undergo spontaneous remission. Lymphomatoid papulosis lesions that initially responded to PUVA therapy flared in the shielded areas of a patient, which suggests that there may be a systemic immunosuppressive effect in addition to the local effect seen from PUVA therapy.

CASE REPORT

A 44-year-old man had self-resolving, recurrent nodules on the trunk and extremities occurring for 2½ years.

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Fig. 1. Lymphomatoid papulosus of the arm, shielded left flank, and axilla that appeared during PUVA treatment.

Examination revealed an indurated erythematous 1.5 cm nodule on the right medial thigh, an ulcerated 1.0 cm nodule on the left inner arm, and several 2 to 3 mm red papules on the right wrist and elbow. There was no adenopathy.