A SECOND COMMUNICATION

ON

SIMPLE ATROPHIC SCLEREMA.

BY

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LOUISA M—, a well-developed, unmarried woman twenty years of age, 5 feet 2 inches high, of fair complexion, with blue eyes and yellowish-brown (light auburn) hair.¹ She was formerly stout² and of a high colour, now she is thin, semianæmic, and, to a great extent helpless, on account of the condition of her hands. Her family history is moderately good. Both parents are living and in good health, the father forty-seven, the mother forty-three, and both have been free from disease and from tendency to rheumatism or dropsy.

Louisa M— is an only child, and her mother was twenty-five years of age at her birth. Her only paternal uncle died of consumption, "brought on by his own indiscretion." Her paternal grandfather died of "typhoid fever" at the age of

¹ The teeth and hair are strong and healthy, as in the former case.

² Her weight at different times is given at p. 106.

forty; her paternal grandmother died of "nervous" debility when over seventy years of age. On her mother's side the grandmother is still living and in good health, and the grandfather died only two years ago. One aunt had rheumatism severely at the age of twelve; she recovered well, but at the age of twenty had a second attack, which completely crippled her, and she was unable to close her hands. She died in her thirtieth year. For five years before her death she was completely blind, and for six months before the final event she was unable to move without assistance. Another aunt, married, but barren, is at the present time attending a hospital for a "tumour of the side." A married cousin (Lydia M—) was bedridden for some years after puberty and could not move without help.¹

To return to Louisa M—. She had variola badly when only a year old. At the age of two and a half years she had a severe attack of "gastric fever," this was doubtless enteric; she was in bed a month and had diarrhoea and bloody stools. Subsequently she had whooping-cough, and about the age of ten a slight attack of measles. As she grew up the health was moderately good. She was unable to bear much excitement, which produced headache; and deviation from plain diet caused indigestion with pain in the epigastrium. The catamenia appeared at the age of fourteen, and the functions were normal up to eighteen months ago, when there was suppression and its results—headache, light-headedness, flushing heats, and pain in the back for six months; since then the flow has been regular, but scanty, and preceded and followed by some leucorrhœa.

In May, 1875, she went from home, and was engaged for about six weeks making ice; her father discovering that she

¹ She died under the care of Dr. Ashurst, of Farningham, who has kindly sent me the following particulars:—"She came under my treatment for arthritic rheumatism described as following an attack of fever. The rheumatism persisted, and was accompanied by inflammatory attacks in the pleuræ, and hepatic congestion; and she ultimately drifted into an asthmatic condition, in which she died. The heart from the first showed great feebleness, but no valvular mischief. She was very corpulent. Her father died of dropsy; her mother of apoplexy."

was living badly removed her to her home after an interval of about four months. Excepting this interval she has always resided at home, where she has been engaged in the work of the house and lived comfortably.

The last few years she has been liable, at intervals of two or three weeks, to attacks of syncope—partial loss of consciousness for some seconds, rushing sound in the head, and deadly pallor, preceded by a flush of heat, and followed by a cold sweat, during which the hands would be quite wet. Some weeks she had had two or three of these attacks.

Two years ago, *i.e.* shortly after her return home, her health began to fail, she became weak, thin, low-spirited, and for a month the fingers were successively affected with sub-epidermic whitlows. After this affection passed off the fingers gradually became cold, stiff, and hard.

In July, 1876, while at the seaside, she had some swelling of the feet, and of one leg as high as the knee. The knee and toes were red and tender, and for two or three days she could not bear movement of the knee nor get her boots on. The whole affection lasted but a week.

The following is her present condition:—The hands are very cold and hard, the fingers semiflexed and almost fixed. They are rather shiny and tense, but are not enlarged, and the integument feels as if it were of the consistence of tendon; it is impossible to pinch together a fold, or to produce a wrinkle. On favorable days the colour is sallowish-pink, but on cold damp days the fingers have a leaden hue. On attempting to close the hands the knuckles become sallowish-white, and after the greatest effort the finger-tips impend over the palm at a minimum distance of an inch. The wrists are maintained straight; flexion and extension are much diminished, but the rotatory movements of the wrist are freer. The wrists ache, and all but the gentlest movements cause pain. A fine creaking is palpable during movement, when a finger is pressed upon an extensor tendon. The finger-tips are attenuated, and the nails strongly incurved from side to side, so as to reduce the interval between the edges of the nails of the little fingers to a quarter of an inch

and to three and a half tenths of an inch in the rest. The nails of the index fingers are not so incurved, but they are coarse from previous suppurative action around them. These features are slightly more marked in the right hand.

The tactile sense is slightly diminished, the fingers cannot anywhere discriminate the two points of the compass when they are separated by an interval of two lines. (This may be in part due to disuse.)

The wrists and lower part of the forearms are always cold.

After sitting for an hour in a warm room of temperature of 58° Fahr., that of the mouth was 98°, of the right fingers 66°, and of the left 69°.

The sclerema decreases upwards, and as a distinct affection ceases just below the wrists. The integument of the forearm, however, is brawny up to the elbows, the muscles are atrophied, and the anterior aspect of the limb is flattened.

The elbow-joint is slightly affected, so that complete extension of the forearm is impossible.

The right hip-joint is stiff and slightly painful, causing her to walk lamely at times. The feet are normal, but just where the ankles escape pressure from the edges of the elastic sides of the boots the integument is firmer than normal. There is no trace of œdema, nor are any of the superficial veins enlarged.

The adipose tissue has entirely disappeared from the neck and upper half of the front of the chest. On the sides of the neck, from the level of the hyoid bone downwards to the lower border of the first rib, the skin presents the characteristic appearance described in my previous case.¹ It is due to the formation of *lineæ atrophicæ*, which take an oblique course downwards and inwards to the root of the neck, and turning over the clavicle and first rib mount upwards over the manubrium to meet those of the opposite side, thus forming a series of semicircles around the root of the neck and the upper limit of the chest in front. The skin itself is thin and pliable, and there is at present no subjacent indu-

¹ See 'Medico-Chir. Trans.,' vol. lx, p. 137.

ration. On twisting the head aside, the lineæ atrophicæ are brought out conspicuously, and apparently in relief, but really this is not so, for the finger cannot detect the least irregularity of surface. The appearance of prominency is due to strong contrast of colour, the lineæ atrophicæ alternating regularly with lines of skin which has not undergone the atrophic change, but which has received an increase of pigment. Thus there is a very regular alternation of lineæ atrophicæ and lineæ pigmentatæ, the former are of ivory whiteness and opacity, the latter are yellowish brown and of the exact tint of the hair of the head. On examination with a pocket lens both kinds of lineæ are seen to be pervaded by a coarse, meagre, network of fine hair-like vessels, the main branches of which run in or close to the borders of the lineæ pigmentatæ. The meshes of the network are oblong and take the direction of the lineæ, the finest branches pass into the lineæ atrophicæ, where they become scant and soon disappear by attenuation. These features are represented in Plate X.

The patient herself thinks the neck has become a little browner of late, and she has noticed that on throwing back the head the skin in front of the neck is very tightly drawn. On doing so the fascia covering the mylohyoid is thrown into three tight thin cords, a central and two lateral, and the pressure upon the larynx excites deglutition.

A few fine blood-vessels are seen by the aid of a lens on the summits of the shoulders, and here and there in the cheeks, but elsewhere (as far as I have examined) they are absent; and on scanning the healthy skin of the shoulders and chest the regular pigmentation of the orifices of the sebaceous follicles arrests attention, for the surface is crowded with regularly distributed yellowish-brown, circular dots, commonly a little raised and surrounded by a very fine, radiated, wrinkling of the white cuticle.

The integument of the face is rather firm, the expression set, and rather sad, features which are not abolished by a smile. I have never seen her features in the least degree contracted; a fact which I noted in my former case, and

which occurred *pari passu* with the advance of sclerema on the face.

The mammæ are well developed. The lungs and heart normal. Of the latter the action is easily excited, but in a state of rest the pulse is from 70 to 80, and under all circumstances the rhythm is perfect and the beats equal; the systole is weak, and the first sound is rather flapping, as in conditions of debility.

The urine is normal, but poor, the sp. gr. averaging about 1012. Last summer I once found excess of uric acid, and a copious deposit of oxalate of lime, sp. gr. 1016.5.

She is liable to heartburn, and of late she has vomited once or twice a week a little frothy mucus, sometimes sour and sometimes bitter.

The bowels act irregularly; they are usually constipated, but she is liable to diarrhœa. The tongue is clean. She cannot walk more than a mile and a half on account of aching in the hips and legs, pain in the left side, and a feeling of nausea.

When warm in bed, she occasionally suffers considerable aching pain in the hips, wrists, and palms. The condition of the hands renders her very helpless, and she is unable to turn in bed.

She is chilly, and wears a complete investment of flannel. A table is appended, showing the measurements of the body and limbs, and the weight:

Measurements—

Round the waist	.	.	.	.	21½ inches.
„ centre of upper arm	.	.	.	.	10½ „
„ „ forearm	.	.	.	.	7½ „
„ wrist above and below the end of the ulna	.	.	.	.	7 „
„ last proximal phalanges	.	.	.	.	2½ to 2¾ „
„ ankle	.	.	.	.	8 „
„ calf	.	.	.	.	11½ „

Weight in her ordinary clothes—

Two years ago she weighed nearly 9 stone. Last summer she was a little over this weight. At the present time she weighs 7 st. 10 lbs. 12 oz.

Such is the history of this patient. Standing alone it would appear to have no definite signification, but side by

side with the patient whose history I have recorded in the last volume (vol. lx) of the 'Medico-Chirurgical Transactions,' it is very interesting.

The exact resemblance of the two cases is remarkable. Six years ago William W— was in precisely the same condition as Louisa M—. Both came to me because they were unable to close the hands, and at the same period of the disease these members were in precisely the same condition. Both had acquired a delicacy of constitution, probably from acute disease—in the one case typhus, in the other variola and enteric fever—in early life. Both had a previous tendency to syncope, and both exhibited the same condition of debility, and the same functional derangement of the stomach. Both exhibit a remarkable placidity.

To conclude, they have one complexion in common, and under the influence of the same disease the same sad fixed expression.

Both patients are here to night, and in the demonstration of this singular condition I esteem this a fortunate circumstance. As these are the only instances which have come under my observation, I have hitherto regarded the affection as a rare one, but like many other presumably infrequent diseases it may prove to be more common than I have supposed it to be.

In my former communication I have fully described the central complications of which the sclerema is, as I believe, but the outward expression, I will therefore, in conclusion, direct attention to two points; 1st, and by way of appendix to my former paper, the present condition of the first of my patients; and 2nd, the formation of the *lineæ atrophicæ*.

1. Those who may remember W. W— a year ago will find him but little altered. He is weaker, more emaciated, and the functional disturbance of the heart and alimentary canal is more severe, prostrating attacks of sickness and diarrhœa with severe colicky pains keep him in bed for days together. Although the radial arteries are wide and healthy, the hands have apparently reached the stage when dry gangrene is imminent. Indeed, the tips of some of the fingers are in-

vaded by a more active form of molecular death than that to which they have been previously subjected. It is accompanied by much gnawing pain. In my description of the hands, I omitted to mention the attenuation of the tips of the third, fourth, and fifth fingers, and the strong incurvation of the sides of the nails, corresponding exactly to what is seen in the other case.

2. As to the morbid process in the formation of the lineæ atrophicæ. If the neck in the more advanced case of the disease be examined with a lens, it will be seen that the ivory-like lineæ atrophicæ are, with the exception of a straggling vessel here and there, entirely destitute of blood-vessels. In the more recent case, however, the whole of the affected area is pervaded by a network of fine arteries, while no such vessels are seen in the healthy skin. It would appear then at first sight, that in the production of the lineæ atrophicæ there is an excessive vascular supply. The reverse of this is really the case: the process is one of atrophy, pure and simple from the first; there is no antecedent congestion, no hypertrophy.¹ By shrinking of the lacunæ—canalicular system and blood-vessels of the connective tissue of the corium and subjacent areolar tissue, it passes into a close, firm, bloodless tendinous structure, which by preventing the flow of blood at frequent intervals causes for a time dilatation of the blood-vessels in the alternate lines or areolæ of healthy skin. These latter receive an increase of pigment just as occurs in the delicate skin of some patients after the application of a sinapism.

The condition depicted in the accompanying illustration is therefore indicative of the early stage of the process. The streaky vessels there represented are really on their way to extinction, and when this occurs the pearl-like lineæ atrophicæ will stand out in greater contrast to the lineæ pigmentatæ.

Appendix, July, 1878.—Both patients are still in attendance at the hospital; the younger, L. M—, is but little

¹ The part was never the seat of irritation, on the contrary, there was diminution both of tactile sensibility and of temperature throughout the process.

altered. She is a trifle more helpless, thinner, and the disease is more marked in the face, the integument having become firmer, and the expression more set. Stiffness and some pain on movement have affected the other hip. The inner ends of the clavicles once were a little tender, and they appear to be somewhat enlarged. Once also she complained of a little aching at the root of the thumb and in the middle finger. These pains appear to be due to stiffness of the joints, rather than to a distinct rheumatismal affection. On three or four occasions she has had an attack of retching or vomiting, but only once a slight return of syncope. I have observed that she is unable to protrude the tongue, the strongest efforts only placing the recurved tip in contact with the retracted lower lip. She is not aware of any previous inability to protrude the tongue, and its forward movement is probably restricted now by sclerema of the connective tissue near it.

William W— is now 51 years of age. He weighs without his clothes only 7 st. 11 lbs., the adipose tissue has everywhere disappeared, and the following measurements indicate the degree of emaciation:—round the middle of upper arm $7\frac{1}{4}$ inches, round the calf $10\frac{1}{8}$, round the right wrist $6\frac{1}{8}$; the left is $\frac{2}{8}$ in. larger on account of a puffy swelling over the end of the ulna. The intervals of retching and vomiting are shorter, and the attacks are more severe and protracted; some weeks they occur every day. Perspiration is still profuse. The skin over the upper part of the chest remains free and supple.

In August he had a more severe attack of diarrhœa and retching than usual, causing for some days alarming prostration, and the hands were quite black. He rallied, however, and in the course of nine days was able to leave his bed. A fortnight later he had another attack which, proved fatal.

DESCRIPTION OF PLATE X.

Simple atrophic sclerema (Dr. John Harley).

Process of cutaneous atrophy in a case of simple atrophic sclerema. Louisa M—, æt. 20. The brown streaks represent the natural skin with an increase of pigment: the intervals, ivory-like areolæ or lineæ atrophicæ. Both are pervaded by a coarse network of fine hair-like vessels, the main branches of which run in or near the borders of the lineæ pigmentatæ, and forming oblong meshes, spread their finest branches in the lineæ atrophicæ, where they become scant and attenuated, and disappear.

The part of skin represented is that which overlies the right clavicle about the centre and the neighbouring integument above and below.