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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 and steatocystoma multiplex in a patient with pachyonychia congenita

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Eruptive vellus hair cyst (EVHC) has been reported as a separate disease entity.\(^1\)-\(^3\) Recently, however, patients with both EVHC and steatocystoma multiplex (SM) and a cyst with histologic features of both SM and EVHC have been reported.\(^4\)-\(^8\) Therefore these disorders were suggested to belong to one disease process. Pachyonychia congenita (PC) is associated with many other diseases,\(^9\),\(^10\) one of which is SM.\(^9\) We describe the first patient with PC associated with both EVHC and SM. This may be further evidence of a unifying concept.

CASE REPORT

A 29-year-old man had numerous nodules and papules and thick nails. Thickening and discoloration of his nails had first been noted on both thumbs and big toes at 10 years of age and later involved the other fingers and toes. About 7 years ago lesions developed on his neck and progressively increased in size, number, and extent. Not infrequently they suppilated spontaneously. Blisters were readily induced by trauma on his palms. Family history was noncontributory.

Examination revealed thickening and a gray-blue discoloration of all nails. Hyperkeratosis was noted on his left palm and both soles. Hundreds of variable-sized flesh-colored to yellow or white-yellow papules and nodules mixed with erythematous papules involved the entire skin (Fig. 1). The scalp overlying large nodules was alopecic.

Biopsy specimens were obtained from the chest and right forearm. One specimen disclosed a keratin-filled cyst in the mid dermis. Several segments of vellus hair were found in the cyst. The cyst was lined by squamous epithelium with focal areas of a poorly developed granular layer. There was no sebaceous element. A focal inflammatory cell infiltrate was present around the cyst wall. This cyst was diagnosed as EVHC (Fig. 2, A). Another specimen showed an empty cyst that had portions of sebaceous elements within its wall. The cyst wall was infolded and consisted of several layers of squamous epithelium. A prominent eosinophilic cuticle was also noted (Fig. 2, B). A third specimen showed a cyst that contained homogenized keratinous material without hair elements. The cyst wall consisted of two to three layers of squamous epithelium and contained a sebaceous lobule. The histologic diagnosis of these two specimens was SM.

Fig. 1. A, Hundred of nodules and papules on anterior chest. B, Thickening and discoloration of nails of both hands.

DISCUSSION

Recently Ohtake et al.\(^11\) described a patient with both SM and EVHC and proposed that these two diseases lie in the spectrum of the same disease pro-
cess and could be termed multiple pilosebaceous cysts.

PC is a rare hereditary disease characterized by
nail dystrophy, palmar and plantar hyperkeratosis,
leukokeratosis of the mucous membrane, follicular
keratosis, and occasionally hyperhidrosis of palms
and soles\textsuperscript{9} that usually develops in early infancy.
This patient had a late-onset form of the disorder.
This subset was reported by Paller et al.\textsuperscript{10} as pachy-
onychia congenita tarda. Because EVHC and SM are
within the spectrum of the same disease process
and because of the close relation between SM and
PC, EVHC alone or both EVHC and SM can de-
velop in a patient with PC. Our patient may be a
good example of this and supports this concept.

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