Pachyonychia Congenita Project

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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.

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PACHYONYCHIA CONGENITA (JADASSOHN AND LEWANDOWSKY). Presented by DR. FOX.

S. W., a boy, aged 3 years, 9 months, born in the United States, of American parents, presented peculiar anomalies of cornification affecting the nails and cutaneous surface. The lesions of the nails had been noticed two weeks after birth, those on the skin when the patient was almost 2 years of age. At the age of 1 year, the child was said to have suffered from an enlarged thymus gland which had interfered with breathing and which had been treated by the roentgen ray. He had suffered from diphtheria shortly before the appearance of the cutaneous lesions. The patient was an only child of young, healthy parents. A similar disease was not present in any of his relatives, including twelve first cousins on the maternal side, and four on the paternal side. Examination showed the patient to be a well developed, mentally active, fine looking child, weighing 42 pounds (19 Kg.). The eyes were blue and the hair was blonde. All the finger-nails and toe-nails were abnormal. The finger-nails were of normal width but were greatly thickened, the thickness increasing toward the free borders. The surface was smooth, and did not present pits, ridges or furrows. There was some diminution of luster. The nails were grayish brown, being increasingly discolored toward the distal ends where there were some whitish streaks on the nails of the left hand. The plates of the nails were extremely hard and firmly attached to the bed of the nails. There was neither paronychia nor abnormality of the skin of the fingers. There was not any hindrance to the performance of fine movements. The rate of growth of the nails was normal and the direction straight. The toe-nails showed changes similar to those of the finger-nails with the exception that the growth, if uncut, was in a curved direction (onychogryphosis). Subjective symptoms were not caused by the changes in either finger-nails or toe-nails. The scalp was normal. There was not any hyperhidrosis of the face, and nothing to suggest granulosis rubra nasi. At the right angle of the mouth there was a crusted area, apparently a simple herpes; the mother said that the patient often had "cold sores" in this location which lasted several weeks. The tongue was coated habitually, but there was nothing to suggest leukokeratosis. The teeth were in good condition and did not show abnormalities. On the extensors of the forearms, there were keratoses from the size of a pinpoint to the size of a pinhead. Over each olecranon there was an irregularly shaped warty mass the size of a bean surrounded by small, warty spinous lesions the size of a pinhead. On the lumbar and sacral regions, there were fairly numerous, grouped, small lesions the size of a pinhead which were rough to the touch, and the largest had red bases and filiform projections. The genitalia were apparently normal. The lower extremities were affected chiefly above the knees and to a less extent on the anterior aspects of the legs. The buttocks showed a similar condition to that of the lumbar region, but with more marked filiform spines. The thighs showed a fairly profuse eruption of follicular keratoses from the size of a pinpoint to the size of a pinhead, the largest having filiform spines. The skin between all of the described, warty lesions was smooth and normal in appearance. On the internal aspect of the great toes, metatarsal regions and heel of the right foot, there were yellowish callosities from the size of a pea to the size of a bean, under which were painful bullae. On puncture of a bulla, clear fluid of alkaline reaction was obtained, and was found to be sterile on culture.

PORT WINE NEVUS TREATED WITH GRENZ RAYS. Presented by DR. MALONEY.

H. P., a girl, aged 3 years, born in the United States, presented a port wine nevus on the left side of the face, extending from the chin to the ear over the inferior maxillary bone. She had had one treatment with grenz rays over the middle third of the nevus four months before presentation, and two treatments over the upper third of the nevus — one four months and the last one five weeks prior to presentation. The first area showed a decided loss of color, and the second area still showed the usual erythematous reaction following the application of grenz rays.

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PORT WINE NEVUS TREATED WITH GRENZ RAYS. Presented by DR. MALONEY.

I. G., a man, aged 43, born in Austria, manager of a restaurant, presented a port wine nevus on the left side of the face, covering most of the left side of the nose and cheek. This nevus had been treated over four separate 1 inch square areas by the grenz rays. One area, on the left temple, had been treated four times; one on the nose three times; one on the cheek under the left eye three times, and one lower down on the cheek twice. All of the treated areas showed a decided lessening in color as compared with untreated areas.

DISCUSSION

DR. MACKEE: I am impressed with these results. There is a definite improvement in each patient. The improvement in the man is remarkable because adult cases are even more stubborn than when occurring in children. This may prove to be the therapeutic method of election, especially if time demonstrates that sequelae never develop subsequent to these repeated strong erythema doses. I hope that Dr. Maloney will demonstrate these patients once or twice a year for the next few years.

DR. ELLER: There has certainly been great improvement and I too would like to see these cases again.

A CASE FOR DIAGNOSIS. CAPILLARY DILATATION. Presented by DR. BECHET.

H. G., a woman, aged 37, had had a dermatosis limited to the face for seven years. It consisted of a peculiar "blush-like," pinkish red, symmetrical patch on either side of the nose and extending to the chin. On close inspection in good daylight, the patches seemed made up of a capillary network. The color was permanent, and there had never been any remission, but it was greatly heightened by alcoholic or dietetic over indulgence. The patient had not suffered from the prevailing cosmetic habit. She had been under observation off and on for three years, and during that time there had never been any change in the appearance of the lesion. There had not been the slightest scaling, or any papule, erythema, or rosaceous outbreak at any time, either in the affected region or elsewhere on the face. Definite telangiectasia was not discernible, except on the cheeks, where there were a few telangiectatic vessels. There was a faint pinkish nevus, about 1 inch (2.5 cm.) in diameter, on the back of the neck, which had been present since birth.

DISCUSSION

DR. WISE: I could not make any diagnosis other than rosacea.

DR. WILLIAMS: I would like to know if there is any history of nose bleeding in the patient or in any of the family. Many of these curious dilatations of blood vessels which develop after infancy are in reality instances of an extremely mild type of hereditary hemorrhagic telangiectasia.

DR. HIGHMAN: I do not agree with Dr. Wise. It is not rosacea. It is a congenital anomaly of the capillaries.

DR. ELLER: I think that one might consider the diagnosis of rosacea, with a mild seborrheic eczema in this case.

DR. WHITEHOUSE: I agree with Dr. Wise.

DR. HIGHMAN: I do not see that the condition can be anything else but a disease of the capillaries of the skin that has not been identified. The striking part of the distribution of the lesions on the face is the curious linear marking, beginning near the nose and running down the cheek in a distinct band; as distinct as one sees, for example, in acrodermatitis on the forearm. I think that the girl has telangiectasia all over the face — though, of course, this might also be true in rosacea. She has a curious distribution of birth-marks at the nape of the neck and another at the root of the nose. In other words, she was evidently born with a tendency to enlargement of the capillaries; and whereas I