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PACHYONYCHIA CONGENITA TARDA

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A case of Pachyonychia congenita is being reported. The interesting feature being the onset of nail changes beyond the first few years of life.

Key Word : Pachyonychia Congenita Tarda

Introduction

Pachyonychia congenita (PC) is a rare, autosomal dominant disorder characterized by discoloration and thickening of the nails, usually begining within the first month of life.[1,2] Reports of the onset of nail changes beyond the first few years of life are rare. The term pachyonychia congenita tarda (PCT) has been suggested by Paller et al for the late onset form of PC.[3] We herein report one such case.

Case Report

A 30-year-old soldier had discoloration and marked thickening of the fingernails and toenails that began at 8 years of age and persisted unchanged. He was otherwise well except for a recent history of paronychial infection. The patient had no history of cutaneous blisters, natal teeth or corneal dystrophy. His niece was similarly affected with thickened nails that developed at 6 years of age.

Physical examination revealed yellow discoloration and marked thickening of the fingernails and toenails, with subungual hyperkeratosis but a smooth surface (Fig. 1). Some of the nails had a "pinched" appearance at the distal aspect, and the nail tip was angulated upward. The patient in addition had mild palmoplantar hyperhidrosis. No leukokeratosis and/or palmoplantar keratoderma was noticed. Rest of the physical examination was unremarkable.

Results of KOH examinations were negative. Fungus cultures from the nails yielded no organisms. X-rays of hands/feet did not reveal any abnormality. Patient refused biopsy.

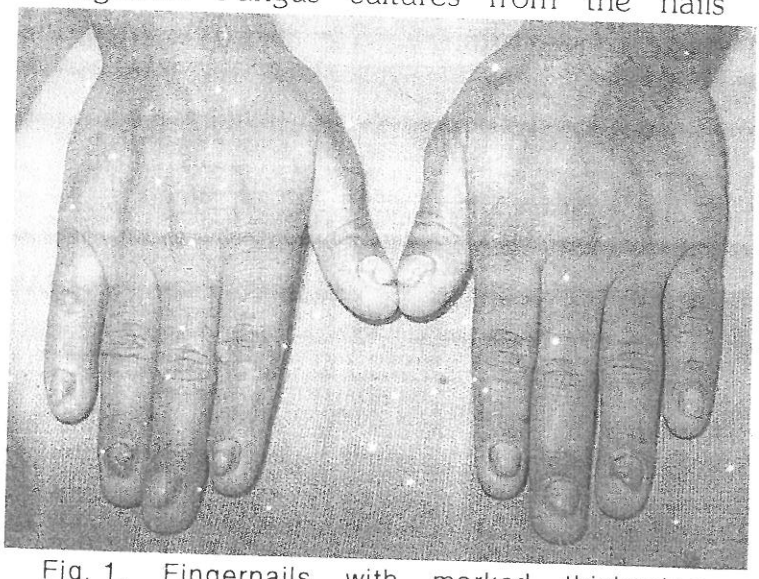

Fig. 1. Fingernails with marked thickening, subungual hyperkeratosis, "pinched" margins and smooth surface.

Comments

PC was originally described by Wilson[4] in 1905, although the association of the disorder with palmoplantar keratoderma and other ectodermal defects was first reported by Jadassohn - Lewandowsky.[5] PC has been subdivided into four groups, based on the clinical features associated with nail changes.[6] Common to almost all patients who have been described, regardless of the

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form of inheritance or subclassification of the disorder, is the onset of pachyonychia in infancy. Only 9 cases have been reported so far with late onset.[1,3] No established classification mentions a late onset form of the disorder. We agree with Paller et al (1991) that this form should constitute a subset of PC and the positive family history of pachyonychia in our patient further supports the opinion that PCT is also inherited in an autosomal dominant manner.[3]

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