A Case of Keratosis Punctata Palmaris et Plantaris

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We report a rare case of sporadic keratosis punctata palmaris et plantaris in a 49-year-old woman. Thirty years ago, firm semitranslucent papules developed on the middle portion of her palms without any subjective symptom. The lesions had flat-topped smooth surfaces. They were concentrated along the margins of the hands, especially on the lateral surfaces of both index fingers and finger tips. Ten years ago, similar hyperkeratotic papules with or without central craters developed on the weight bearing portions of her soles. Her family and past histories were negative. The biopsy specimen revealed a dense hyperkeratotic stratum corneum which fitted into a cup shaped depression in the stratum malpighii and a turning inward of the rete ridges at the margin. The Masson trichrome and elastic stains showed no specific dermal changes.
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Key Words : Keratosis punctata palmaris et plantaris, Sporadic

Keratosis punctata palmaris et plantaris(KPPP) was first described by Davis-Colley(1879)[1] following which numerous appellations have led to much confusion. It is characterized by asymptomatic, discrete, firm, semitranslucent papules on the palms and soles[2]. This autosomal dominantly-inherited disease usually begins at or soon after puberty, but frequently sporadic cases are encountered. Most of the patients have no associated features, but a few cases were associated with internal malignancies[3,4].

We report a rare case of sporadic KPPP in a 49-year-old woman with a short review of literatures.

CASE REPORT

A 49-year-old woman visited due to hyperkeratotic papules on the both palms and soles. Thirty years ago, firm semitranslucent papules developed on the middle portion of her palms and have since spread over her entire palms. Ten years ago, similar hyperkeratotic papules developed on her soles, especially on the weight bearing portions. But, she had no subjective symptoms at all. Her family history was non-contributory. She was diagnosed with irritable bowel syndrome seven years ago but has remained symptom free for the past four years. Despite careful questioning about possible exposure to arsenic, no history was found.

On physical examination, multiple papules were observed on both palms(Fig. 1A) and soles(Fig. 2A). The sizes of palmar papules varied from 2 to 5mm in diameter. They had flat-topped smooth surfaces with or without central craters. They were especially concentrated on the lateral surfaces of both index fingers and finger tips(Fig. 1B). Similar semitransparent hyperkeratotic papules with or without central craters were seen on the soles around the toes and heels(Fig. 2B), especially on the weight bearing portions. Other skin manifestations suggesting atopic dermatitis or icthyosis vulgaris were not found.

Routine laboratory examinations, including CBC, LFT, U/A, electrocardiogram, and chest roentogenogram were all within normal limits. The results of a VDRL test for syphilis and fungal culture were negative. An extensive physical examination, radiological, and laboratory evaluation for the purpose of detecting occult internal malignancy

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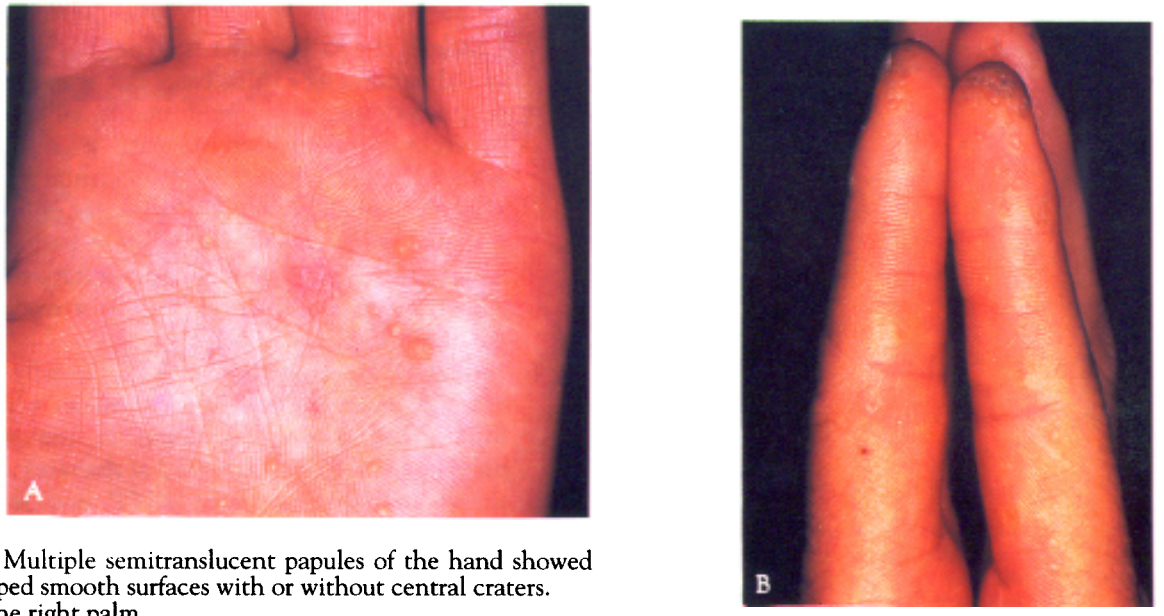

Fig. 1. Multiple semitranslucent papules of the hand showed flat-topped smooth surfaces with or without central craters.
A. on the right palm
B. on the lateral surfaces of both index fingers and finger tips

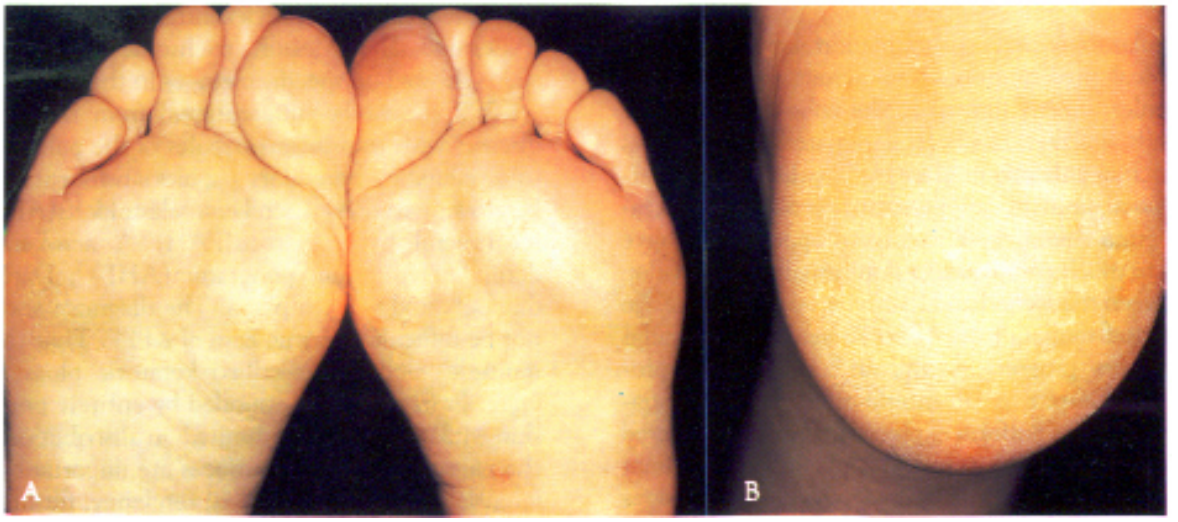

Fig. 2. Multiple semitransparent hyperkeratotic papules & plugs of the feet.
A. on both sides of the soles
B. on the left heel

revealed no abnormal findings. The radiological evaluation included UGI, barium enema, and mammography. The laboratory evaluation included stool occult blood test, tumor markers. A biopsy specimen taken from the papule of the right palm revealed a hyperkeratotic horny layer with a slight central depression. The epidermis beneath such depression showed acanthosis with moderate elongations of rete ridges(Fig. 3). The Masson trichrome and elastic stains showed no specific changes in the derm

As a treatment, 0.05% topical tretinoin and 30% TCA peeling were performed. But the lesions did not improve.

DISCUSSION

Keratosis punctata palmaris et plantaris (Davies-Colley, Buschke, Fischer, Brauer) is one form of

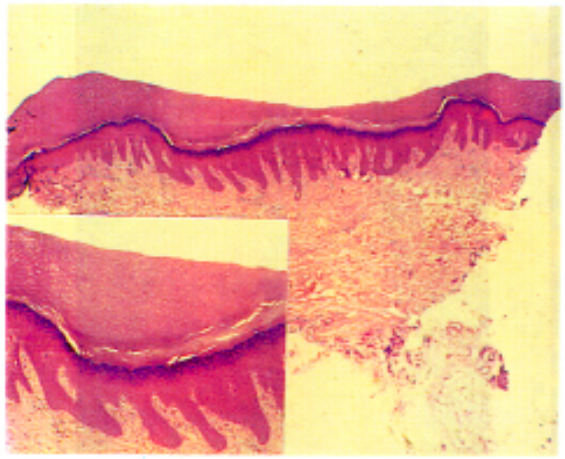

Fig. 3. Hyperkeratotic stratum corneum fitted into a cup shaped depression in the stratum malpighii and a turning inward of the rete ridges at the margin. No parakeratosis (right palm, H&E stain, ×40).

papular hereditary palmoplantar keratoses[5]. The clinical presentation of numerous tiny keratotic papules, strictly limited to the volar aspects of the hands and feet, has been designated porokeratosis punctata palmaris et plantaris[6,7], palmoplantar keratosis acuminata[8] and punctate porokeratotic keratoderma[9,10] and these numerous appellations have led to much confusion. Lesions usually first develop between the second and fourth decades, with the age of onset ranging from 12 to 70 years. The papular keratoses progress slowly, and remain asymptomatic. Despite great interfamilial clinical variation, there is uniform expression within an affected family. Localized forms limited to the palmar creases have been described[11]. In this case, her lesions began at puberty on the middle portion of the palms and spread over her entire palms and progressed to the soles with no subjective symptom. She also had no specific family history.

The cause of the KPPP is not known, but dual influence of genetic and environmental factors may trigger the disease in many cases. A strong association between KPPP and hard manual labour has even been postulated. Hyperhidrosis does not accompany this disorder. Previously, it has been stated that the prevalence of dermatophytosis in patients with diffuse palmoplantar keatoderma of the Unna-Thost variety was 36.7%, but in KPPP, affinity to dermatophytes has not been demonstrated.

Most of the KPPP patients do not have any associated features as in this case. But spastic paralysis[12], ankylosing spondylitis[13] and facial sebaceous hyperplasia[8], have been reported in association with KPPP. In addition, a coincidental[4,14], and a possible familial[3] association with gastrointestinal malignancy have been discussed. The pathogenesis is not certain but abnormal keratinization of the acrosyringium, mechanical trauma or maceration can be related[15].

Histologic examination of KPPP usually shows dense hyperkeratotic stratum corneum which fit into a cup shaped depression in the stratum malpighii and a turning inward of the rete ridges at the margin[18,19]. Sometimes, KPPP may reveal a compact column of parakeratosis resembling that of a cornoid lamella seen in porokeratosis[4,5] . A biopsy specimen taken from this case revealed a hyperkeratotic horny layer with a slight central depression and acanthosis with moderate elongation of rete ridges beneath a central depression in the epidermis. The Masson trichrome and elastic stains showed no specific changes in the dermis.

KPPP should be differentiated from the other diseases with translucent papules of the palms and soles including hereditary papulotranslucent acrokeratoderma(HPA)[20], acrokeratoelastoidosis(AKE)[5], focal acral hyperkeratosis(FAH)[21-24], acrokeratoderma hereditarium punctatum(AHP)[25], Darier's disease[26], and verruca plana. The diagnosis should not be difficult in a true case of KPPP. That these multiple, well-circumscribed keratotic plugs and crateriform pits are surrounded by entirely normal skin and that they are limited in distribution to the palmar and plantar surfaces are important differential factors. The familial tendency, futility of therapy, and the characteristic histological observations are additional factors. Generalized hyperkeratosis or erythema surrounding the individual lesions should not be present.

HPA is known as a rare genetic cutaneous disorder. Clinically it shows white translucent papules with a smooth surface affecting the hands and feet, especially the palms and soles, but there is no subjective discomfort. Histologic study reveals signs of focal hyperkeratosis, hypergranulosis, and acanthosis without any alteration of the dermis. AKE is clinically characterized by small, yellowish, round to oval keratotic papules, mainly confined to the margins of the palms and soles. The

keratotic papules may become confluent in the center of the palms and soles, to produce a diffuse keratoderma. The process begins in adolescence or adult life. The number of papules gradually increases over several years. Local hyperhidrosis is present. AKE can be distinguished by confinement of yellow keratotic papules on the margins of the palms and soles, occasional diffuse keratoderma, and local hyperhidrosis clinically, and elstorrhexis histologically. Histologically, AKE differentiated FAH. FAH, which has an insidious onset in childhood, reaches a maximum in early life, and causes only cosmetic embarrassment. In addition to the typical papules along the borders of the hands and feet, hyperkeratotic papules may be present over the interphalangeal joints of the fingers and toes, and on the heels. On histologic examination, there is no elastorrhexis in FAH. AHP shows wart-like papules with central dimpling on the hands. Histologic findings are hyperkeratosis, hypergranulosis and acanthosis. In Darier's disease, the palms and soles may show punctated keratoses or minute pits. The pits are pathognomonic of Darier's disease whereas the punctate keratoses may be found in other conditions or may also occur on their own as a separate and distinct autosomal dominant trait. But the histology shows a distinctive form of dyskeratosis, with corps ronds and suprabasal acantholysis. Verruca plana can be histologically differentiated from KPPP by vacuolization of cells in the upper epidermis. The differential diagnosis of KPPP is particularly important due to the possibilities of inheritance and internal malignancy.

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