Thesis for M. D. degree.

# Sclerodema Adultorum

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# Scleroderma Adultorum.

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# Sclerodermia Adultorum: — by D. McCalman, M.D.

This is one of those affections which rarely come under the observation of the general practitioner and even in the skin-disease wards of our hospitals it is seldom seen. Even when a case is admitted into an hospital, owing to the chronicity of the disease and the slight inconvenience it causes to the patient, it seldom remains sufficiently long to allow of the protracted observation necessary for watching the slow changes which take place.

Though now well recognised, this rare disease was, according to Auring (in "Wurzburg med. Zeitschrift", 1861 Band II, p. 186) first described by Curzio in 1752 under the title "Dissertation Anatomique et pratique sur une maladie de la peau d'une espèce fort rare et fort singulière"; but the name Sclerema Adultorum ("Du Sclérème chez les adultes") was introduced by Thirial in 1845 in an account which he published in the "Gazette Méd. de Paris for that year of two cases of this disease that were under the care of Trousseau in the Hôtel Dieu. After the publication of Thirial's two cases communications

from various authors, among whom we may mention Forget, Bouchut, Pulegnat, Brück, Pelletier, Eckström, Gillette, Fuchs and W. Donnell, appeared in different Journals, Chiefly French and German, till Anning in 1861 was able to collect seventeen cases previously recorded, and publish a critical examination of them along with a case observed by himself in the II Vol. of the "Wurzb. Med. Zeitschrift" above referred to. Subsequent to the communication of Anning contributions came from Bazin, Förster, Nordt, Moosler, Köhler, Köbner, Bärmann, Brück, Läger, Pierquin, Fiedler, Mosler, Bing, Wernicke, Villemin, Gamberini, Auspitz, Plu, Rasmussen, Paulicki, Fagge, Arnold, Barton, Neumann, Rosbach, von Hebra, Erasmus Wilson &c., Hebra in 1868 met with only five cases in his extensive practice as a dermatologist, four in the Allgemeines Krankenhaus and one in private. Of the two last of his public cases he published a very interesting account in the annual reports of the hospital for the years 1867, 1868 (Abtgl. Ber. des k. k. Allgemeines Krankenhaus) which his son-in-law, Prof M. Kaposi, published subsequently in Hebra's

work on Skin diseases. In 1867 the number of cases reported amounted to thirty-one; twenty-four of which occurred in females and seven in males (see Journal of cut. med. and diseases of the skin" 1867 Vol I. p. 218.)

In recent years the number of cases of Sclerodema recorded has increased in much the same proportion as formerly, and, as far as I am aware, without diminishing the proportion which appeared in women - indeed it has rather increased it. Among the contributors may be mentioned Kaposi, Hallopeau, Moxe, Colley, Bernhardt, Schwalbach, Westphal, Hader, Chiari, Dorozynski, Haller, Caspari, W. Wall Anderson &c.

As an example of the rarity of the disease we may mention that W. Wall Anderson had seen only three cases in eleven thousand cases of skin disease (Brit. Med. Journal 1877, Vol II page 812) which came before him. The greater number of the cases known have been recorded in German and Austrian Journals, though a number appeare in French, British and American Journals.

Considering the comparative rarity of the disease and the paucity of our knowledge of it, the following case which has been under my care for a considerable time may not be altogether unworthy of being recorded and studied, as it is a good example of the disease generally and presents some peculiarities which I fail to find in any of the records of cases which I have read.

The patient is a girl, Christina M.D., twelve years of age and enjoyed good health previous to the present illness. The exact date of commencement is not quite certain, but about two years ago her mother noticed that she was less inclined to activity than formerly - that she was listless, sleepy, and did not take her food well - but complained of no pain. After a month or six weeks of this state her mother, while undressing her for bed, noticed a a large bluish mark over the sacrum and on examining her more particularly found that the upper and lower left limbs "felt very hard and appeared withered". With the exception of the above mentioned drowsiness and loss of appetite no history of previous complaint could be elicited either

from the patient or from the mother. After a time, however, the mother observed that the disease was increasing slowly in extent and intensity at the distal parts. Sharp, shooting pains now made its appearance, slight during the day but pretty severe at night. Pressure with the finger and particularly rubbing, caused considerable suffering. The girl continued in this condition, attending school regularly and without apparent inconvenience except that the affected limbs were more sensible to cold and felt somewhat stiff; neither getting better nor worse, unless that she became generally emaciated and that her appetite became consiederably impaired, till her mother, getting at last anxious, called me to see her in the beginning of December 1880.

On examining the upper left limb I found that all the tissues corresponding roughly to the area occupied by the extensor muscles, beginning at the base of the scapula, involving the ~~lower~~ two-thirds of the deltoid and extending to the wrist, were firmly bound down to the bone, leathery to the feel; and the skin would not "slide" nor could it be pinched up

into a fold. From the middle of the forearm, where the lines formed by the outer and inner borders of the affected area met in front, to the wrist the whole circumference of the arm was bound down, but the hand and wrist-joint were perfectly free, i. e. normal. With the exception of a small area on the back of the forearm and immediately above the wrist-joint, and a narrow band passing obliquely upwards and outwards to the elbow-joint, where the surface had a mottled brown appearance, the surface of the affected area had a markedly bluish appearance, probably due to capillary stasis, and the arm seemed atrophied. There were also some narrow bands more bound-down than other parts, particularly one along the course of the musculo-spiral nerve — the outer anterior border of the disease — and two transverse bands bands, one immediately above the elbow, and one about the middle of the humerus. These bands, as well as others less marked in the forearm, gave the surface an irregular appearance. In the skin of the affected parts numerous, minute, starlike or circular white patches were seen, and these were very numerous in the

parts most bound down; indeed, there seemed to be an obvious proportion between the number of these white patches and the firmness with which the particular spot was bound down. In the groove formed by the extreme binding-down in the course of the musculo-spiral nerve and also over a very firm band in the forearm which passed from the outer supra-condyloid eminence obliquely downwards and backwards for four inches, these white patches almost coalesced. They are all very equal in size — about two lines — and in the band above mentioned in the forearm, they are surrounded by brownish pigment giving this part a peculiar appearance as compared with the rest of the affected area. There has been no broken skin from the beginning except a small boil or pimple the anterior and upper aspect of the deltoid; but the skin here is, and was all along, perfectly normal. Tactile sensibility as tested by pin-points, hot and cold applications &c., does not appear to be affected in the least. With the exception of an occasional sharp, shooting pain, there is no suffering during the day, but

there is still, though not so severe, considerable pain during the night. Pressure with the finger still causes pain and rubbing gives rise to pretty severe suffering. Flexion and extension of the elbow-joint is considerably interfered with and when forcible extension is attempted the vertical bands become very tense and white.

The affection of the lower limbs is similar to that of the upper, only that the tissues here are thicker and consequently don't feel so much bound-down. It begins at a point on a level with the base of the sacrum, and three inches to the right of the middle line, in a band, two and a half inches broad, passing obliquely downwards and outwards over the sacrum and involves the whole of the left buttock, thence to the front of the limb and down to the ankle-joint — just the area of the muscles of extension. Like the arm, the lower part of the leg is surrounded. The hamstrings, adductors and gastrocnemeus are not involved, but immediately below the muscular part of the gastrocnemeus, and forming the upper border of the part which surrounds the leg, is a

transverse band very firmly bound-down and presenting them cicatrix-like patches very numerously. The foot and ankle-joint are perfectly free as are also the body and head and neck. The colouration is bluish. The edges of the affected areas are generally well marked and give, in contrast to the adjacent healthy tissues, the idea of loss of substance - as marked is the depression. This depression is extremely well marked in the upper arm and over the caecum and parts to the right of it.

Careful percussion and auscultation of the lungs and heart, elicit no abnormality. The urine was examined from time to time but nothing abnormal could be detected. Temperature has also been normal. Tongue clean; bowells regular.

Although patient's immediate relatives are to all appearance quite healthy and free from nervous taint, there are relations of hers in the neighbourhood - four families of cousins - some of whom have very marked nervous affections - one an idiot, three lunatics and almost all the females markedly hysterical. Patient is the eldest of her own family -

The treatment employed consisted of good nourishing food, attention to the functions of the body, 2 M. Liq. Arsenicalis, 2 M. Liq. Strychniae, 5 M. Tinct. Ferri Perchlor. thrice daily; and having discovered accidentally that the affected parts were much softer when patient was for some time in a warm room, I got her well clothed in flannels and a liniment consisting of three parts of Lin. chloroformis and one part of Lin. Aconiti was rubbed well over affected parts night and morning.

During the last seven months the conditions above described have changed considerably. Both limbs present a marked improvement; but the improvement is very much greater in the lower than in the upper limb. If I accept the mother's statement as correct, and I have no reason to doubt them, the disease advanced distally both in area and intensity, and my own observations show me that recovery began at the proximal ends and is steadily, though slowly, progressing towards the distal ends; i.e. towards the ankle and wrist. In the upper arm no trace of the induration appears above the point of origin of the outer head of the

triceps, and even as far as the elbow-joint The improvement is very marked. Below the elbow-joint the improvement is less but still obvious, and the brown mottling is more extensive. Over the shoulder-blade and lower half of the deltoid the only trace of the disease consists of a marked prominence of the superficial vessels and the persistence of the minute, white spots already refered to. These white spots are pretty numerous here and are now depressed considerably under the level of the surrounding skin which is soft and natural; thus giving the part a pitted appearance not unlike that following a slight attack of smallpox.

In the lower limb the only trace of the induration above the knee-joint consists of a vertical band about two and a half inches broad on the outer-anterior aspect, and extending from the patella to the middle of the thigh. Below the knee-joint the tissues are still pretty firmly bound-down but the improvement is obviously marked. The transverse band below the muscular part of the gastrocnemeus has almost disappeared. In the

upper and recovered part of the thigh, and especially over the sacrum where the depression was so marked, but which is now scarcely if at all perceptible, the surface is glistening, has a darkish-blue appearance, prominent superficial vessels and is marked by those, now pitted, minute white spots. Where there were longitudinal or transverse bands there are now soft, white cicatrices not unlike those seen on the abdomen of women who have born children. The buttock has no longer the pinched, atrophied appearance it had, but is soft and scarcely less in size than the right. There is now no pain at night nor does pressure produce any pain but rubbing still produces slight suffering. Patients' general health is very much improved.

The etiology of Scleroderma Adultorum is very obscure. Certain conditions and morbid processes, such as severe colds, severe wettings, articular rheumatism, erysipelatous inflammation &c., have been described by various authors as having immediately, or within a comparatively short

period, preceded the development of the disease; but in many cases no obvious symptom or state which could be justly connected with the disease was observed or at least recorded. No doubt such morbid conditions as above mentioned precede the majority of cases, and some observers, and I think with good reason, consider them, especially rheumatic fever, as undoubtedly connected with the subsequent Sclerodema.

The pathological conditions found in the affected parts are great increase of the connective and elastic tissues of both the corium and subcutaneous cellular tissue, accumulation of lymph-cells forming sheaths round the vessels, and deposits of brownish pigment in the corium. Dr. Chiari of Vienna published in the "Vierteljahres-schrift für Dermatologie und Syphilis", 1878, Heft II, page 185, an account of a microscopic examination of the spinal coard and ganglia, as well as parts of the affected skin, of a case of this disease which was under the care of Dr. Mader in the Rudolfspital, but found nothing abnormal in the coard or ganglia. The abnormalities found in the skin agree with

other observers, except that he does not mention the presence or absence of lymph-cells under he means their absence to be inferred from his statement "that no obstruction was met with in the vessels of the lymphatic system" (Kein Anhaltspunkt im Lymphgefässsystem sich vorfand). He found the brown pigment in the papillary layer occupying the situation described by others as occupied by the lymph cells, i.e. the neighbourhood of the blood-vessels. He, however, calls particular attention to numerous partial indurations of the coverings of the brain which were found by Westphal in a case of this disease of which he published an account in the "Charité-annals," III Band.

It will thus be seen that neither the pathological anatomy as far as it is yet known nor the morbid processes or conditions preceding the development of the disease, help us much in the elucidation of the cause of Scleroderma. But if we take them in connection with the clinical symptoms, mode of extension and treatment found most advisable, they will, I think, help us considerably in forming an opinion of the nature of the disease. The frequency with which the affection

is preceded by rheumatism, especially articular rheumatism, or severe colds, which are somewhat related, and the almost constant symmetry with which the disease spreads, point, I think, to an undoubted neurotic origin. This conclusion I arrived at from the wonderful analogy between the area affected in connection with the upper and lower limbs of the case observed by myself; and I was further confirmed in this opinion when I considered the course of the cutaneous, as well as the muscular, nerve supply, especially in the upper limb, of the area affected. In the upper limb the nerves which supply the affected area, except the integument over the back of the scapula which derives its nerve supply from the posterior division of the dorsal nerves, are the suprascapular, circumflex, external cutaneous and musculo-spiral nerves. The fasciculi of which these nerves are composed may be traced to those of the 5th, 6th, 7th & 8th cervical nerves. The nervous supply of the affected area in the lower limb is more complicated. I have been also considerably impressed with the prevalence of nervous disease

in the family, and I direct attention to it de= spite the consummate modesty of Dr. A. B. Shepherd in denominating a similar statement by Hallopeau in connection with a case of Scleroderma which he published in the "Gazette Méd. de Paris", 1873. page 584, as "irrelevant!" (see Sydenham Society's Bi= ennial Retrospect of Med. & Surgery for 1873-1874. p. 194)

I am now aware that Scleroderma has been attributed to a nervous cause by previous observers, but my conclusions, whatever they may be worth, were arrived at independently; i. e. before I became aware of the fact that others had already attributed it to a nervous cause. In a letter I wrote to my friend professor Gairdner of Glasgow on the 16. Feby. 1887. regarding the case under my care, I expressed my opinion that the disease was due to a neurotic cause pretty strongly. Dr. Mader of Vienna was, as far as I am aware, the first to en= unciate the idea of a nervous cause, and I am know from a private letter that Prof. Kaposi is of Dr. Mader's opinion. Dr. Mader, after criticizing Prof. Kaposi's conclusions as published in "Virchows Extracts" (they are the same as those

given in the Sydenham Society's translation of Hebra's work on Skin diseases) writes:- "Es scheint mir viel plausibler anzunehmen, dass der Sclerodermia universalis, wie so manchen anderen Hautkrankheiten, eine Affection der trophischen Nerven zu Grunde liegt, deren Sitz in den medullaren Centres gesucht werden muss." (cu "Vierteljahresschrift für Dermatologie und Syphilis". 1878. Z II. p. 186). It is somewhat unfortunate that W. Mader is so precise. However plausible such a theory may be, we cannot for= get that the very existence of the trophic nerves is purely hypothetical. Prof. Herrmann of Zürich- au his Human Physiology - though not absolutely denying, seem to doubt their existence, and holds that they are by no means essential to the phe= nomena cited to prove their existence:- that tho phenomena may be refered to the action of the motor and especially the vaso-motor nerves. To put my own opinion of the etiology of Sclerodema Adultorum in a few words:- I believe it to be an abnormal state of the nutritive processes due to a local affection of

the nerves, the site of the affection being most likely in the cerebro-spinal axes — just Dr. Mader's opinion minus his precision.

— I am not sure that any significance can be attributed to the small, circular or star-like white spots already referred to. I mention their existence simply as a prominent feature which I fail to observe recorded by others unless Kaposi refers to them when he says that the diseased parts were "marked with dark-yellow brown pigment spots and streaks and dots, between which were intervals of white colour without pigment", or Chiari who writes, in the article already referred to, when describing the appearance of the skin-" Dieselbe war glänzend, blass, mit einem bräunlichen Teint versehen, im Gegensatze zu welch' letzterem an einzelnen Stellen, wie an den Zeigefingern und Daumen unregelmässig begrenzte Flecken durch ihre weissliche Farbe sich scharf abhoben". That they are an entity in the pathology is shewn by the permanent marks, in the shape of pitts, which they leave behind them.

Prognosis in this disease ought to be very guarded, especially when it is extensive and involves the trunk. And though it may be confined to a very limited patch when first seen it sometimes spreads slowly but steadily till it affects very large areas. It appears that the disease per se is seldom or never fatal; yet, even when it affects only a small spot, it is often very obstinate. But when the skin is extensively involved, especially that of the trunk, other complications, such as phthisis, cardiac disease, kidney disease &c. are to be feared. Such complications are just what would be expected from an extensive interference with the important function which the skin performs in the economy, and also the depressed state of health, and frequent mental suffering, which accompany the disease. But though the prognosis should be guarded, it should be by no means hopeless. We have some examples of cases of complete recovery, and of many cases in which considerable improvement took place. The case under my own care is almost completely recovered, and shews that recovery may take place

even in parts which are tendenous and in parts depressed below the level of the adjacent normal skin. Formerly such conditions were held to be hopeless: on Sydenham Society's Translation of Hebra's work on Skin diseases, Vol III. page 116.

As regards treatment a good nourishing diet and attention to the bodily functions are essential. In medicine those remidies, such as iron, Strychnine, Arsenic, &c., which long experience has taught to be most efficacious in improving the tone of the nervous system, should be employed. To this must be added warmth which can be best secured by plenty of woolen cloth. Liniments may be used — those whose application cause heat in the part — but whether they are beneficial or not is doubtful. Professor Kaposi recommends ferruginous compounds and cod-liver oil internally, combined with the external use of otid of iron and copper baths, and ointments containing copper. Baths, no doubt, seem to have been employed with advantage — improving cases which ultimately terminated fatally from complications — and perhaps of all the forms, hot-air is preferable. This is easely obtained

in private practice by means of a spirit-lamp, cane-bottom chair and crinoline. Change from a cold to a warm climate might be useful. Electricity has been employed but with doubtful success. Scleroderma, however, seems sometimes to undergo involution spontaneously so that we must be careful and not attribute to certain remedies more efficacy than is their due.