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Scleroderma.*

BY

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A CASE OF SCLERODACTYLE WITH DIFFUSE SCLERODERMA.

BY H. C. GORDINIER, M.D.,
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As I can find no recorded American case of sclerodactyle, I report the following case, hoping it may contribute to a better understanding of the disease. But thirteen cases are recorded—one English, one German, three Scandinavian, and eight French.

August 16, 1888. Mrs. S. K., æt. forty-five years. Mother and father both died of phthisis; has a brother living, and in perfect health; another brother died several years ago of some cerebral difficulty, the nature of which I was unable to learn. Has three step-sisters living and in good health. The patient has had seven living children, one stillborn, and one miscarriage. Of her seven children, four are now living and in excellent health—none of them presenting the slightest trace of their mother's disease. Of the three children dead, one died from the results of a deep cellulitis of the neck at seventeen months; one died when seventeen days old, and one but a few days after birth. Of the seven births, four were breech cases. She suffered greatly during each confinement, and they were long and tedious.

Prior to the occurrence of her present illness, which first made its appearance in the winter of 1868, she had been in excellent health and had never seen a sick day since childhood. Her menstruation first made its appearance at the age of eighteen; she has always suffered considerable pain prior to the onset of the flow, and was usually ill for six or seven days, always losing too much blood. Her menstruation has continued with perfect regularity until two years ago, save in January of 1868, from which time she dates her present illness. Two years ago her menstruation appeared very irregularly, coming and going at intervals of from three weeks to as many months. Since January, 1888, until July 15, 1888, she had not menstruated. July 15th, was unwell, and continued so for seven days, losing a great deal of blood. At the date of this examination she is ill, losing more blood than ever before. She never has had hemorrhage from the nose, stomach, intestines, or bladder.

The disease first made its appearance in January, 1868. About the middle of January she went to a dance; while in a condition of profuse perspiration she sat exposed to a draught of air, and had a severe chill. She was menstruating at the time; this ceased. A doctor was called, and gave her a hypodermatic injection of morphia in the left thigh; in a few days was better and able to be about.

In the early part of April she first noticed, in walking, that her legs were getting stiff, at the same time pains, neuralgic in character, made their appearance in both legs, and ankles and knee-joints. The stiffness continued, and became very noticeable on going up or down stairs. Soon local feelings of cold appeared. This stiffness and pain became so severe that she sent for her doctor, who told her that she was suffering from rheumatism, and applied on the outer side of the right leg a large blister, the effects of which were very painful, and resulted in an ulcer, which did not heal for three months. She was, during that time, able to walk only with considerable difficulty. This impediment to locomotion was not due to the ulceration, but to the increasing stiffness and pain.

During the months of July, August, and September she was not able to walk and was moved about in her sick-chair; this she attributes to the too rapid healing of the ulceration. She began to walk a little in October. November 15th, gave birth to her first child; was unable to walk for eighteen months after her confinement. She then walked occasionally, but with great difficulty and pain. She has been able to walk a little about her house until seven years ago; since then has never walked.

Eighteen years ago the little finger and thumb of her right hand became swollen, very stiff, and cold; this condition continued from bad to worse, and the thumb and little finger became the seat of very severe pain. Two years after, the stiffness, pain, and swelling made their appearance for the first time in the thumb and index finger of the left hand, soon followed by local feelings of cold; very gradually the stiffness and swelling extended to the index and middle fingers of the right hand. The left little finger became affected next in order; the middle and ring fingers were the last to become involved with this stiffness and pain. She does not know when the infiltration and thickening of the skin first began, nor does she remember about the thickening of the skin of the face and its swollen condition; she states that her friends have frequently told her that her face was much bloated for several years.

In the winter of 1880, a large ulcer made its appearance on the dorsum of the right hand, occupying its entire surface, superficial and secreting a serous-like material, which did not heal for six weeks. The following winter an ulcer, having exactly similar characteristics, appeared on the dorsal surface of the left hand. This ulcer healed in three weeks. Neither ulcers left scars. There has never at any time during the existence of this disease been any ulcerations of the fingers, and no discharge of fragments of bone or bone dust has occurred. She complains bitterly of cold fingers, which are always made worse by the use of cold water. She dreads the winter very much; she is always pleased with the approach of warm weather, which brings with it marked relief to her sufferings of cold and pain. During cold, rainy days in summer the cold sensations return, and are immediately attended with severe pain. She was able to sew quite well until six years ago; since then, on account of her deformed fingers, can use a needle but very little. She can distinguish between very small objects, and, apparently, has no sensory disturbances. Plunging her hands in hot water gives considerable relief to the pains and cold sensations; but if, by accident, her hands be plunged in cold water, she suffers severe pain and the cold sensations remain a long time. There have been no scales or horny epidermic growths on the hands.

Examination.—Patient is short, rather stout, and weighs about 170 pounds. She is bright and intelligent; there is no mental disturbance, no affection of speech, no difficulty in swallowing; has an excellent memory. The skin of the face, forehead, neck, arms, forearms, thighs, and legs is thickened; that of the face is glossy, of a pinkish hue, and can be moved with great difficulty on the underlying tissues—this is especially marked with the skin covering the malar processes and that on the forehead. One cannot pinch the skin into folds. There are no wrinkles to be seen on the face or forehead, and that of the forehead does not wrinkle whether she laughs, speaks, opens her mouth, or tries to cough. The naso-labial folds are nearly effaced. The eyelids do not appear to be thickened and are not œdematous. The patient is able to open and close them in a normal manner; there is no overflow of tears. The ears are large and prominent, and look as if made of wax; they are universally thickened, which is especially noticeable in the lobes. Slight friction of the ears with a bit of cotton produces a pinkish hue. The skin covering the nose is very tense and glossy, particularly near its root; the *alæ nasi* are freely movable, and present no atrophic processes or ulcerations. The lips are cyanotic and thickened. The movements of the lower jaw are quite restricted by the thickening of the skin covering the rami and that of the chin and neck, preventing the patient from opening her mouth in a normal manner. The tongue is slightly coated, about its normal size, and can be protruded a little beyond the teeth. The gums are spongy and bleed readily when irritated. A number of teeth are missing, and those remaining are very friable. Her hair is becoming sparse, is brittle, and quite gray. Along the margins of the scalp and just over the eyebrows a scaly seborrhœa-like deposit is seen. This thickening and immovability of the skin, and the peculiar wax-like appearance of the face, with the absence of wrinkles and folds, give a remarkable expression. The countenance is changed and the face has a fixed immobile condition, as if covered by a mask.

The skin of the neck and chest is less thickened than that of the face, and more easily movable; it has a wax-like color and is glossy. Three large transverse folds are to be seen crossing the front of the neck. The lateral movements of the head are much restricted, while those of flexion and extension can be performed with ease. The patient is unable to raise her arms to a right angle with the body; can with difficulty touch her nose with her finger-tips, and cannot place her hands on her head. These restrictions are not due to any interference on the part of the normal action of the shoulder or elbow-joints, but to the hidebound condition of the skin covering the shoulders and arms, preventing the underlying muscles of these localities from performing their physiological functions. There is no ankylosis of the shoulder- or elbow-joints. The upper extremities remain in a condition of adduction, motions of supination and abduction cannot be performed. The skin covering the forearms is less thickened than that covering the arms and shoulders, and is a little movable; it cannot, however, be pinched into folds. It has a wax-like appearance. The wrist-joints are stiff, immovable, and ankylosed. There is a little lateral motion in the wrist-joint of the right hand. The skin on the front and back of the joint is indurated, tense, and thickened. The thickening of the skin about the wrist does not prevent one from finding the pulse, which is a little rapid, but of good strength—a number of counts give an average of

eighty-five beats per minute. Perspiration has nearly ceased; a little perspiration takes place about the neck and forehead during very warm weather.

There is universal tenderness on deep pressure over all the body save the abdomen. This deserves special mention, as the tenderness is not confined to those parts most diseased, but to those presenting the least sclerodermatous changes. The patient dreads to be pinched or to have too firm pressure applied on the surface. The hands look as if made of wax, and are quite puffy. On the dorsal surface of the left hand near the outer side of the wrist, a small white cicatrix is seen. The skin covering the hands is more movable and less thickened than that of any part of the body save that of the abdomen and feet; is puffy and wax-like. This puffiness is very marked on the dorsal aspect; the skin and underlying tissues in this situation are atrophic. The skin covering the metacarpo-phalangeal and phalangeal articulations is tense, thickened, stiff, and glossy, and not as movable as that of the dorsum. There is an entire absence of hairs on the hands, and they are sparse on the forearms. The ulcerations which the patient described as having occurred on the dorsal surfaces of the hands in the winter of 1880-1881, doubtless were large pemphigoid bullæ, as there can be found no trace of a cicatrix on either hand. As the deformities, distortions, and dislocations of the fingers of the right hand are a little in advance of those of the left, it will be first described. The skin on the dorsal surface of the hand is much less thickened than that of the forearm; has a wax-like color, is smooth, puffy, and swollen. The wrist-joint permits of slight lateral movements, but extension and flexion are prevented. A small conoidal tubercle is seen on the outer side of the hand near the wrist.

The points of most interest are the peculiar deformities, the apparent dislocations, and the atrophic changes of the phalanges. The thumb is fore-shortened, the terminal phalanx small, atrophic, and congested; the skin on the outer surface of the articulation with the second phalanx is thickened and atrophic, so that the terminal phalanx is flexed on the second, almost at a right angle. Movements of extension and flexion can be easily executed at this joint, while lateral movements are restricted. There is no ankylosis at the metacarpo-phalangeal joint, and movements of the joint are unimpaired. The skin of the index finger appears very puffy, particularly that covering the first phalanx; this, with the atrophic condition of the second and third phalanges, gives the fingers a marked spindle shape, which is fore-shortened: the motions of the metacarpo-phalangeal joint are free; movements of flexion and extension of the first phalanx are limited, but lateral movements are unimpaired. The third phalanx presents its normal length and thickness; the second is about half its natural length and thickness; the unguis phalanx is very small, its tip red; the articular surfaces between it and the second phalanx have undergone extensive atrophic changes, the phalanx is freely movable, slightly flexed in a lateral position, and dislocated laterally. The middle and ring fingers are of equal length and not much shortened; the skin covering the first phalanx of each is puffy, giving them the spindle form. They are freely movable at all the phalangeal articulations, the unguis phalanges of both fingers present the same redness as those of other fingers: the second phalanges are shortened, and the terminal ones over-extended. The little finger is very short, puffy, and flexed at an acute angle, the skin is thickened

on its outer side, the articulations are freely movable, and there is no stiffness. The unguis phalanx is about one-third its natural size. The nails of the fingers and thumb are small, brittle, and striated. The fingers look like wax, except their tips, which are red and congested.

Left hand. The thumb presents the same deformity as that of the right, but in a less degree. The thumb, as a whole, is flexed at a right angle at the metacarpo-phalangeal joint; its first phalanx is over-extended and laterally flexed. The index finger is more deformed than its fellow. The motions of the metacarpo-phalangeal joint are free; there appears to be a dislocation of the second phalanx to the outer side at its articulation with the first; the second and third phalanges are more extended and laterally flexed toward the middle

FIG. 1.

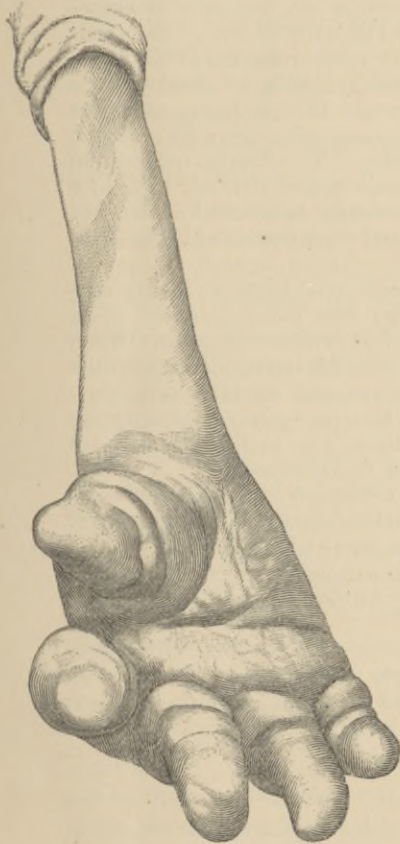


FIG. 2.



finger. The middle and ring fingers have the same appearance as those of the opposite side. They are of equal length, are freely movable in all directions. The little finger is much deformed, is fore-shortened: the second and third phalanges are small and atrophic; the terminal phalanges of fingers and thumb are like their fellows—red and con-

gested, and the nails are small and brittle. (Figs. 1 and 2 are from photographs of the palmar and dorsal surfaces of the left hand, showing deformed fingers and the position of hand.)

Looking at the hands placed close together, that which is most striking is their extreme smallness, due to the atrophic changes in the bones and joints. They appear symmetrical in the distribution of deformities and the conditions of articulations and general character of the skin. On close inspection, one sees that the disease is more advanced in the right hand, and that it presents, with few exceptions, the greatest deformities. The skin of the palms is not thickened; is of a pinkish hue, analogous to that of the ends of the fingers. The hands are habitually held in a sort of claw-shape; the fingers are flexed in a peculiar manner. The normal grooves of the palms are much exaggerated.

The skin of the feet is not thickened, and is freely movable; there are no deformities of the toes; deep pressure on the skin produces intense pain. They are constantly cold. The skin covering the upper parts of the legs is much thickened. Very limited motion at the knee, but a little flexion and extension remaining. Skin of chest a little thickened; that of abdomen appears normal and is freely movable. Heart and lungs normal; passes normal amount of urine, acid reaction, specific gravity 1010, no albumin, no casts. No affection of cranial nerves: muscular, thermal, tactile, and pain senses normal. Bladder and rectum normal. The tendon reflexes cannot be elicited on account of the ankylosed condition of the joints and thickened and hidebound condition of the skin.

Electrical reactions. Chloride of silver combined battery; faradic current; indifferent pole on nape of neck, other over motor point for facial nerve, in front of ear. No response with cylinder fully in: when withdrawn one inch, left facial nerve muscles respond with quick contraction; right facial nerve muscles respond equally well when cylinder is out one and one-half inches. Extensor muscles of forearms respond sluggishly when the tube is withdrawn two inches. Muscles of ball of thumb and the interossei give quick contractions.

Galvanic current. Left facial nerve, seventeen cells, K. C. C.: response being prompt of all muscles supplied by the facial. Right facial nerve, twenty-five cells, K. C. C.: response more sluggish than that of opposite side. Reversal of poles, with same number of cells, produced no reaction; but when thirty cells were introduced, left facial nerve muscles A. C. C.

Extensor muscles of forearms, with thirty cells, respond quickly to negative closing: positive closing, a slight contraction. There is no reaction of degeneration; a decided quantitative diminution in extensor muscles of forearms and those supplied by right facial nerve. Muscles of the legs respond in a normal manner.

As to etiology, very little is known. A very noticeable feature of the disease is the fact of its having developed, without an exception, in women. Distinct neurotic family histories were traceable in the cases of Hallepeau,¹ Ball,² Dufour,³ Rapin,⁴ and my own. In no case could

¹ Société de Biologie, December, 1872.

² Société de Biologie, June, 1871.

³ Société de Biologie, October, 1877.

⁴ Journal Bulletin de la Société Médicale, 1875, page 383.

a history of *syphilis* be elicited. Heredity could be traced in no case. Several of the patients had had children, but they never presented traces of the disease.

That which seems to play the most important part in the production of the disease is the action of cold. The starting-point of the disease in three patients was attributable to the sudden checking of perspiration, after exposure to draughts of air. In all cases the patients complained bitterly of cold; very much dreaded the approach of cold, rainy weather. When the disease had apparently been stationary during the warm summer months, it made rapid progress in winter. In six patients the disease first made its appearance in winter. All the patients were much worse during the cold season. Dufour's patient was treated by Rilliet with cold baths, which, instead of improving her condition, made her decidedly worse. Several patients have had recourse to hot baths, and especially of plunging their hands in hot water, with decided relief to all the symptoms.

The onset of the disease was characterized, in most cases, after exposure to the influence of cold, to repeated acute attacks in the hands and feet. The fingers and toes become the seat of intense lancinating pain, localized especially over the small joints; subjective sensations of numbness, tingling, burning, and formications, followed by circulatory changes; the toes becoming violet, very cold, and finally assuming a waxy appearance. This condition would last about half an hour, when gradually they would recover their normal sensibility and temperature.

After several of these attacks of pain, tingling, formication, local feelings of cold, and alterations in the circulation, in four cases, small bullæ made their appearance, especially over the phalangeal articulations and near the roots of nails, which, after a short duration, ruptured, leaving small superficial ulcers, which were very difficult to heal and often secreted pus freely; when healed, the skin covering them was often hard, indurated, sometimes pigmented, adherent, in most instances, to the underlying tissues, and often the seat of yellowish crusts. In three cases ulcerations occurred on the elbow-, wrist-, and ankle-joints, and, after healing, their places were occupied by yellowish, horny excrescences resembling species of epidermic corns.

The lesions of the hands are of most interest; they are symmetrically distributed in most cases. In most cases the deformities were more marked in the right hand. That which is most striking is their apparent smallness, which, in most part, is due to the atrophy and fore-shortening of the fingers. The skin of the dorsal surface, in most cases, is of a waxy color, with a yellowish cast, resembling old wax, is thickened, cannot be pinched into folds, and is usually adherent to the underlying tissues. In a few cases marked pigmentation was noticed. In three cases there was an absence of hairs on the dorsal surface of the hands and fingers,

and a scarcity on forearms. Ankylosis at the wrist-joint occurred in three patients. In a number of cases yellowish crusts were discovered near the wrists, and at times over the phalangeal articulations. The skin of the palmar surface has been found normal in regard to thickening and movability, but it was most generally of a pink color, rather swollen and puffy, and having the palmar grooves markedly accentuated.

The fingers are the parts most compromised. They present several deformities. Some are dislocated at their phalangeal articulations, being in positions of flexures; others freely movable. The phalanges were, in all cases, atrophied, both in length and thickness, producing marked fore-shortening. The skin covering the fingers, in most cases, has been described as waxy, thickened, in some cases adherent to underlying structures, usually quite puffy, particularly over the first phalanx, which, with the atrophic condition of the second and third, and the smallness of the nails, give the fingers a spindle form. The distal phalanx of the thumb, in three cases, was atrophied, flexed almost at a right angle with the second, and partially ankylosed in that position; the index, middle, and ring fingers have their terminal phalanges flexed, producing a sort of claw-hand; the little finger, in most cases, was markedly deformed, much shortened, its phalanges atrophied to an extreme degree, and the terminal one at a right angle with the second. The nails, in all cases, have been deformed; they are very small, sometimes a third their natural length, variously curved and hooked; friable, rough, and deeply striated in their long diameter.

In six cases the feet were the seat of pain, numbness, formication, local changes in the circulation, producing a violet, and later a waxy color. In three cases ulcerations and the formation of horny excrescences and yellowish crusts appeared similar to those in the hands; in all cases the patients complained bitterly of cold feet. In no case have the feet and toes become deformed to anything like the extent which occurs in the hands. In the cases of Dr. Radcliffe Crocker¹ the skin covering the dorsal surface of the feet was hard and indurated, but the toes were not affected in their movements. In Dufour's patient, after an acute attack in the toes such as described, they remained semiflexed, and could not be straightened. In three cases ankylosis at the ankle was observed. There is no sclerodermatous change in the feet or toes of the patient under my observation; the skin is freely movable, and not the least thickened, and no deformities of the toes exist. But a few weeks ago she suffered an attack of severe pain in the great toes, followed by vascular changes. During this attack the terminal phalanges of the great toes have both been dislocated inward and distinctly flexed, and at present no scleroderma exists. The perspiration in a few cases has been absent,

¹ Lancet, July, 1885, page 192.

and in all cases much diminished. The temperature is not recorded in other cases; in my case it has always been 99° F.

Six cases presented diffuse sclerodermatous changes in the skin covering the face, neck, upper part of the chest, upper extremities, and thighs. In the cases of Lienville¹ and Huchard² the neck and thighs escaped, and in Arnozan's sclerodermatous changes appeared on the back.³ The condition of the face is quite remarkable: no wrinkles can be seen, and emotional changes do not bring them out, thus giving to the patient a rather stupid appearance; face looks waxy and swollen, the skin is glossy, thick, and indurated, and but little movable. In a few cases the patients, because of this condition of the skin, were unable to open their eyelids in a normal manner, and the thickening and hidebound condition of the skin covering the lower jaw prevented the mouth from being opened naturally. In four cases the changes in the skin were confined entirely to the hands and fingers, and in the case of Haslund⁴ they were confined to the skin covering the second and third phalanges.

It is difficult to form an opinion from the observations of but one case as to the nature or primary origin of the disease. The number of autopsies and clinical observations are too few to determine with certainty the nature of the disease. There has been but one autopsy. The case of Hallopeau, after a duration of eleven years, died of phlegmonous erysipelas. An autopsy was made by Budin and Lagrange, which I copy from Senator's paper.⁵ Several autopsies have been made in cases of diffuse scleroderma. In Schimmers's case disease of the peripheral nerves was found.⁶ Westphal found pathological changes in the brain, and in a recent case of Eulenberg progressive facial atrophy coexisted with scleroderma. The mode of inception by intense pain, numbness, tingling, burning, and formication, with circulatory change, probably dependent on vasomotor changes, followed by the formation of blebs and ulcerations, very stubborn and difficult to heal, and the repetition of the attacks before the deformities make their appearance, and their symmetrical distribution, the nutritional disturbances in the small joints, the dislocations, the atrophic changes, marked local changes in the temperature, can best be explained by a *trophic* affection of the nervous system.

In two cases the changes in the bones and joints were explained by the pressure of the sclerodermatous skin. But after a careful study of the reported cases I am inclined to think that the trophic changes in the bones and joints precede those of the skin. As to the condition of the case here reported, I can state positively that there is no sclerodermatous

¹ Société de Biologie, 1874.

² L'Union Médicale, No. 93, page 372.

³ Journal Médicale, Bordeaux, 1881, page 97.

⁴ Medical Society, Copenhagen, March, 1886.

⁵ Berliner klinische Wochenschrift, August, 1888.

⁶ Scleroderma, Ziemssen's Handbook of Skin Diseases.

change in the skin covering the feet or toes, and that after a very severe attack of pain, followed by coldness and circulatory changes in the great toes, they both became dislocated and flexed inward.

Autopsy. Budin and Lagrange. In certain phalanges the cortical substance had disappeared, so that the cancellous tissue appeared to be in direct contact with the periosteum, in others the atrophy was less marked. Microscopical examinations in other bones, in which the process was of an older date, showed scanty, irregular infiltration of embryonic cells; about the vessels and in spots, aggregation of fat cells occurred, these again were more numerous in the long diseased and more atrophic bones than in those later attacked; the bony substance showed quite an irregular framework, with large mesh-work, which contained fat and embryonic cells, the latter especially heaped up in the borders of the framework or about the vessels, and more sparsely in the bones with marked atrophy, more numerous in the neighborhood of the joints. In the more recently diseased and less atrophied bones, the joints showed the same changes as are found in chronic arthritis: loss of cartilage, a disappearance of bone through fibrous, and in other parts connective tissue. In the spinal cord and nerve-trunks of the arms nothing abnormal was found. In the little finger of the left hand there was found in two or three places in the nerve-sheath an infiltration of embryonic cells.

Lagrange and Budin agree in regarding the changes as dependent upon an irregularly advancing chronic inflammation, which extends from the skin to the underlying tissues.

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