Palmoplantar Keratosis Acuminata With Facial Sebaceous Hyperplasia

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We examined a patient with acuminate keratoses of the palms and soles and sebaceous hyperplasia of the face. No similar cases were found in an examination of the literature.

We recently had the opportunity to study an interesting patient with an unusual genodermatosis, characterized by an acuminate keratosis of the palms and soles and sebaceous hyperplasia of the face. We believe this case represents a variant of the congenital punctate keratoderma. It is well known that there may be interfamilial variation of the punctate keratoderma. In a recent search of the literature, however, we were unable to find a report of this unusual phenotypic combination. We therefore believe this case deserves reporting.

Report of a Case

A 50-year-old, retired marine corps officer first noted at the age of 9, "hairy" growths on his hands and feet that involved the entire palmar and plantar surface. These lesions were asymptomatic and were not associated with hypohidrosis or hyperhidrosis.

At the age of 12, the patient developed multiple 2- to 3-mm grayish-yellow papules of his face and anterior cervical region, which were asymptomatic. Over the course of the next 30 years, these lesions approximately doubled in size.

The patient has been told that the lesions of palms and soles have occurred in approximately 50% of the male members in his family over the past nine generations. He personally knows of his grandfather, father, and six of ten siblings being affected with this condition. About 59% of the female members have been affected with palmoplantar lesions without having the associated sebaceous hyperplasia.

The patient has had an active life and has experienced excellent health, as have all the affected family members. There is no history of dental, hair, or nail abnormalities or other cutaneous problems.

Physical examination revealed on the palmar surface of the hands and the plantar surface of the feet numerous, discrete, 1- to 3-mm yellowish-brown, acuminated verruca-keratotic lesions that did not coalesce. These lesions could not be removed from their noninflammatory bases (Fig 1 and 2). The nails, hair, oral mucosa, and dentition were normal. The entire face and neck, extending to and over the shoulders, manifested 3- to 5-mm discrete, brownish-yellow papules. The remainder of the skin was normal.

A starch iodine test using strong iodine solution (Lugol's solution) and filter paper showed normal to slightly decreased eccrine activity of the palms and soles. The VDRL test for syphilis and fluorescent treponemal antibody and acute brain syndrome (FTA-ABS) and serologic studies were both normal. Ophthalmologic examination did not disclose any corneal abnormalities.

A biopsy specimen of a characteristic palmar lesion showed marked focal hyperkeratosis in a verruciform pattern. Focal parakeratosis, thickening of the granular layer, and slight irregular acanthosis characterized the epidermis. The subjacent corium was normal.

Fig 1.—Hand showing typical acuminata lesions.
Fig 2.—Feet showing typical acuminata lesions.
A biopsy specimen of a facial papule revealed follicular plugging in an otherwise normal epidermis. There were increased numbers of mature sebaceous glands of varying sizes and shapes. The accompanying hair follicles contained normal keratinaceous structures. There was a mild, chronic, nonfocal, inflammatory cell infiltration in the papillary corium.

The patient did not seek medical attention for his lesions until the age of 38. The palmoplantar lesions remained essentially unchanged despite various topical treatments, except when the lesions were subjected to occasional planing with sandpaper. For a period of more than one year at four-month intervals, respectively, the patient was treated with 10% salicylic acid in anhydrous lanolin (Aquaphor), 45% urea in an emollient (Keri Lotion), and 0.5% tretinoin (vitamin A acid) in propylene glycol without marked effect. The sebaceous hyperplasia has never been treated.

Comment

Palmar and plantar keratodermas are characterized by focal or diffuse thickening of the stratum corneum and may be congenital, acquired, or secondary to other disease processes. Examples of the congenital disorders characterized by diffuse palmoplantar keratoderma include progressive palmoplantar keratoderma (Greither disease), mal de meleda, Papillon Lefevre syndrome, mutilating keratoderma, and keratosis circumscripita. Those congenital disorders characterized by focal discrete lesions include keratosis striata and keratosis punctata palmoplantaris. Our case probably represents a variant of the latter two types of disorders.

Classification of this case or of any case of palmoplantar keratoderma is made by accurate documentation of the following characteristics: the age of onset; whether the palmoplantar hyperkeratosis is diffuse or local; the presence or absence of involvement of cutaneous sites other than palms and soles; the pattern of inheritance; and the presence or absence of the ectodermal or systemic defects.

The palmoplantar lesions in this propositus and all affected relatives become manifest at or around pubescence, a development consistent with the previously reported cases of punctate palmoplantar keratoderma. All affected male patients show focal acu¬minate lesions of the entire palmar and plantar surfaces with associated facial and cervical sebaceous hyper¬plasia, whereas the affected female patients lack facial sebaceous hyper¬plasia. All affected individuals lack extracutaneous, other ectodermal, or systemic defects. The previously reported palmoplantar lesions were punctate, in sharp contrast to the acu¬minate spiny features reported here. In addition, sebaceous hyperplasia has not been reported to occur with any other congenital focal kerato¬dermas.

The cases of punctate palmoplantar keratoderma reported in the recent literature by Scott and Costello all showed an autosomal pattern of inheritance. There are sufficient cases in this pedigree to be certain that the patient's family also follows an autosomal dominant inheritance pattern.

Based on the above criteria, the features of this case are best classified as keratosis punctata palmoplantaris. However, the spiny configuration of the keratosis and the associated sebaceous hyperplasia are features that have, to our knowledge, not been previously reported with this condition.

We report this case not to attach a new name to this variant of a previously reported congenital keratoderma, but rather to document a familial variation of a condition known to have marked interfamilial variations. We hope that with subsequent recording these disorders and their variations some day will be classified into syndromes that are not only clinically but also genetically understandable.

References