

Mutilating Palmoplantar Keratoderma Successfully Treated with Etretinate

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Three patients, a mother and her two sons, with mutilating palmoplantar keratoderma (Vohwinkel's syndrome) were successfully treated with oral etretinate (Tigason®). All of them had keratotic constriction furrows of one or more digits (pseudo-ainhum) with

threatening spontaneous amputation. Oral treatment with etretinate brought about disappearance of the pseudo-ainhums and normalization of the digital blood circulation. The hyperkeratotic skin became thinned and pliable with a dose-dependent trend towards redness and atrophy. Continued long term etretinate medication has made it possible for these patients to fulfill their social and occupational activities. All 3 patients had acoustic impairment of high-pitched tone perception. *Key words:* Pseudo-ainhum; Retinoid; Vohwinkel's syndrome. (Received May 22, 1984.)

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Hyperkeratosis of the palms and soles with starfish-shaped projections to the dorsal aspects of hands and feet in a young mother and her 2-year-old child was described by Vohwinkel in 1929 (2). This skin disorder starts in early childhood. In the adolescence or young adult life, it is often complicated by keratotic constrictions (pseudo-ainhum) of fingers and toes with the risk of eventually leading to spontaneous digital amputation. A proper designation for the condition is keratoderma hereditaria mutilans (KHM); it is, however, often referred to as Vohwinkel's syndrome. In patients suffering from KHM, other clinical signs are sometimes seen, such as ichthyosiform skin changes, knuckle pads, cicatricial alopecia, nail anomalies, high-tone acoustic impairment or deaf-mutism (1).

Keratolytic topical remedies such as salicylic acid or high concentration carbamide creams may be of some help in KHM. However, treatment of the phalangeal constrictions, in order to prevent amputation, was previously inadequate. Attempts with plastic surgery (excision of the constrictions and skin transplantation to cover the defects) was beneficial in some cases, but the cosmetic result was often poor.

In 1981, Chang Sing Pang et al. reported successful treatment of KHM in an 11-year-old boy with an aromatic retinoid, etretinate (Tigason®) (1).

The cases of three patients with KHM, a mother and her two sons, efficaciously treated with oral etretinate, are reported.

CASE REPORTS

Case 1

This housewife was aged 51 when first seen in the clinic in the spring of 1981. In addition to palmoplantar keratoderma since early childhood, she had poliomyelitis when she was 15, resulting in a persistent mild paresis of arms, hands and legs. Subsequently, the volar keratotic callus of her hands encircled her fingers which became sclerotic and tapered, some of which developed constriction furrows jeopardizing the blood circulation in the distal portion of the digit. In order to prevent spontaneous amputation of her right thumb and ring-finger, a plastic surgeon in 1961 performed excision of the pseudo-ainhums and skin transplantation.

On examination, she had thick hyperkeratosis of palms and soles, also affecting the dorsal aspects of her digits and the margins of the dorsum of hands and feet where the borders of the keratoderma formed small starfish-like projections. The fingertips had a tapered appearance and were tender, somewhat painful, cold and pale. The previously transplanted skin on the right thumb and ring-finger was soft and pliable, but bulged outward with ulcerated margins against the surrounding keratotic skin as if a rejection of the transplants was about to occur. An audiogram showed a considerable deficit in the perception of high-pitched tones.

Treatment and progress. Oral treatment with etretinate, 50 mg daily, was started in October 1981. In the course of 3-4 weeks, dramatic loosening and detachment of her palmar and plantar keratoses occurred, leaving the skin red and scaly, but soft and pliable. The blood circulation of constricted fingers improved and the ulcerations and pseudo-ainhums disappeared. She has since continued the etretinate medication for 2½ years, regulating the dose herself, changing between 50 and 25 mg daily according to the thickness of the palmar skin. Treated like this, she has, as a housewife, regained full manual function and is very pleased with the result of therapy. On attempts to stop the medication, she has noticed prompt recurrence of the keratoderma.



Fig. 1. Constriction (pseudo-ainhum) of little finger in Case 3 before etretinate treatment.

Case 2

This youngest son of the previous patient was aged 17 when first seen in the clinic in June 1981. His palmoplantar keratoderma first appeared when he was 2 years old. Because of bilateral severe and increasing constrictions of the fifth finger and toe, plastic surgery had been considered in order to prevent spontaneous amputations. He complained of numbness and pain in the distal parts of the constricted digits.

On examination, he had marked hyperkeratotic skin of the hands and feet, resembling the palmoplantar keratoderma of his mother. There were deep keratotic constriction furrows located over the proximal interphalangeal joint of the aforementioned fingers and toes. An audiogram showed a marked hearing loss of high-pitched tones.

Treatment and progress. Oral treatment with etretinate was started in June 1981. The palmoplantar keratoderma did not diminish as dramatically as it did in case 1. Slowly, in the course of two months, the hyperkeratotic skin became less thickened and the pains and paraesthesias of the constricted fingers disappeared. However, even after six months' continuous therapy with etretinate, there was still a persistent keratoderma with remnants of the pseudo-ainhums. The dose of etretinate was therefore increased to 75 mg and later 100 mg daily. On these dosages, he complained of, at times, marked cheilitis, while the condition of his hands and feet improved. Currently, on an etretinate maintenance dose of 50 mg daily, his hands and feet still show some redness and scaling. Nevertheless, he has completed a training school for carpenters and his working capacity is unimpaired.

Case 3

This older brother of the previous patient was aged 21 when he first came to the clinic in January of 1982. As in the case of his brother, and mother, he had, from early childhood, suffered from keratoderma of palms and soles and his hearing was impaired. During the last 6 months, he had noticed a slowly progressing constriction furrow on his left little finger.



Fig. 2. Little finger in Case 3 after treatment with etretinate.

On examination, he presented the very same type of hyperkeratotic skin changes of the hands and feet as were seen in cases 1 and 2. There was an incipient pseudo-ainhum at the base of the middle phalanx of his left little finger together with a slight numbness and paraesthesia of its distal portion. An audiogram showed acoustic impairment of high-pitched tone perception.

Treatment and progress. Oral etretinate medication was started in January 1982. After two months of treatment, his pseudo-ainhum had disappeared almost completely. His palms and soles became soft, but with some redness and scaling. He has now been treated continuously with etretinate, 50 mg daily, for 2 years and is working full time as a glazier.

COMMENTS

The beneficial effect of the aromatic retinoid etretinate has been noted in a wide range of hyperkeratotic skin disorders. In the three cases reported here, not only did etretinate reverse the hyperkeratosis of hands and feet, but it also brought about disappearance of the digital constrictions and thereby restored normal digital blood circulation. The retinoid treatment not only prevented mutilating ulcerations and amputations, but enabled the patients to conduct a normal occupational and social life. During the course of more than 2 years of etretinate therapy, blood tests, including complete blood cell counts, serum level of liver enzymes, blood urea, serum creatinine and serum protein levels have all been within normal range in all three cases.

The fact that etretinate can indeed reverse the digital constrictions in KHM suggests that they are caused by excessive and pathological keratinization.

The bilateral neurogenic acoustic impairment, described in all three patients, was not influenced by the retinoid therapy.

REFERENCES

1. Chang Sing Pang AFI, Oranje AP, Vuzevki VD, Stolz E. Successful treatment of Keratoderma hereditaria mutilans with an aromatic retinoid. Arch Dermatol 1981; 117: 225-228.
2. Vohwinkel KH. Keratoderma hereditarium mutilans. Arch Dermatol Syph 1929; 158: 354-364.