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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.
ACQUIRED LOCALIZED KERATOSIS PALMARIS ET PLANTARIS

REFERENCES.


plantar warts appeared on the palms (Fig. 1), which were entirely unaffected by external applications.

It was finally decided to test the effect of vitamin A, and 100,000 i.u. daily was ordered. After three months it was found that the hyperkeratotic nodules and slight hyperkeratosis had entirely disappeared, and that the palms were covered with soft erythematous sweating skin. The soles remained as before. The vitamin A was increased to 200,000 i.u. daily for three months and then stopped. Six months later there had been no recurrence of hyperkeratosis on the palms, while the skin of the soles was softer.

Case 2.—A girl, aged 9, complained of thickening and deformity of the toe nails and bluish discoloration of the margins of the finger nails.

Past history.—Pneumonia, chickenpox, measles.

Family history.—Mother has thickened skin.

Examination showed a healthy girl with thickened skin over the toes and raised from the nail bed. On the fourth and fifth fingers the "claw" formation (Fig. 2). No sign of fungal infection or monilia were found microscopically.

Treatment.—Vitamin B, 1 mg. t.d.s., was given. Vitamin A, 30,000 i.u. daily, was then added to the diet.

One month later: within two months improvement, the nails had become normal, excepting the fingers and toes. Slight hyperkeratoses. The thickened skin was unaffected.

Comment.—It is, of course, possible that both of these conditions are due to vitamin A, and that the improvement observed was due to the improvement of the diet described by Brooke (1891). In Case 1 it is possible that the condition persisted five years' treatment by our
ACQUIRED LOCALIZED KERATOSIS PALMARIS ET PLANTARIS

Past history.—Pneumonia, chickenpox, measles and mumps.

Family history.—Mother has thickened skin over pressure areas on the soles.

Examination showed a healthy girl with no ectodermal or other defects apart from the toe-nails and thickened skin over pressure areas on the soles.

The distal ends of all the toe-nails were thickened, deformed, of a yellowish colour and raised from the nail bed. On the fourth and fifth toes there was a tendency to have "formation (Fig. 2). No sign of fungus infection was seen clinically, and no pus or monilia were found microscopically.

Treatment.—Vitamin B, 1 mg. i.d.s., was tried for one month without effect. Vitamin A, 30,000 i. u. daily, was then added, and increased to 75,000 i. u. daily one month later; within two months improvement was noted, and after four months the nails had become normal, excepting the free margin of the right big toe, which showed slight hyperkeratoses. The thickened skin on the soles, however, remained unaffected.

Comment.

It is, of course, possible that both of these patients would have got better without vitamin A, and that the improvement while taking it was a coincidence. It is known that localized hyperkeratosis on the palms, when appearing for the first time late in life, may respond to simple remedies, as in the case described by Brooke (1891). In Case 1, however, the nodules on the palms resisted five years' treatment by ourselves and others with keratolytic

Fig. 1.

Fig. 2.

appeared, and that the palms were covered with their supernumerary skin. The soles remained as before. The vitamin was given for three months and then stopped. Six months later there was hyperkeratosis on the palms, while the skin of the soles had returned to normal. The distal ends of the toe-nails showed the same condition, with the exception of the right big toe, where slight hyperkeratoses were noted. The improvement in the nails was noted within two months, after which the treatment was continued for a further month. The skin on the soles remained unaffected, while the palms showed a return of the condition described above.

Fig. 1.

Fig. 2.
and other ointments, and the malady had been present on the feet, and on the palms in a modified form, since birth. Although the palmar lesions which corresponded to vitamin A were of recent origin, the disorder was clearly of the congenital type, which is generally resistant to any kind of therapy. Many (1949), however, gave vitamin A to a mother and children with congenital tylosis and noted that the palms became softer, though there is no mention of the hyperkeratosis disappearing.

The second case was probably one of pachyonychia congenita, although we have referred to it as "acquired" because the nail changes began later than usual. Wright and Guequiere (1947), in describing two cases of this nature, say that they found vitamin A and emollients of value in treating the dry keratotic skin in an infant, but apparently the nails were unaffected.

It is noticeable that the hyperkeratotic nodules in Case 1 and the nail changes in Case 2 were of comparatively recent origin and short duration for such disorders, and were actively progressive. Possibly these factors may help to account for the response to vitamin A.

It is not suggested that deficiency of vitamin A is an aetiological factor of importance in tylosis, but it is possible that the malady so disturbs the utilization of vitamin A that the process of keratinization is affected.

**SUMMARY.**

The beneficial effect of vitamin A in a case of keratosis palmaris et plantaris and in one of pachyonychia is described.

We wish to thank Dr. R. T. Brain for permission to publish the second case.

**REFERENCES.**


**LEPRA, PSORIASIS, OR THE WILLAN-PLUMBE SYNDROME.**

**BRIAN RUSSELL, M.D., M.R.C.P.**

The ancient history of the condition known today as "psoriasis" has been obscured by the unfortunate terminological confusion that arose not only between the Greek, Latin and Arabic schools of medicine, but also between succeeding generations of physicians. The precise detailed description of management of the "rising, scab or bright spot" in Leviticus XIII shows the age-long difficulty of differential diagnosis between various skin diseases, from the serious to the banal. Biblical leprosy (Zaraath), of which Naaman was healed by dipping himself seven times in Jordan, was probably the same as our psoriasis, and the term "leprosy" used by Bateman in 1817 for the first cases, an eruption apparently identical to macular leprosy, psoriasis, squamous eczema (psa = the itch) was apparently applied to lepra. Paulus Aegineta, early in the disorders as "characterized by a roughness of the skin..." Another term, alja = (white), which was apparently used to be applied to vitiligio and leprosy. Hippocrates, however, regale, "and, probably referred to lepra as "generally believed to be the most fatal kind." The (white) was applied to a slowly developing disease, with very white scales, and without pigmentation. Lepra nigricans of cold, damp, and a precocious or improper name, "lichen," terminological "confusion not psoriasis." Robert Willan (1808) gave the first clear distinction, under the title "lepra vulgaris," a more distinct form and character adopted "psoriasis," "which seems to be a type of psoriasis by a different appellation," which from the ulcerated psora, placed by the Greek and known as psoriasis guttata, psoriasis guttata correspond to those conditions in our term.