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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.
PACHYONYCHIA CONGENITA (JADASSOHN-LEWANDOWSKY) AND KRYLE’S DISEASE IN THE SAME PATIENT

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ABSTRACT: An 11-year-old boy with typical lesions of pachyonychia congenita (Jadassohn-Lewandowsky) type II and with keratic lesions histologically characteristic of Kryle’s Disease is described.

In 1906, Jadassohn and Lewandowsky described a 15-year-old patient with congenitally abnormal thick fingernails and toenails, keratoderma palmoplantaris, hyperhidrosis, disseminated follicular keratosis and lingual leucokeratosis. We were able to record 73 published cases through 1975.

Congenital pachyonychia is a disease with generalized dyskeratosis. Thickness of the nails is its most constant manifestation. It is probably inherited in an autosomal dominant pattern. The largest series was published by Kumer and Loos covering 23 cases in 5 generations. Akeson reported another important series, covering 18 cases in 6 generations. Several other series of 5 to 7 affected members in 3 generations have been published.

Aside from these alterations, association with dystrophic hair, xerosis, multiple steatocysts, epidermoid cysts, gingival or premature eruption of the teeth, corneal alterations and nasal deformations have been described. The association to mental retardation and cataracts has been considered as incidental.

Case Report

An 11-year-old boy was admitted to the Department of Dermatology of IMAN Children’s Hospital because of cutaneous lesions of palms and soles and deformed nails. The patient’s parents were both normal, Mexican, mestizo, not consanguineous. The family history was negative for the disease. The propositus had 2 apparently healthy sisters and a brother. The mother had 9 pregnancies, 5 of which resulted in abortion in the first trimester. The propositus was the premature product of the third normal pregnancy, of 32 gestational weeks. Delivery was normal. Weight at birth is unknown.

Two weeks after birth, the mother noted a yellowish color in the fingernails and toenails of the patient. Soon after, the child’s nails thickened, especially in their free edges, and became convex.

At age 3 the patient walked, began to speak and was continent. At 4, the boy’s skin thickened in palms and soles. The mother thought that keratosis in the feet was due to walking. A year later, she noted hyperkeratotic patches with horny plugs in the center over elbows, buttocks, lumbar region, and knees. Four years ago and thereafter, he developed bullous lesions and painful grooves on both soles that secondarily became infected and made walking a difficult task. Three years ago, the mother noted a whitish plaque on the medial portion of the tongue which gradually spread (Figs. 1–3). The patient had bronchopneumonia and jaundice at 3 days of age. At age 7, he fell 15 meters down and was unconscious for 3 days; he had a fractured elbow and femur.

School performance was deficient.

The patient weighed 57 kg and was 1.52 m tall. He had first and second degree caries, aside from his cutaneous lesions. The tympanic membrane of the left ear was retracted due to a scar. The following tests were within normal limits: com-
COMPLETE blood count, urinalysis, skull X-ray, roentgenographic study of intestinal transit, EEG, and audiometry.

Speech tests suggested that alterations in voice and language rhythm were due to the social and cultural level of the propositus and to his sentenceism from school.

Genetic study suggested a de novo mutation with 50% chance of inheritance. The ophthalmologic study revealed small scattered pin-point precipitates in both crystalline lenses at the periphery; the test was not significant for cases. IQ was of a normal slow intelligence. The bone in both feet showed demineralization probably due to partial immobilization.

Biopsies of 3 keratotic cutaneous lesions buttocks had similar alterations: epidermis and mild acanthosis and papillomatosis. Large keratinous plugs were seen, showing hyperkeratosis with ortho and parakeratosis penetrating the epidermis to the point of breaking it and raising the dermis. The dermis showed mononuclear inflammatory infiltrates. The lesions were related to follicular structures (Fig. 4). A biopsy of lingual leukokeratosis showed acanthosis, papillomatosis, excessive hyperkeratosis, parakeratosis, lymphomonocytic infiltrates, and dilated capillaries in the superficial dermis.

**Discussion**

In the literature, we found 30 published cases of Kyrle’s disease throughout 1975. Some have been family cases suggesting a hereditary origin. The etiology is still unknown. Kyrle’s disease is more common in women over 50. Clinically, the lesions are cone-shaped papules that develop a horny mass which penetrates the skin and arc by an erythematous halo. In weeks, the horny plugs disappear a scar depression. Lesions may affect any part of the body.

The evolution is chronic.15

Histologic examination shows keratotic and parakeratotic epithelium in the epidermis at an angle as well as in parafollicular areas. Under the pressure of the horny mass, the spicule and basal layers break down and disappear. Through this horn-filled mass penetrates the skin until it is in contact with the dermis. In our patient, the altered nails and tongue are characteristic of pachyonychia congenita or Lewandowsky syndrome. A Kumer and Loos,2 there are 2 types of pachyonychia congenita: pachyonychia congenita with hyperkeratosis palmo-planteris with fortes of the body2, (Riehl type I) but with leukokeratosis palmo-planteris. (Riehl type I but with corneal calcification and leukokeratosis). Our patient’s case is interesting because histologically, the keratin plugs of chest and buttocks are “hy...
Hyperkeratotic and ulcerated areas

of the propositus and to his sebaceous glands of the nose as well as to histology, and to the patient's history. The lesions were not familial, and no familial history was obtained. The patient has been followed for 10 years, and no new lesions have developed.

Histologically, the lesions are characterized by the presence of hyperkeratotic plugs that invaginate into the epidermis at the follicular level, as well as in parafollicular position. Under the pressure of the plug, the prickle and basal layers become thinner and disappear. Through this pathway, the horny mass penetrates the epidermis until it is in contact with the dermis.

In our patient, the alterations in toenails and tongue are characteristic of pachyonychia congenita or Jadassohn-Lewandowsky syndrome. According to Kummer and Loos, there are 3 different types of pachyonychia congenita: pachyonychia congenita with hyperkeratosis palmo-plantaris with follicular keratoses of the body; (Riehl type), like type I but with leukokeratosis oris; like type I but with corneal changes.

Our patient's case is interesting because histologically, the keratotic lesions of chest and buttocks are "hyperkeratosis follicularis et parafollicularis in cutem penetrans," or Kyrlé's disease. This association had not been described before. It is possible that the lack of histologic studies in other patients with keratotic lesions similar to those of our patient prevented observation of their nature.

The keratotic lesions in the chest and the buttocks of our patient were clinically and histologically characteristic of Kyrlé's disease. We quote the opinion of Pinkus, to whom we sent the histologic slides of the patient: "The lesions of the boy obviously deserve the name of hyperkeratosis in cutem penetrans. Actually, they are the best example that I have ever seen of what I believe Kyrlé must have described."

The fact that the lesions are not directly associated to hair follicles and that the follicles observed looked normal, eliminates the possibility of perforating folliculitis. The case is not typical of Kyrlé's disease if we consider the age of the patient and location of the lesions.

The benign lingual leukokeratosis in our patient showed similar changes to those described in other similar cases of pachyonychia congenita.
study of nails was not carried out; in other cases, abnormal keratinization of the terminal matrix\(^{20}\) and of the proximal matrix\(^{21}\) was reported.

The treatment for nail lesions in pachyonychia congenita has been surgical with variable results.\(^8,22\) Joseph\(^{21}\) reported good results with topical applications of 3\% iodochlorhydroxyquinoline in chloroform.

References


Sebaceous Glands

Sebaceous gland activity may also be influenced by caloric intake. Sebum production was measured in fasting subjects, and diminished secretions occurred 2-4 weeks after the onset of fasting. The composition of surface lipids was also altered. Lower caloric intake resulted in a reduction of the triglycerides, fatty acids, wax esters, cholesterol, and cholesterol esters. However, no significant change in squalene occurred. In a similar study a change in the sebaceous gland lipids of the forehead in humans (occurred) after 5 days without food. Decreased synthesis of all lipids occurred except for squalene, and this change was reversed by returning to a normal diet.—Marzulli, F. N. and Maibach, H. I.: Dermatotoxicology Pharmacology. New York, Halsted Press, 1977 p. 107.

ABSTRACT: A mild dermatitis pro
to Cheyletiella yasguri was obse
ted when the dog was in a
to this parasite. Cheyletiella mites c
topically from sarcoptic mit
cause canine and human scabies.
gran and dog with 1% gamm
hexachloride is usually successful.

The medical literature is req
reports of human scabies result
increased physician awareness a
dosis of this malady. The dia
human infestation with mites af
found on other animal hosts
made. Canine scabies in h
caused by Sarcoptes scabiei car
morpologically identical to th
mite.\(^2\) While dermatis c
ectoparasites of Cheyletiella sp
scribed, most reports of this ha
occurred in the Europea
ure.\(^2\) We report human an
infestations with Cheyletiella sp
New Mexico, and consider its
and treatment.

Case Report

One of the authors of this report is a
an 8-week-old male puppy. After sev
the author's wife developed a pruritic
abdomen, chest, breasts, and neck. W
were tiny papules, 2-3 mm in diameter
surrounding erythema. Several days a
author developed a similar eruption
terior torso.

The pup was noted by his previous a
a veterinarian to have "dandruff" or t
This had been treated with milk a

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