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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.
A FAMILIAL CASE OF JADASSOHN-LEWANDOWSKI SYNDROME


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Pachyonychia congenita (Jadassohn-Lewandowski syndrome) is a rare genodermatosis characterized by thickening and change in color (yellow, brown) of the nail plates, keratodermia symmetrica (palmaris et plantaris), focal hyperkeratosis on the extremities (chiefly in the area of the knee and elbow joints), hyperkeratosis follicularis of the epidermis, hyperhidrosis, leukokeratosis, and pilar and dental anomalies. These alterations may be accompanied by mental disorders, microphthalmia, corneal opacity, cataract, blindness, etc.

Most authors now classify this syndrome, described by Jadassohn and Lewandowski in 1966 [sic], as an ectodermal dysplasia and categorize it among hereditary diseases with varying types of inheritance (N. D. Sheklakov et al.; K. M. Suvorova et al.).

Three basic types of the syndrome are distinguished with regard to degree of affliction (Komer and Loos; Vermenouze): (1) pachyonychia in combination with focal symmetric keratoses of the palms and soles; (2) pachyonychia, symmetric palmar-plantar keratoses and leukoplakia; (3) pachyonychia, diffuse cutaneous keratoses and leukokeratosis, in combination with anomalies of the skin, teeth, and skeletal system and mental retardation.

Patients with this dermatosis have been described by I. Ya. Markin; N. D. Sheklakov et al.; K. N. Suvorova et al.; N. D. Sheklakov, N. A. Yegorov et al.; V. A. Grebennikov and R. A. Chalimova; and A. V. Khamaganova and Ye. I. Shakhtmeyster.

We have observed 4 patients with Jadassohn-Lewandowski syndrome in one family.

Patient M., 22 years of age, presented on 1/27/1977 complaining of changes in her nails, formation of pustules under the nail plates, their tenderness, and focal keratinizations on the palms and soles. Early loss of teeth was also noted.
Soon after birth the infant was found to have yellowing of the nail plates and their accelerated growth with thickening of the free borders. As she grew older, foci of keratinization appeared on her palms and soles, for which she was given outpatient symptomatic treatment.

She was born the second child. Her growth and development were unremarkable. She had an occurrence of pneumonia during childhood. Menstruation at age 15, regular, lasting 4 days. She married at age 18. Three pregnancies and 3 deliveries. 8th grade education. The patient’s father died of pneumonia at age 43. Her mother had been an alcoholic since age 35 and died of cirrhosis of the liver. No data on disease similar to the patient’s could be established in her parents.

A patient of normal constitution, with adequate nutrition. Skeletal system normal. No superficial lymph nodes palpated. No anomalies found in the internal organs.

There were isolated foci of keratinization of a dense consistency, yellowish in color. A drastic change of the hypertrophic type could be seen in the nail plates, and they were of a dingy yellow color (Fig. 1).

[Fig. 1. Left hand (a) and feet (b) of patient M., 22 years. Jadassohn-Lewandowski syndrome. Onychogryposis.]

Complete blood count: erythrocytes 4,100,00; hemoglobin 127 g/L; leukocytes 6600; eosinophils 3%; polymorphonuclear 2%; segmented 65%; lymphocytes 24%; monocytes 6%; ESR 35 mm/hr.

Our patient’s three daughters (Nadya, age 5 years; Zhanna, age 1 year 8 months; and Inna, age 8 months) suffer a similar ailment (Fig. 2).

[Fig. 2. Genealogy of patient M., 22 years of age, with Jadassohn-Lewandowski syndrome. Autosomal dominant type of inheritance. White square and circle indicate healthy males and females; hatched square and circle, deceased; black circle with arrow to left, observed patients; black circle with arrow to right, proband.]

All of the children were found at the time of delivery to have yellowish-colored nail plates, subsequently with accelerated growth of the hypertrophic type. Their physical development was appropriate for age, but they all were seen to have retarded hair growth, leukoplastic alterations on the tongue, and a Gothic palate.

Complete blood count for Nadya M., age 5 years: erythrocytes 4,200,000; hemoglobin 110 g/L; leukocytes 13,800; eosinophils 1%; segmented 58%; lymphocytes 34%; monocytes 6%; ESR 34 mm/hr. Complete blood count for Zhanna M., age 1 year 8 months: erythrocytes 4,000,000; hemoglobin 120 g/L; leukocytes 8900; eosinophils 3%; polymorphonuclear 1%; segmented 47%; lymphocytes 40%; monocytes 7%; ESR 40 mm/hr. Complete blood count for Inna M., age 8 months: erythrocytes 4,320,000; hemoglobin 120 g/L; leukocytes 13,000; eosinophils 1%; polymorphonuclear 2%; segmented 55%; lymphocytes 36%; monocytes 6%; ESR 31 mm/hr.

It should be noted that all of these patients in their blood counts had an accelerated ESR in the 31-40 mm/hr range.

Thus, the clinical findings in these observations correspond to Jadassohn-Lewandowski syndrome, with clear inheritance from generation to generation.

All of the above provides a basis for classifying this syndrome as a monogenic genodermatosis with an autosomal dominant type of inheritance.
REFERENCES


Komer and Looz, cited by R. S. Babayants in *Dermatologicheskaya Sindromologiya* [Dermatological Syndromes], Moscow, 1974.


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