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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.
Eruptive Vellus Hair Cyst in a Patient with Pachyonychia Congenita

Hong-Tak Lee, Seung-Ho Chang and Tae-Young Yoon

Abstract

Pachyonychia congenita is characterized by symmetrical nail dystrophy, palmoplantar keratoderma, oral leukokeratosis, and follicular hyperkeratosis. In addition to these features, multiple cutaneous cysts of various kinds have been described.

We report a case of pachyonychia congenita associated with eruptive vellus hair cyst.

Key words: pachyonychia congenita; eruptive vellus hair cyst

Introduction

Pachyonychia congenita (PC) is an uncommon autosomal dominant disorder with variable expression (1-3). Although the unique nail changes are the most striking feature of PC, various cutaneous manifestations can be associated. Associated manifestations include palmoplantar hyperkeratosis, oral leukokeratosis, follicular hyperkeratosis, and corneal abnormality (1, 3). Multiple cutaneous cysts of various kinds have been described as well (4).

We report the second patient with PC associated with eruptive vellus hair cyst (EVHC).

Case Report

A 25-year-old woman was seen for nail deformities and palmoplantar keratoderma that had been present since the first month of life. She also noted multiple papules on both posterior thighs at the age of 17. Her 6-month-old son had similar nail deformities and palmoplantar keratoderma, but other family members had no history of any similar disorder.

All nails of the hands and feet showed symmetric thickening with yellow-brown discoloration, subungual hyperkeratosis, and upward growth of the distal nail with hypercurvature (Fig. 1). Focal white thickening of the tongue and diffuse hyperkeratosis and fissuring of both palms and soles were also observed. Fungal cultures were negative. The lesions of the thighs were multiple, 5 to 10 mm in diameter, firm, flesh-colored papules (Fig. 2). A biopsy was taken from the posterior aspect of the right thigh.

The biopsy specimen showed a keratin-filled cyst in the middermis (Fig. 3). The cyst was lined by squamous epithelium three to five cell layers
thick that contained laminated keratin and vellus hairs cut in cross and tangential section (Fig. 4). There was no sebaceous element. This cyst was diagnosed as EVHC.

The patient and her son were otherwise well.

Discussion

PC is characterized by symmetrical nail dystrophy, palmoplantar keratoderma, oral leukokeratosis, and follicular hyperkeratosis (1–3). In addition to these features, multiple cutaneous cysts may be present on the face, neck, or trunk. These are usually steatocystoma multiplex (SM) (1, 5, 6) or epidermoid in nature (2, 7). To our knowledge, EVHC associated with PC has been reported in only one case (8).

Although EVHC has been reported as a separate disease entity, patients with both EVHC and SM have been reported (9, 10). Furthermore, a patient with EVHC, SM, and epidermoid cyst was described recently (11). Therefore, Ahn et al. (11) suggested that EVHC, SM, and epidermoid cyst are within the spectrum of the same disease process, and each pure type represented one end of a spectrum. In 1994, Moon et al. (8) described a patient with PC associated with both EVHC and SM.

Our case confirms that the previous report by Moon et al. was not fortuitous and suggests that PC may be associated with not only SM and epidermoid cyst but with EVHC.

References

4) Dahl PR, Daoud MS, Su WP: Pachyonychia con-


