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PACHYONYCHIA CONGENITA

Regression of Plantar Lesions on Patients Wearing Specially Made Rubber Base Foot Mold and Shoes

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NEW YORK

An interesting therapeutic result is herewith reported. A patient with large callouses and bullae on the soles due to pachyonychia congenita was able to walk with comfort immediately on wearing specially fitted rubber base foot molds. There was gradual and steady involution of the lesions so that by the time of writing, except for the flat and diffuse calluses on the heels, only one cherry-sized callus remained on each metatarsal arch. Each of these was also gradually undergoing involution after the depressions in the foot molds corresponding to the sites of these painful lesions had been deepened.

REPORT OF CASES

Case 1.—History.—L. H., a young man aged 18, was first seen at the Department of Dermatology and Syphilology of the New York University Post-Graduate Medical School and the Skin and Cancer Unit of University Hospital on Jan. 23, 1941. His general health had been good. His mother had a similar congenital skin disease, and her case is reported here as case 2. His sister and her child had no such disorders. There was no history of consanguinity of his parents or grandparents.

His mother noticed one week after his birth that the middle finger of his right hand was discolored. Within a month all the nails of the hands and feet became yellowish. The mother also noticed soon after birth that his tongue was "coated at all times." A white patch developed on the buccal mucous membranes as his teeth erupted. When he began to walk, very painful erythematous patches appeared on the soles. These patches formed into blisters and then developed into calluses which gradually enlarged. The process had reached its height when the patient was 6 years old and had been the same ever since. He was able to walk only with great difficulty and pain because of these blisters and thick calluses, which were located particularly on the balls of the feet and toes. The hard calluses had to

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K. (a) Wise, F.: Pachyonychia Congenita with Keratoderm of Palses and Soles and Leukoplakia Oralis, Arch. Dermat. & Syph. 58:608 (Nov.) 1948; (b) Pachyonychia Congenita (Keratoderm Palmare et Plantaris; Dystrophia Unguim et Leukoplakia Oralis), J. Bid. 66:610 (Nov., pt. 2) 1949; (c) Gurt, J.: Pachyonychia Congenita with Keratoderm of Palses and Soles, Dystrophia Unguim et Leukoplakia Oralis, Considerably Improved by Specially Constructed Rubber Base Foot Molds, to be published.
be pared with a razor about once a month in order to raise the pain on walking, because at his dermatologist the patient was unable to go to school from the age of 9 to 12.

Dermatological Examination.—The patient had extensive, almost symmetrical, well-defined, yellowish, very thick keratic lesions on the heels, soles, and toes (Fig. 1), which were tender. There were no calluses on the hands. The nails of the fingers and toes were elongated, curved, and discolored. The distal ends were eroded and some thickened to about 0.5 cm. There were continuous leukoplakic patches on the buccal mucosa along the interdental lines posteriorly, tapering off gradually toward the buccal commissures. The teeth appeared in good condition except for some caries and fillings. There was gingivitis of the front and sides of the upper
Pain on walking. Nails from the age
symmetric, well
hairs (fig. 1),
all of the fingers

Fig. 2.—The imbeds provided for L. H. (case 1).

The color index was 1.2. The leukocyte count was 7,250 cells with a differential
count of 35 polymorphonuclear neutrophils and 20 lymphocytes. The Maizn
reaction was negative. The basal metabolic rate was −9 per cent. Total
erythrocytes and vitamin A content were normal. Findings and slit lamp examina-
tions of the corneas did not reveal any observations of significance.

Fragment.—Topical extract, 1/4 grain (0.01 Gm.) once a day, was given con-
comitantly with vitamin A, 25,000 units three times a day, for six weeks until
Jan. 21, 1947. From Feb. 19, 1947, to May 25, 1948, the patient took vitamin A
17.500 units), vitamin E (30 mg.), a preparation containing the vitamin B complex and thyroid extract (64 grains [3.954 mg.] daily). Folic acid and placebo, 40 per cent, was applied to the calluses on the feet every other day. Room of podophyllin, 20 per cent, in a vehicle consisting of eucamol oil, aquaphor® (an unsaturated-petroleum distillate base) and petrolatum, was applied experimentally to two lesions on the soles for one month without any improvement.

"Other" was used for six weeks without any noticeable change in the lesions.

Fig. 5.—Photograph taken May 30, 1949, after the patient had worn the rubber base foot molds for a year, showing the regression of the growths and bullae.

On May 25, 1948, the patient was instructed and begun to use specially made rubber base foot molds supporting the entire soles and heels. They were made according to foot prints on which the calluses were outlined. Medium hard resilient rubber was used to fill in the cavity and the base to support the heels and soles. The foot prints were made on paper before the application of the molds. The patient was instructed to wear the molds for 24 hours each day, and to wear them every night after getting well. During daytime, the molds were removed and the lesions treated with a suitable solution for a few minutes. The rubber was removed with a rubber ointment. The expectorant was not used. The symptom was seen in the patient for three years. The foot prints were made before the application of the molds. The calluses were removed and the rubber base rubber was removed with a rubber ointment. The expectorant was not used. The symptom was seen in the patient for three years. The foot prints were made before the application of the molds. The calluses were removed and the rubber base.

rubber was used for the base of the molds. The pressure areas were then traced out of the rubber, cups being formed corresponding to the callouses shown in the last print. The full molds were balanced to adjust for weight bearing and then were covered with soft pliable leather.

The inner soles of the shoes were also grooved with a hydraulic pressure machine to give greater cupping and pressure over freedom from pressure at each scene pressure peak. The molds were then set in the shoes and cemented to

Fig. 4.—Photograph taken on Jan. 23, 1947, showing the calluses of the mother, with the callus anterior to the left heel which recurred in the full thickness skin graft taken from the left calf (arrow). The callus in the graft still has a few hairs growing.

given shifting. These molds served the purpose of relieving pressure on the calluses and transferred part of the pressure to the longitudinal arch.

As soon as the rubber molds were used, the patient felt relieved and was able to walk fairly comfortably. By September 1948 three boot-sized and larger calluses on the right side, a cherry-sized callus on the base of the left little toe and several blisters on the soles had disappeared.
In order that the patient might obtain still greater relief at pressure points on the soles, new shoes were specially made on Sept. 22, 1948, with enough height to accommodate a field of at least ½ inch (9.54 cm.) in thickness, thus allowing the pressure points to be more deeply hollowed and obtunding the necessity of grooving the inner soles of the shoes. The wearing of this pair of shoes resulted in regrowth of all the lesions except two large protuberant growths, one on the middle third of the foot and the other on the inner third of the right and left metatarsal arches respectively (Fig. 3).

Case 2.—S. H., the mother of the patient in case 1, suffered from phrynoglyphic onychogryphosis of the fingers in both hands, and was treated for some time with a solution of ichthammol ointment. The nails became soft and could be easily lifted from their beds. The case showed no signs of improvement. The patient was then placed on a diet of hard-boiled eggs, milk, and bread. The nails began to grow again and became hard and strong. The treatment was continued for three months, and the nails showed no further improvement. The patient was then given a course of x-ray treatment, and the nails began to grow again and became hard and strong. The treatment was continued for three months, and the nails showed no further improvement. The patient was then given a course of x-ray treatment, and the nails began to grow again and became hard and strong. The treatment was continued for three months, and the nails showed no further improvement. The patient was then given a course of x-ray treatment, and the nails began to grow again and became hard and strong. The treatment was continued for three months, and the nails showed no further improvement. The patient was then given a course of x-ray treatment, and the nails began to grow again and became hard and strong. The treatment was continued for three months, and the nails showed no further improvement. The patient was then given a course of x-ray treatment, and the nails began to grow again and became hard and strong. The treatment was continued for three months, and the nails showed no further improvement. The patient was then given a course of x-ray treatment, and the nails began to grow again and became hard and strong. The treatment was continued for three months, and the nails showed no further improvement. The patient was then given a course of x-ray treatment, and the nails began to grow again and became hard and strong. The treatment was continued for three months, and the nails showed no further improvement. The patient was then given a course of x-ray treatment, and the nails began to grow again and became hard and strong. The treatment was continued for three months, and the nails showed no further improvement. The patient was then given a course of x-ray treatment, and the nails began to grow again and became hard and strong. The treatment was continued for three months, and the nails showed no further improvement. The patient was then given a course of x-ray treatment, and the nails began to grow again and became hard and strong. The treatment was continued for three months, and the nails showed no further improvement. The patient was then given a course of x-ray treatment, and the nails began to grow again and became hard and strong. The treatment was continued for three months, and the nails showed no further improvement. The patient was then given a course of x-ray treatment, and the nails began to grow again and became hard and strong. The treatment was continued for three months, and the nails showed no further improvement. The patient was then given a course of x-ray treatment, and the nails began to grow again and became hard and strong. The treatment was continued for three months, and the nails showed no further improvement. The patient was then given a course of x-ray treatment, and the nails began to grow again and became hard and strong. The treatment was continued for three months, and the nails showed no further improvement. The patient was then given a course of x-ray treatment, and the nails began to grow again and became hard and strong. The treatment was continued for three months, and the nails showed no further improve
and tender plantar calluses she had had a plastic operation performed on the left foot on Jan. 2, 1932, in the orthopedic department of another hospital. A quadrant of the transverse arch of the left sole was excised and a graft taken from the left calf. It was contemplated to graft skin for the entire soles if calluses did not form. However, a large callosity, 1.5 cm. circular, developed in this graft while two months (figs. 4 and 5). Some hairs are still present in this graft and even in the callus. When this therapeutic surgical attempt had proved of no avail, the patient was told by the orthopedic surgeon to stay home, relax, and keep off her feet.

On May 15, 1947, she was hospitalized at the Post-Graduate Hospital and put to bed. Within six weeks the calluses began to regress, but they recurred again soon after she resumed walking.

On June 10, 1949, she received specially prepared modil and shoes similar to those made for her son. The next day she stated that she could walk with fair comfort. Aluminum chloride solution, 25 per cent in distilled water, was recently prescribed for her to be applied three times a day to the feet for the purpose of counteracting the excessive sweating of the soles, thus enabling her to wear the shoes continuously at home unless she is resting. Tincture of belladonna in 5 per cent (0.30 cc.) does twice a day is occasionally given in addition to the local applications. There has been great and sustained subjective improvement (fig. 5).

**COMMENT**

Apparently in this congenital anomaly there are inherent weakness and lowered resistance of the skin and mucous membranes to pressure, irritation and friction. Because of these factors and because of the differences in the tissues, the abnormality is manifested clinically at the areas of greatest pressure, on the weight-bearing points of the soles, by calluses and bullae (fig. 1); in areas of irritation, such as in the nails, by hyperkeratotic changes, and in areas of frequent friction, as in the mucous membranes of the mouth and tongue, by leukoplakic patches.

That the susceptibility to callus formation is not limited to or dependent on the local disposition of the skin itself is dramatically illustrated in case 2 by the formation of the hard callus on the left sole in a full thickness skin graft removed from the left calf (figs. 4 and 5).

It seems logical to assume that for the patient in case 2, perhaps because the surfaces of her soles were considerably smaller than were those of her son, the results should be less spectacular. This outcome could be attributed to the smaller size of areas which can be made to transfer and distribute the pressure points.

It is obvious that these patients will be obliged to wear these costly, specially constructed modil and shoes for life and that every few months the footwear may have to be adjusted. Nevertheless this method certainly appears worthwhile, as is shown by the ability of the patient to walk with comfort after the modil were used. The younger patient is now a totally transformed young man both physically and mentally and is for the first time able to do a full day's work.
Similar casts with modifications to fit each individual case might be used in treatment of other disabilities of the soles, such as painful exostoses, tender plantar verrucae, acquired calluses that do not respond to treatment, painful synechiae, verrucae on the soles, and perhaps epidermolysis bullosa.

A partial review of the literature, including authoritative books on rehabilitation and orthopedics, failed to reveal that such molds as were fitted to these 2 patients had ever been used before.

SUMMARY AND CONCLUSIONS

Two cases are reported, in a mother and son, of pachyonychia congenita. Large calluses and blisters on the feet of these patients had defied every form of therapy applied and caused them to walk only with great difficulty and with considerable pain. They had to resort once a month to paring down the calluses with a razor. In the first case, that of the son, several calluses and blisters regressed within two months after the patient began wearing specially constructed molds. Further regression occurred later as a result of molds fitted in specially constructed shoes of sufficient height to contain molds ¾ inch (0.94 cm.) thick. These allowed for deeper grooving so that to date only one callus on each sole remains. These calluses were gradually undergoing regression, at the time of writing, the grooves corresponding to these growths having been deepened. The patient is now able to walk with little discomfort and to work steadily. The patient in case 2, the mother of the patient in case 1, with the same congenital anomaly, was given similar molds and shoes on June 10, 1949. She was able to walk with complete comfort at the time of writing. An additional topical remedy was recently prescribed for her to combat the excessive sweating of the feet, thus enabling her to wear the shoes consistently at home, particularly during the hot weather, as well as on the street.

That the predisposition to callus formation is not a local phenomenon is illustrated in case 2 by the formation of a large callosity in the great toe taken from the left call to replace the callused skin removed from the left sole. A few hazes are still visible in this callus, showing the independent viability of the flap from the callus.

From these observations it appears as though the skin and mucous membranes of patients with pachyonychia congenita may be congenitally weak or defective in their capacity to support pressure, friction and other irritations and lashed to respond to such local trauma with callusses, hyperkeratotic changes in the nails and leukoplakic patches.

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Mrs. Berenadette Kelley and the Social Service Department of University Hospital of the New York University-Bellevue Medical Center obtained the funds for the shoes for the mother and son.