A CASE
OF
SIMPLE ATROPHIC SCLEREMA,
ASSOCIATED WITH
DISORDER OF THE CIRCULATORY AND ALIMENTARY
FUNCTIONS.

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THE following history will form a fitting supplement to the preceding paper by Mr. Gaskoin, as, while the case presents many points of agreement, the apparent differences are probably only those which may be expected to result from a difference in the exciting cause, and to the circumstance that in Mr. Gaskoin's case the morbid action has hitherto chiefly affected the peripheral parts, while in my own case the central organs have been from the first more deeply implicated.¹

William W—, æt. 45, a stonemason, came to me in March, 1872, complaining that he was no longer able to work from inability to close the hands upon his tools.

He stated that the disease began with "a little cold and

¹ I am indebted to Mr. Gaskoin for opportunities of studying his most interesting and instructive case.

indigestion," in the preceding September, when he was taken with a sensation of "great weight" in the epigastrium after food. It was accompanied by "terrible retching," but nothing was ejected except a little watery fluid. Under medical treatment the symptoms subsided, and then the hands gradually assumed their present condition.

He has remained constantly under my care up to the present time, so that my observation of his condition has extended uninterruptedly over a period of more than five years.

Speaking generally, the hands have undergone no change, but his health has declined, and changes associated with those affecting the hands have been gradually developed during this period.

The patient, now in his fiftieth year, is of moderate build. He is fair and has blue eyes, scanty reddish whiskers, a bushy head of hair just turning grey, and a thin, delicate, and only slightly pubescent skin; the lobes of the ears are prolonged and adnate.

Although he has latterly become somewhat anæmic, he still retains traces of a naturally ruddy complexion, due to the permanent injection of fine networks of minute cutaneous vessels.

He is the father of five healthy children, aged twenty-three, twenty, seventeen, thirteen, and nine, respectively; he has lived a careful, temperate, and industrious life, and has suffered but one illness, a severe attack of well-ascertained typhus, ten years before the present condition came on. He was able to resume work about thirteen weeks after he succumbed to this fever. His recovery was complete and was retarded by weakness only.

From the age of twenty-five to thirty-seven, he was occasionally troubled with small sluggish boils appearing about the back of the neck, and the white scars of two or three of these boils are still present below the nape.

His normal weight is about eleven stone, with very slight variation. He considers himself to have possessed average muscular strength, but from boyhood he has had

a tendency to faintness with outbursts of perspiration when in a close atmosphere. He has also been liable to flushings, and has always perspired freely, and on the least over-exertion, profusely. He has smoked since the age of twenty, his usual daily allowance of tobacco being from a quarter to half an ounce. He never experienced uncomfortable effects from its use, and he still smokes about half of the above-mentioned quantity.

His remote ancestors were mostly long-lived; his father and mother died early, but all their children are alive and moderately strong.

The father died after eight months' illness, at the age of thirty-eight, and when William W— was fourteen years of age, "of enlargement of the heart followed by dropsy." His son does not know whether it was of rheumatic origin or not. The mother experienced an attack of bilious and nervous fever (probably enteric) shortly after her marriage, and was ever afterwards a delicate woman.

She married a second time, and the issue of this marriage, a son, is alive and well. She eventually died of "consumption" at the age of fifty-one.

To return to William W—. When he first came to me, and for a year or more afterwards, he presented nothing unusual in appearance, nor, perhaps, would a casual observer have noticed anything amiss with his hands. There was no enlargement or œdema of any part of them, but on near inspection, the fingers from the first joints to the tips were seen to be sometimes rosy, but more commonly dusky, and on some days livid; and on taking either hand into your own it felt from the wrist downwards as cold as that of a corpse. The fingers were rounded, smooth, and hard, like tendon; no pitting could be produced by pressure, and it was impossible to pinch up the least fold of skin, the mobility of which was reduced one half, and it appeared to be tightly drawn over a hard substratum, so that the hands were constantly kept open, and it was impossible to close them completely.

After prolonged efforts he could, indeed, bring the finger-

nails of the left hand in contact with the palm, an inch and a quarter below the upper border of the hand (the anterior fold of the wrist), but he could not bring any nail of the right hand in contact with the palm, and after the strongest effort the finger-ends impended at the distance of an inch over the upper limits of the palm.

This induration of the areolar tissue was marked around the wrists, and gradually ceased about four inches above the joints.

The wrists were kept straight, and were still capable of limited movement; coercive attempts to flex or extend the hands caused pain, and the slighter disturbances incurred by their necessary use produced an intermittent aching, which was much complained of. The rotatory movements of the forearms were reduced to one half, *i.e.* the plane of the palm in passing from extreme supination to extreme pronation, described a quadrant only.

A fine creaking or grating is often felt immediately under the skin, by a finger placed on the back of the wrist when it is in motion.

The hands perspired freely, and the fingers were usually damp—some days even wet—as well as cold.

The actual temperature of the fingers was usually from 3° to 5° only above that of the external air; and from 30° to 40° below that of the axilla. After sitting in a warm room before the fire, and when warm in bed, the temperature of the fingers has become nearly 10° higher than that of the external air.

These facts, and the comparative temperatures of various parts of the body, are given in the accompanying table.

Temperature (Fahr.) of the fingers and hands, as compared with that of the apartment and other parts of the body.

	Air.	Fingers	Palm.	Axilla.	Mouth.	Toes.	Sole.	Ankle	Remarks ; condition of fingers.
1872, Sept. 18	60°	60°	65°	98°	...	81°	82°	83°	
Oct. 2	60	65	66	97·5	98				
10	60	65	70	98		65	75	80	
16	59	60	63	93	...	...	...	...	Very dusky.
23	58	63	63	97·2					
30	57·2	63·5	65	...	99				
Nov. 13	52	55·5	59·25	...	100	...	...	...	Very livid.
1873, April 30	54	60·7							
1874, Sept. 22	67	66							
Oct. 6	61	64	...	...	90				
1876, Aug. 15	61·5	70·5 ¹	...	...	99				

The sense of touch was very slightly, if at all, diminished ; and the tips of the index fingers, which certainly had their full share of the disease, maintained their natural superiority. Thus, the limit of confusion for the whole hand occurred when the points of the compass were less than two lines apart ; whereas the tips of the index finger discriminated two points when they were separated by an interval of only a line and a half. The nails were perhaps a little more incurved at the edges than usual, but their nutrition and growth were normal.

Such, I repeat, was the state of the hands nearly five years ago, and as there were no other indications of disease at this time, I was greatly puzzled as to the nature of the local disorder. I was inclined to attribute it to disease of the arteries of the hands, leading to dry gangrene, and thought of the stories of ergotised grain. But the patient had always resided in London, and the radial arteries, in common with those of the rest of the body, were soft and full. Subsequently, I was satisfied that the impediment to the circulation was due to the induration of the integument, which necessarily interfered with the movements of the arteries. From the further course of the disease it

¹ After sitting two hours before a fire.

may be assumed that the adventitia of the arteries, at least, was involved in the sclerema, thus preventing the expansion of the vessels.

Up to the present time the hands have remained unchanged, excepting that a hard, linear scar has formed obliquely across the palm, firmly binding the skin to the subjacent fascia, and that gangrene has actually declared itself, although in the very slightest degree. A year ago the first indication of this change showed itself as a minute, depressed brown scale on the extreme tip of the right middle finger, attended by sharp throbbing and shooting pain in the part. A speck-like cicatricial shrivelling now marks the seat of this change. Three months later on, the tip on the right index finger was similarly affected, and healing was attended by the discharge of a little thin pus. In November last this action was renewed at the tip of the same finger, and was accompanied by a moderate degree of inflammation, resulting in the separation of the cuticle of the forepart of the finger. A little discharge still continues; the forepart of the nail is separated, and the finger tip is slightly truncated.

The face was observed to be gradually losing expression, the mobility of the skin, especially over the malar bones and forehead and of the scalp, decreased, and the subcutaneous tissue grew firmer. Simultaneously the affection spread over the neck and upper part of the chest. The subcutaneous tissue of the abdomen is unaffected, but the aponeurosis of the external oblique muscle at least is implicated, for the moderately protuberant walls of the abdomen are hard and unyielding, and this condition is a source of discomfort to the patient. The fibrous tissue around the knees appears to be similarly affected. The other parts of the body, even the back immediately below the vertebra prominens, are at present free from the disease.

The expression is that of melancholy impassiveness; the mouth is always closed, and the lips somewhat compressed. A permanent hard ridge is seen across the forehead. It is produced by pressure of the hat.

The pleasant emotions may still evoke a ghastly smile, but the opposite passions have no longer power to contract the brow.

The characteristic features are, however, best displayed on the neck. When the head is thrown back, complete extension is prevented by the superficial fascia in front of the neck, which forms a thin, hard, resisting plane beneath the skin, gathered from the sternum and clavicles upwards into slight vertical ridges, feeling like knotted cords. Here the skin is free, but lower down, as far as the second costal cartilage, it is drawn very tightly over the subjacent parts, and in passing from the root of the neck, over the sternal ends of the clavicles and the manubrium, it attains its maximum degree of immobility, and is not only highly polished but partially eburnated in appearance. The surface is still further diversified by pink spots or little patches of minute straggling vessels, rendered conspicuous by the poverty of the skin in which they lie. The sides and back of the neck are brawny. On a cursory examination it appears seamed or furrowed, coarsely about the nape and just above the clavicles, and finely below the ears. The skin, however, is perfectly smooth, and the alteration is due to a partial disappearance of blood-vessels from the naturally rosy surface, which is now broken up, in the regions above stated, into red and white areolæ, varying in size from half a line to three lines in width, and arranged alternately with considerable regularity. The pink areolæ exhibit to the naked eye ramifications of minute blood-vessels, such as are seen in this and other parts of the surface in persons freely exposed to the weather. The extremities of these vessels terminate abruptly in the margins of the colourless areolæ. These latter are of ivory-like whiteness and opacity; here and there a minute blood-vessel from a contiguous vascular areola may be seen streaking it with pink, but usually the colourless areolæ are absolutely bloodless. These characters are clearly and conspicuously displayed under a low magnifying glass.

Towards the shoulders and back the red and white

mottling gradually disappears, the white areolæ becoming larger, more diffused, and, losing their ivory-like appearance, gradually assume that of the healthy skin; correspondingly the pink areolæ becomes smaller and less vascular, until they are reduced to mere points, and ultimately disappear; and thus the morbid and natural tissues are imperceptibly blended.

The skin of the affected parts retains, as far as I can determine the fact, the normal amount of sensibility. A distance of five lines between the points of the compasses is the limit of confusion for the whole of the side of the neck, but when the points are placed on the *areolæ atrophicæ*, at the root of the neck, they are rarely discriminated as two when fully an inch apart.

The skin generally has a slightly freckled and somewhat dirty appearance, particularly about the sides of the chest, due to a little increase of pigment. The adipose tissue has entirely disappeared from the integument of the upper extremities, the neck, chest, and back, the muscles of the arms are much wasted, and the borders of the scapular muscles are apparent under the thin skin. The lower part of the body and the lower limbs are in fair condition, and if the muscles of expression, the platysma myoides, the lumbricales doubtless, and perhaps the external oblique of the abdomen, be excepted, the muscular system must be regarded as free from disease. The wasting of the muscles of the upper extremities is clearly due to inaction.

Thus much respecting the local disease. I pass now to the general condition of the patient. He is weak and anæmic, and the neck and shoulders stoop. The curvature of the neck is mainly, if not altogether, caused by the contraction of the superficial fascia in front of it producing a forward inclination of the head.

But the most serious troubles are those which arise from associated disturbance of the circulatory and digestive organs.

During the first two years of the disease the pulse was

normal, tranquil, regular, of good volume, and moderate force, and ranging between 70 and 80. Shortly afterwards it began to intermit once or twice in the minute. This defect increased rather rapidly, and attained its maximum during the fourth year, when the heart's action exhibited remarkable variations, which have continued ever since. Sometimes the pulse is only 40 to 42, the beats regular and equal; but on listening over the heart 80 to 84 regular, but unequal ventricular contractions, are clearly audible, a feeble one regularly alternating with the stronger, which alone are appreciable at the wrist. But the most remarkable feature is that occasionally the feebler beats are altogether eliminated, the ventricular contractions being reduced to the same number (40 to 42) as the radial pulsations, the action meanwhile being quite equal and regular.

This regular intermission of the pulse and reduction of the cardiac contractions gives place either to the normal state—a regular pulse of 72 to 80, synchronous with the ventricular contractions—or to the usual form of irregular action in which the cardiac contractions being of unequal force the radial pulsations have a corresponding character with frequent intermissions. This latter is the common state of the pulse at the present time.

Associated with the gastric symptoms, which I will now describe, the patient has severe attacks of cardiac pain, with breathlessness, palpitation, and faintness. In the patient's own words, "at one time the heart seems as if it would jump out of his chest, and at another that it is not going at all," and he "thinks that he shall die."

The cardiac irregularity above described, has from the first been attended with dyspeptic symptoms, varying in frequency, but often occurring twice or thrice a week.

For the last two and a half years he has almost always complained of a pain, variously described as "heavy, burning, and gnawing," across the sternum on a line with the nipples. But occasionally pain of a more

severe character attacks the cardiac, epigastric, and left hypochondriac regions. It is usually accompanied by flatulency, and ends in severe retching and vomiting of sour or bitter mucous fluid.

Apart from these attacks the patient considers his digestion good. They are not provoked by errors of diet, for his food is of the simplest kind, and his drink only milk and water and weak tea. He has a constant tendency to diarrhoea and the motions are rarely formed.

To sum up. The patient inherits a rather delicate constitution, and this was probably somewhat enfeebled by a severe attack of typhus fifteen years ago. He is now greatly debilitated. A walk of two miles is as much as he can accomplish at best, but oftentimes a much shorter journey proves too much for him; "he feels faint as if he had no breath at all," an inclination to action of the bowels comes on, and a little watery mucus is discharged with slight prolapsus of the bowel.

Frequently after sitting in the cool waiting-hall an hour or two, he presents himself in the prescribing room with beads of sweat on his brow. The pupils are moderately dilated; the frontal vein is large and lax; it lies in a hard deep channel, and becomes full and prominent on the slightest inclination of the head, and a dilating wave of blood is at once thrown back into the vessel by rendering the tissues between the malar bone and root of the nose tense by a slight depression of the cheek, thus plainly indicating the complete absence of tone in the coats of the vein. A narrow line of dusky congestion occupies the margins of the gums; the inner surfaces of the thighs are pervaded by a visible network of fine blood-vessels, and an irregular festoon of similar vessels radiating upwards is seen over the anterior margins of the ribs and epigastrium.

The larger veins everywhere appear healthy. It is necessary to state that the patient is free from both varix and hæmorrhoids.

Apart from his graver troubles, and as a matter of

common complaint, he speaks of "a heavy sensation across the forehead, and a sharp pain across the chest."

The lungs and kidneys are normal, and the urine is poor, the sp. gr. being usually under 1008.

Treatment.—I have used tonics and stimulants, general and local, including ample trials both of digitalis and belladonna without avail. He has taken iodide of potassium at intervals, and has spent some months in the convalescent institutions of Bognor and Walton-on-Thames without advantage. He has lost strength in spite of all that has been done for him, and I cannot but regard the issue as unfavorable.

Pathology.—I have described with some minuteness the condition of the skin of the neck, for here, by the help of a pocket-lens, we may read the morbid anatomy of the disease. We see areas, so regularly disposed as to resemble broken lineæ atrophicæ, from which the blood-vessels have more or less completely disappeared, and the soft, clear, pink skin is converted into a firm, opaque, white, and bloodless structure. While the blood-vessels have collapsed and disappeared the system of lacunæ and canaliculi pervading the healthy connective tissue has shrunk; the adipose tissue for the most part has disappeared, and the soft, moist, and open cushion has collapsed into a hard, close, comparatively dry, and ill-nourished tendinous structure.

I hoped to have secured the attendance here to-night of a spare woman, otherwise healthy, in whom the sub-cutaneous tissue of the inner surfaces of the fingers and of the palms has become everywhere indurated and compacted with the palmar fascia contracting and fixing the fingers and seaming the narrowed and hardened palms. But such cases are familiar to all of us, and the subject of this paper himself exhibits this condition as it is usually seen, the centre of the right palm being seamed, tucked in, and firmly fixed to the fascia beneath.

The nature of the disease in this patient is illustrated very simply and clearly by these common instances of

partial sclerema of the palm, and the close associations which I here recognise will be a great aid in the proper classification of such examples of sclerema as that under consideration, for it may at once be isolated and distinguished from the greater number of the forty or fifty cases which have been described under the term sclerema and its synonyms. Only three or four of these have furnished opportunities for post-mortem examination.

In Förster's¹ case, which Dr. Fagge regards as an example of Addison's keloid, the corium and connective tissue were confused, forming a homogeneous white tissue, from which the fat had vanished, and the sclerosed connective tissue was firmly bound to the muscles, fasciæ, and tendons, and the author regards the change as due to "a chronic process of proliferation in the connective tissue of the corium, and particularly of the subcutaneous tissue, unattended by fever or local inflammatory symptoms."¹ It is implied that there was an increase of connective tissue in this case. Kohler,¹ Auspitz,¹ Arning,² and Day,³ expressly state that such was the case in their patients, and they especially mention an increase in the elastic element.

I cannot introduce a comparison with Rasmussen's³ case, for it appears from the progress of the disease, as well as from its anatomical characters (free cell infiltration of the connective tissue), to have been a case of cancerous induration of the skin.

The question of hypertrophy of the skin and subcutaneous tissue, when these have coalesced, excluding due consideration of the adipose element, is one that must be answered very cautiously in any case, for it is certain that before we can know that any absolute increase has taken place we must previously know the

¹ Referred to in Day's paper, 'American Journal of Med. Sciences,' 1870, vol. lix, p. 357.

² For references, see Classification, p. 149-150.

³ 'Scleroderma, and its relation to Elephantiasis.' Translation by Dr. W. Moore, Edinburgh, 1867.

thickness and consistency of the pre-existing adipose layer. Of the "proliferation of the connective tissue," so freely spoken of by Förster, neither he nor any other observer gives us any proof. Kaposi, indeed, found in one of the cases under his observation a "stasis of lymph-cells in the perivascular spaces," and he assumes that "this stasis and stagnation of lymph in the interstices of the tissues occurs in the production of scleroderma, the connective tissue being formed in excess out of the accumulated superfluity of nutritive material."¹

That the integument may be thickened and hardened by lymph stasis is obvious enough, but that growth of the connective tissue takes place as a consequence is a pure assumption. Nor, indeed, can Kaposi's case be regarded as typical, for the patient had lupus erythematosus, œdema of the legs, repeated attacks of "urticaria" in the affected parts, recurring rheumatic pains, and thickening and hardening of the bones of the legs—symptoms which are commonly associated with a specific irritation of the lymphatics.

Watching, as I have done, the gradual development of the disease in my patient, I can affirm that there has been no "proliferation of connective tissue" in his case. At no time has there been the slightest enlargement of the affected skin; on the contrary, the parts affected,—excepting perhaps the fingers, which have continued throughout in a chronic state of venous congestion from mechanical impediment to the return of blood,—have shrunk.

The atrophy is, of course, due in part to the more or less complete absorption of fat, but an examination of the skin in front of the neck of this patient, in which the disease is still progressing, will, I think, be sufficient evidence of that for which I contend, namely, that the disease in this particular case is due not to an increase of the subcutaneous connective tissue, but to its atrophy *ab initio*.

¹ Hebra, 'On Diseases of the Skin,' Syd. Soc., vol. iii, p. 123.

But what, we may ask, is the cause of this atrophy of the nutrient canals? of this conversion of skin into tendon? "Doubtless, some affection of the vaso-motor nerves" is the reasonable answer.

That the sympathetic nervous system is generally involved in this case, appears to me certain. The functional derangements of the heart, stomach, and intestines, the lax condition of the coats of the blood-vessels, the easy provocation of sweat, and progressive inanition, are to me marks of a grave depression of the sympathetic—of a slowly advancing paralysis of this system of nerves, of which the subcutaneous atrophy is but the superficial indication.

The case narrated is remarkable for the slow progress of the sclerema, and for its unmistakable association from the first with derangement of the central organs. In the cases recorded, disorder of the stomach or heart, or of both, is mentioned in only four or five.

In Rilliet's¹ case, the disease was ushered in by a sudden and violent pain in the epigastrium, accompanied by very intense palpitation, but these symptoms, as well, indeed, as the sclerema, may properly be referred to inflammatory irritation of the serous surfaces (in which I include the areolæ of the connective tissue), for at the end of a week or ten days, when the induration was subsiding, slight ascitis, and effusion into the right pleura and pericardium were noticed.

In Barton's² case the sclerema was preceded by dyspeptic symptoms, failure of appetite, and sickness after meals, with occasional vomiting.

But the cases which in several particulars most closely resemble that under present consideration, are those related by Dr. Arnold of Baltimore,³ and Dr. Day of New York.⁴ In both of these, derangement of the digestive and circu-

¹ 'Med.-Chir. Rev.,' 1848, p. 79.

² 'Dublin Quart. Journ. Med. Sci.,' Aug., 1869.

³ 'American Journ. Med. Science,' July, 1869, vol. lviii, p. 89.

⁴ *Ib.*, April, 1870, vol. lix, p. 350.

lating organs, were prominent symptoms. The troubles in the latter case were proved, however, both by the post-mortem examination, and by the physical signs during the progress of the disease (pain and fine crepitant râles in the upper part of the lung, and thence to the præcordia, &c.), to be due to chronic pleuritis, pericarditis, and peritonitis.

In the case before us, the disease has entered its sixth year, but there are no positive indications of organic disease in any of the viscera; the chest movements are free, the percussion note is good, and the breath-sounds for one in his debilitated condition, may be considered normal. Expiration is, indeed, slightly audible in all parts of the lungs, and the gentle vesicular murmur of health is somewhat encroached upon by a proportionate increase in the bronchial sounds—changes which, in an ordinary case, one should unhesitatingly refer to diminished elasticity of the lung, but which in this may be regarded as an indication of fibrous degeneration of the lung tissue.

At present the heart cannot be charged with organic disease, and the gastric symptoms are referable rather to defective or vitiated secretions than to peristaltic action hindered by adhesions.

Dr. Arnold's is really the only case which resembles that under consideration, for the two agree both in the duration of the disease and the attendant troubles of the alimentary canal. That these were merely functional in Dr. Arnold's case, appears from his statement, that "the cardiac and thoracic functions were normal, but the pulse was weak and slow."

So far then this case is almost unique, and as the disease is still in its mid-career, further opportunities will be afforded of watching its progress, and of testing the accuracy of the conclusion which I have drawn, respecting its nature and associations.

The slow progress of the disease suggests one other consideration. In most, if not all, of the recorded cases, the peripheral disease—the scleroderma—has been the prominent affection, and few fatal issues are recorded; in

the case under present consideration, the sclerema has become quite secondary to the grave disturbances of the circulatory and nutritive functions. How far the central disturbances interfere with the peripheral affection, or whether they do so at all, is a question worthy of consideration. I find it noted in Dr. Day's case, that an attack of vomiting and purging, lasting some days, was followed by "a great amelioration in the condition of the skin, which lost much of its bronze colour, became softer and more supple, and in parts could be moved over the subjacent tissues."¹ If we may recognise cause and effect here, then we have an explanation why the sclerema in my patient has during these five years made so slow a progress, for vomiting and occasional diarrhœa have never been absent from the first, and during the last three years they have been frequent and severe.

In conclusion, I venture to make a few remarks on the classification of the variety of disorders of the skin and subcutaneous tissue, which have been or may appropriately be included under the term sclerema and its synonyms.

Dr. Hilton Fagge² has done good service by bringing together "Keloid, Scleriosis, Morphœa, and some allied affections," and pointing out the relations between them; but the case which I have narrated to the Society, and that very interesting one recently brought under its notice by Mr. Gaskoin, throws additional light on the subject, and call for some further generalizations.

The terms *sclerema* and *scleriosis* are well suited for embracing these affections; *scleroderma* as a generic term is insufficient, since it implies an affection of the skin alone, whereas the morbid process in every case, involves the subcutaneous connective tissue, and in most, if not all, has its origin in this structure, the derm being usually involved in the later stages of the disease only.

Dr. Kaposi³ defines sclerema (scleroderma) as "diffuse

¹ Loc. cit.; see also the Appendix to this communication, p. 151.

² 'Guy's Hospital Reports,' 3rd ser., vol. xiii, p. 255.

³ Hebra, 'On Diseases of the Skin,' New Syd. Soc., vol. iii.

hypertrophy of the connective tissue," and of course places it in a class quite distinct from "xeroderma¹ or parchment skin," since he defines this as "diffused idiopathic *atrophy* of the skin." He says, "the course and development of sclerema and xeroderma, seem to distinguish them in a marked degree from one another. In the former, the disease commences as a board-like infiltration of the subcutaneous connective tissue, the corium being involved later and in certain parts only, the epidermis not at all, or only in the later periods of the disease. In xeroderma, the shrinking, the atrophy, appear to begin in the papillary layer and the epidermis, and from thence only to spread to the corium. This is evidenced by wrinkling of the epidermis, disturbance of the pigment, and the quantity of small superficial teleangeiectases." (p. 256).

He disregards the fact that atrophy of the papillary layer is attended by a proportionate loss of sensation. Thus, in the first case, a girl of eighteen, the xeroderma had existed "from early childhood," but "the sensibility of the skin was not diminished." In the other cases, the disease had existed in one, a girl of ten years, "from earliest childhood," and in the two others, "from the age of twelve months." The tactile condition of the skin is mentioned in the first case only, and it is obvious that if the papillary layer of the derm had been the seat of diffuse atrophy for a period of ten years only, the sensibility of the skin would not have remained intact as it is stated to have been. Hence it may be concluded that the special seat of the disease in so-called xeroderma is not the papillary layer. Then as to the condition of the epidermis, "disturbance of the pigment," "checkered pigmentation," certainly does not exist less frequently in sclerema than it does in so-called xeroderma. In Mr. Gaskoin's case, where xeroderma (ξηρός, parched) and sclerema are most

¹ Those authors to whom these high-sounding but trivial words are apparently of so much importance, should be careful to give them their proper meaning. "Xeroderma" no more denotes "parchment" skin than does the term "cutis anserina" or "pachyderma."

completely associated, there is not only confusion, but frequent interchange of all their supposed distinctive characters. As to wrinkling and shrivelling of the epidermis, this is merely the effect of the atrophy of the subjacent corium, and may be taken generally as a mark of the age of the disease. In only one or two of the recorded cases of sclerema had the disease existed so long as five years, and in none of Kaposi's cases of xeroderma had it existed for less than this term, and he had no opportunity of seeing either of them, until the disease had attained this and a greater age; he has, therefore, no ground for assuming that the morbid changes commenced in the papillary layer. Indeed, if supposition is to take the place of fact, it were more reasonable to assume that these cases of xeroderma originated in "Sclerema neonatorum," a condition indistinguishable from the "Sclerema (Scleroderma) adultorum."¹

There is only one more of Dr. Kaposi's distinctions that requires notice. It is the presence of small superficial teleangiectases. These are not infrequent in sclerema; they are characteristic of one form at least of keloid, and they form a very prominent feature in the case which I described.

If the preceding considerations have the force I consider them to possess, it follows that the lines which have been set to differentiate sclerema from xeroderma, and Scleroderma adultorum from S. neonatorum, can no longer be maintained, even as an aid to classification. The only question that remains is, the relationship that elephantiasis, leprosy, and tubercular diseases of the skin generally, bears to some cases of sclerema—such as that of Mr. Gaskoin's notably.

Since the essential feature of these affections is hypertrophy and induration of the subcutaneous connective tissue and skin, I think they must be included with sclerema in a general classification. With this view, I would define

¹ A comparison of the records of the cases included in the oedematous variety of sclerema (see p. 149 of this paper) with "Sclerema neonatorum" will prove the truth of this statement.

sclerema as an induration of the subcutaneous connective tissue, sooner or later in some degree involving the derm and epiderm, and causing proportionate alteration of structure and disturbance of function. Two or, perhaps, three classes may then be formed:—1. SIMPLE SCLEREMA, or induration without actual hypertrophy of the connective tissue or skin; 2. IDIOPATHIC HYPERTROPHIC SCLEREMA, in which the induration of the connective tissue is accompanied by positive hypertrophy; and 3. SPECIFIC HYPERTROPHIC SCLEREMA, in which may be included the tubercular disease of the skin.

1. SIMPLE SCLEREMA; under this may be grouped the following varieties:

(A) SIMPLE ATROPHIC SCLEREMA, marked by the gradual disappearance of blood-vessels, and shrinking of the connective tissue, so as to attach the corium, more or less completely, to the subjacent fasciæ, muscular aponeuroses, tendons, ligaments, or bones, and unattended by any previous changes in the parts affected.

This may be—(1) *General*, as in the case narrated, and in those given by Bruche,¹ Barton,² Day.³

(2) *Local*, as in the sclerema of the fingers and palm; in one of Mosler's⁴ cases (W. J.); in Case 3, related by Arnold,⁵ and further exemplified by Alderson's⁶ case; and in those cases of so-called Addison's keloid,⁷ which are simply atrophic.

(3) *Scattered*, which will include morphœa, and many cases of partial sclerema.

(B) ŒDEMATOUS SCLEREMA, such as occurs in children (Scleroderma neonatorum), in chlorotic girls, and those

¹ 'Hannov. Ann.,' vii, 5 and 6, 1847.

² 'Dub. Quart. Journ. Med. Sci.,' Aug., 1869.

³ 'American Journ. Med. Sci.,' April, 1870, vol. lix, p. 350.

⁴ Virchow's 'Archiv,' B. 23.

⁵ 'American Journ. Med. Sci.,' new ser., vol. lviii, p. 89.

⁶ 'Medico-Chirurgical Transactions,' vol. xxxvii.

⁷ Ibid.

in whom the catamenia are suddenly suppressed (see Thirial's¹ cases, Arnold's case,² one of Mosler's³ cases, two of Rilliet's⁴ cases, Fantonetti's⁵ case, and Sir W. Gull's two cases quoted by Fagge).⁶

(c) INFLAMMATORY SCLEREMA, including those cases in which the local affection has been preceded by some amount of inflammatory irritation. It may be—(a) *Acute*, as in the cases related by Henke,⁷ Bouchut,⁸ Gillette⁹ (which followed the application of a blister), Eckström (which followed an attack of erysipelas),¹⁰ Arning,¹¹ and by Fuchs.¹²

(b) *Chronic*, as in the cases given by Fiedler,¹³ Forget,¹⁴ M'Donnell,¹⁵ Nordt,¹⁶ in the very interesting and typical one by Mr. Gaskoin,¹⁷ and those by Sedgwick¹⁸ (partial) Mosler (in one), perhaps that by Grisolle,¹⁹ and that by Förster.²⁰

(d) TRAUMATIC, following upon injuries, such as variculous and syphilitic inflammations, burns, the use of the cat, and ulceration.

¹ 'Gaz. Méd. de Paris,' 1845, p. 523.

² 'Amer. Journ. Med. Sci.,' new ser., vol. lviii, p. 89.

³ Virchow's 'Archiv,' B. 23.

⁴ 'Traité des Malad. des Enfants,' 1861, vol. ii, p. 107.

⁵ 'Annali Universi di Milano,' 1847.

⁶ 'Guy's Hosp. Rep.,' 3rd ser., vol. xiii, p. 286.

⁷ 'Handb. zur Erkenntniss und Heilung der Kinderkrankheiten,' 1809.

⁸ 'Gaz. Méd. de Paris,' 1847, p. 771.

⁹ 'Archives Gén. de Méd.,' 1854, tom. ii, p. 657.

¹⁰ 'Hygiea,' Band ii, No. 2.

¹¹ 'Hygiea,' Band ii, No. 2, and 'Würburger Med. Zeitschrift,' 1861, Bd. ii p. 186.

¹² 'Bericht. über die Med. Klin. zu Gottingen,' 1855, p. 192.

¹³ 'Deutsche Klinik,' 1855, p. 34.

¹⁴ 'Gaz. de Strasbourg,' No. 6, 1847.

¹⁵ 'Dub. Hosp. Gaz.,' 1855, vol. ii, p. 6.

¹⁶ Virch. 'Arch.,' 1861, vol. xxii, p. 198.

¹⁷ Page 113 of this volume.

¹⁸ 'Pathological Trans.,' vol. xii, 1861, p. 234, and vol. xvi, 1865, p. 260.

¹⁹ 'Gaz. des Hôp.,' 1847, p. 209.

²⁰ 'Würzburg Med. Zeitschrift,' 1861, Band ii, p. 294.

2. IDIOPATHIC HYPERTROPHIC SCLEREMA.—

That an increase of the connective tissue and of the skin can occur during the progress of simple atrophy of the integument is obviously impossible. Nor can I consider that actual hypertrophy has been proven in any of the recorded cases, for the reasons stated at pp. 142, 143, but that such a condition may arise in consequence of prolonged subacute inflammatory action so insidious as to exclude it from B and C, Class 1, is possible, and such cases would be properly included in this provisional class.

3. SPECIFIC HYPERTROPHIC SCLEREMA, including the following :

- (A) SYPHILITIC HYPERTROPHIES AND CONDYLOMATA.
- (B) ELEPHANTIASIS.
- (C) LEPRA ARABUM.
- (D) MOLLUSCUM.
- (E) KELOID (the tubercular as distinguished from the cicatricial form).

Such a classification as the above has the advantage of bringing into a single view a number of morbid conditions more or less closely allied, and each of which will, I believe, throw some light on the etiology and pathology of the others.

All classification is, of course, to some extent artificial ; and just as occurs in grouping animals or plants, so here we shall surely meet with cases which will serve as links between the classes above defined.

APPENDIX.—During the present summer (1877) there has been a slight increase of weakness, and the patient has occasionally been obliged to keep his bed two or three days in succession, and the hands have become duskier.

After a sojourn at Walton-on-Thames, the patient presented himself at the hospital on the 3rd of August, when the external shade temperature was about 80° Fahr.

He stated that he was no better, and showed extreme debility. He was perspiring freely; the hands were darker than I had ever seen them, the ends of the fingers being bluish-black and cold. Dry gangrene, indeed, seemed imminent, and the integument over the summit of each metacarpo-phalangeal knuckle of the right hand was occupied by a minute dark adherent scab, like those which formed on the tips of the two fingers, which have undergone slight gangrenous shortening.

The sclerema, however, is less for the first time during the progress of the disease. The integument can now be slightly pinched up over the backs of the proximal phalanges of the right hand, the integument of the neck has lost its brawny consistence, and the skin over the upper part of the chest has resumed its normal appearance and mobility. There is no amelioration of the cardiac and gastric symptoms; the poor patient is a wretched object, the neck being very thin, and the features drawn and expressionless, like those of a mummy.