KERATOSIS PALMO-PLANTARIS DIFFUSA —A TYPE WITH SO-CALLED GRANULAR DEGENERATION—

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According to Franceschetti and Schnyder,¹ diffuse hyperkeratosis of the palms and soles unassociated with other cutaneous or systemic abnormalities, inherited as an autosomal dominant trait, is classified as keratosis palmo-plantaris diffusa circumscripta (Thost-Unna).

The clinical picture of this type is characterized by symmetric, diffuse, sharply demarcated (circumscribed) hyperkeratosis of the palms and soles. Frequently, localized hyperhidrosis is present and the lesion is surrounded by a band of erythema.

It is generally accepted that the histopathological findings of keratosis palmo-plantaris diffusa are nonspecific, showing considerable hyperkeratosis, thickening of the granular layer and acanthosis with normal architecture.² On the other hand, however, it is also known that there are some cases of palmo-plantar keratosis having histopathological finding which has been termed granular degeneration³ by Lapière.

In review of the literature, reported cases⁴⁻⁸ of diffuse palmo-plantar keratosis with so-called granular degeneration are very rare. The first report of such condition was that of Vörner⁴ in 1901, in which he described the clinical and histopathological findings of the disease in details which occurred in four generations.

Recently, three Japanese cases of diffuse palmo-plantar keratosis associated with so-called granular degeneration, were observed in the Dermatological Clinic, School of Medicine, Keio University. In view of the rarity of such condition, a brief report seems warranted.

CASE REPORTS

Case 1.— A 20-year-old Japanese female was first seen in our clinic in October, 1965, with the complaint of thickening of the skin of her palms and soles.

Examination showed diffuse marked hyperkeratosis of those regions without any other abnormalities of the skin and its appendages. The diffuse thickening of her palmar and plantar skin was noted at birth. There was no episode of bullae formation. Her elder brother, mother and maternal grandfather are said to have the similar skin lesions. There was no consanguinity between her parent.

Clinical diagnosis of *keratoma palmare et plantare hereditarium* (Unna-Thost) was made.

Case 2.— A 17-year-old male student was first seen in our clinic in June, 1970, for keratotic change of his palms and soles, present since 2 or 3 months of age. There was no family history of similar dermatoses. No consanguinity was noted between his parent.

Except for the skin of palms and soles, physical examination was negative. The palmo-plantar skin lesions, sharply demarcated with an erythematous border, were diffusely hyperkeratotic and yellowish-gray colored (Fig. 1). No hyperidrosis of the lesions was noted.

Clinical diagnosis of palmo-plantar keratosis (Unna-Thost) was made.

Case 3.— A Japanese girl, 7 years of age, was first seen in our clinic in June, 1970, for keratotic change of her palms and soles. According to the patient's mother, the eruption was not present at birth but first appeared at 3 weeks of age. Episode of bullae formation was not noted. There was no family history of similar dermatoses. No consanguinity was noted between her parent.

Examination showed diffuse, dirty yellowish-gray colored, marked hyperkeratosis of her palms and soles (Fig. 2). Deep fissures were also present over these regions. The patient had no other skin complaints and the rest of the physical examination was normal.

Clinical diagnosis of *keratoma palmare et plantare hereditarium* (Unna-Thost) was made.

HISTOPATHOLOGY

Serial sections of paraffin fixed specimens of palm (case 1) and soles (case 2 and 3) were stained by hematoxylin and eosin, Mallory, Gram, Giemsa, colloidal iron and Feulgen methods.

Biopsy specimens of all three of these cases showed essentially the same pictures. They were summarized as follows:

The horny layer is extremely thickened with or without focal parakeratosis. The epidermis is acanthotic and the rete ridges are widened. In the middle and upper portions of the stratum malpighii, the prickle cells show so-called granular degeneration with small and large basophil-staining keratohyalin-like granules and shell-like structures (Fig. 3 and 4). Such disintegrated cells lose their boundaries, and are surrounded by variously sized clear spaces. The basal cell layer is well preserved. The upper dermis shows slight inflammatory infiltrate.

The keratohyalin-like granules noted in the upper stratum malpighii are stained vivid red by Mallory's method. Some of these granules are Gram-positive, purple red by Giemsa's stain and green color by colloid iron method. The granules are Feulgen-negative.

These histochemical findings of the keratohyalin-like granules in our three cases, were similar to those found in a case of bullous congenital ichthyosiform erythroderma studied by Weibel and Schnyder⁹ in 1966.

COMMENT

In this report, the clinical features of three unrelated patients were indistinguishable from those of Thost-Unna type of palmo-plantar keratosis.¹ The mode of inheritance in case 1 was autosomal dominance. Other two patients (case 2 and 3) with no family history of similar dermatoses were solitary cases. Histopathologically, however, all three of these cases showed so-called granular degeneration with marked hyperkeratosis. This characteristic histopathological finding was distinct from that of Thost-Unna type of palmo-plantar keratosis showing only hyperkeratosis.

In Japan, until 1970, about one hundred and sixty cases of various types of inherited palmo-plantar keratosis had been reported. Unfortunately, however, it is impossible to determine the frequency of palmo-plantar keratosis with so-called granular degeneration (Vörner type of palmo-plantar keratosis), because the histopathological study had been performed only in a quarter of these previously reported cases. The first report of Vörner type of palmo-plantar keratosis in Japan was that of Yoshida⁷ in 1967, in which he presented a case of 11-year-old boy with family history of similar dermatoses.

Recently, especially in the American literature, the term epidermolytic hyperkeratosis has been applied to a spectrum of dominantly inherited ichthyosiform dermatoses associated with so-called granular degeneration.

The term epidermolytic hyperkeratosis was introduced by Frost and Van Scott¹⁰ in 1966, and they described that epidermolytic hyperkeratosis "consists of a clinical spectrum of disorders; generalized forms may be bullous, and localized forms may resemble linear nevi."

In 1970, Klaus, Weinstein and Frost⁸ reported nine patients with a familial type of diffuse hyperkeratosis of the palms and soles. Gross clinical features of their cases were similar to those found in Thost-Unna type of palmo-plantar keratosis, however, skin biopsies showed so-called granular degeneration, the characteristic histopathological changes of epidermolytic hyperkeratosis. Moreover, in one case, the changes of epidermolytic hyperkeratosis were obtained by epidermal kinetic studies. Based on the clinical, histological, and kinetic data accumulated in their study, Klaus and his co-workers concluded that it is reasonable to include this special form of diffuse keratoderma of palms and soles in the category of epidermolytic hyperkeratosis.

It is well known that bullous congenital ichthyosiform erythroderma and hyperkeratotic nevus associated with so-called granular degeneration, can occur in one family.¹¹ Schnyder and his co-workers¹² have the opinion, that they are apparently different manifestations of the same mutation. None of our patients, in this report, had a personal or family history of other skin problems and there is no evidence to indicate that Vörner type of palmo-plantar keratosis is associated with bullous congenital ichthyosiform erythroderma.

Reported cases of Vörner type of palmo-plantar keratosis, in the literature, are very rare, and association between such disorder and bullous congenital ichthyosiform erythroderma has not been completely clarified yet. It is desired to accumulate many more case presentations of Vörner type of palmo-plantar keratosis.

SUMMARY

Three Japanese patients with Vörner type of palmo-plantar keratosis were reported.

The clinical features of these patients were indistinguishable from those of Thost-Unna type of palmo-plantar keratosis, but histopathologically they showed so-called granular degeneration with marked hyperkeratosis, which are the characteristic changes of epidermolytic hyperkeratosis.

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Fig. 1. Diffuse hyperkeratosis of the left sole, case 2.

Fig. 2. Diffuse hyperkeratosis of the palms, case 3.

Fig. 3. Biopsy of palmar skin, case 1, showing so-called granular degeneration in the middle and upper stratum malpighii. (Hematoxylin and eosin).

Fig. 4. Biopsy of the right sole, case 2, showing so-called granular degeneration with numerous shell-like structures. (Hematoxylin and eosin).