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ON A RARE FORM OF

# IDIOPATHIC LOCALIZED OR PARTIAL ATROPHY OF THE SKIN

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# ON A RARE FORM OF IDOPATHIC LOCALIZED OR PARTIAL ATROPHY OF THE SKIN.*



Although our knowledge of the various forms of symptomatic atrophy of the skin is quite complete, there are many points in the origin and course of the idiopathic localized or partial variety which yet need careful study. Except the paper, by Mr. Wilson,† in the Journal of Cutaneous Medicine, and the chapter by Kaposi ‡ in Hebra's handbook, we have not any noticeable article on the latter condition, and it must be confessed that these are in a measure incomplete. My own observation and study lead me to agree with Kaposi, who recognizes two distinct forms of idiopathic partial atrophy of the skin, the one linear, the other in the shape of round or oval spots. The linear form, though not very rare, still is not common, and is most frequently met with in persons of a delicate skin, particularly in those who have grown fleshy rapidly. As this variety is well described, I shall say very little of it in this paper, except to mention certain features presented by a case, in which I observed the integument during the whole course of this atrophic process. Besides this case I have also seen a number of others, males and females, in which these white atrophic streaks were formed in certain localities without apparent cause. Both Wilson and Kaposi state that the form, in which round and oval spots occur, is quite rare, and they only allude to it in a superficial manner. For these reasons it occurred to me that the Society would be interested in the details

* Read before the New York Dermatological Society, Dec. 14, 1875.

† On Striæ and Maculæ Atrophicæ Cutis. Journal of Cutaneous Medicine, Vol. I., p. 140.

‡ Diseases of the Skin (Sydenham Translation), Vol. III., p. 260

of a case showing the lesion, which came under my observation, and which I studied very closely at the time. Though it does not throw much light upon the etiology of the affection, I think that it brings out its clinical history much more clearly than a didactic description could, and that it will materially increase our knowledge of this rare and obscure affection. Besides these points of interest, certain considerations as to the resemblance of the lesion with one form of morphœa are suggested, which, when studied carefully go to prove that the two affections are radically different. I am in hopes that by our study, certain heretofore obscure points as to the so called atrophic morphœa are cleared up.

I think that the term idiopathic localized atrophy is to be preferred as being more definite than is the term partial atrophy.

KATE C.——, aged 45, Irish, a seamstress, came under my observation in February, 1872. She was a thin and tolerably healthy woman, but she had suffered often and severely with rheumatism during her life, and of late years had not been as well as before. She had never had any skin affection until the appearance of the one for which she sought relief, nor had she prior to that time experienced any abnormal sensations as to the nervous distribution in the integument. There was no history of wound or injury of any nerve of the body. She had been married and had borne children who were in every respect healthy. She applied to me for treatment of an affection of the skin, which was situated mostly on the flexor and slightly on the outer border and exterior surfaces of both fore arms slightly on the flexor surface of the arms, and sparsely on the abdomen and on the anterior and outer surfaces of the thighs. The affection consisted of groups of round and oval patches of skin, of various sizes, from the diameter of two lines to that of half an inch as a limit, having a whitish glossy appearance resembling very much mother of pearl. Their surfaces were very smooth, but over some of them a few pores were very noticeable. These were evidently the sites of follicular ducts, but they were much larger and more striking in appearance than the latter when normal, being perceptibly trumpet shaped. The patches ended by a sharp, well defined margin, and the surrounding integument seemed normal. They were evidently very slightly depressed below the level of the skin, to the extent of the thickness perhaps of thin or tissue paper, as shown by looking at the limb obliquely. If examined with a magnifying glass it was seen that the natural surface lines of the skin were wanting and the appearance noted was in a measure that which is seen when certain very thin and delicate syphilitic cicatrices, which have become thoroughly blanched, are examined in the same way. There were no hairs, even downy ones. If pinched between finger and thumb the fact that marked atrophy existed was appreciated; but this fact was better made out by delicately pinching up a fold with dressing forceps and comparing its thickness between the blades with that of the normal integument similarly treated It was evident that the subcutaneous tissue was also

thinned. As the fingers were passed over a group a marked sensation was communicated both as to the surface and the condition, of the deep layers of the corium. These patches were soft, smooth, and I may even say slippery to the touch. A group of these patches, four inches wide and four long, reaching to and ending abruptly at the wrist, was seated on each forearm, while a few existed just above the elbow joint, on the flexor aspect. The chief part of the group was seated on the inner and flexor aspect, and the outlying patches of the group extended in a straggling manner on the outer and extensor surface where they were very few and small in size. The centre of the group was formed by the larger ones, the small and intermediate sizes being outside. Over the abdomen, there were in all about one dozen of these patches of larger size and they were sparsely scattered, among the atrophic lines due to pregnancy. On the thighs the distribution was in all particulars like that of the forearms, the groups being situated at the lower part of the upper, and the upper part of the middle thirds. Such were the patches which were strikingly noticeable of this affection, but there were other patches seen which also presented a peculiar appearance. These latter were irregularly scattered among the white ones ; being in numbers and size somewhat less than the latter. These patches were of a light brown color, resembling very much tinea versicolor, but being somewhat darker. Their surface was rather rougher to the touch than the normal skin, but they were not at all elevated. In size they corresponded very closely to that of the medium sized white patches, except that the smaller ones of the diameter of one line which were most numerous. There were also a few minute punctae of much less than a line in diameter. The patient evidently had watched the eruption closely for she said that the brown patches would become white after immersion of the parts in warm soap suds. or if they were scraped. This led me to scrape the surface of several and when the brown epithelial scales had been removed patches exactly like the white ones were seen. This convinced me that the brown patches were associated very intimately with the white ones. I observed a peculiar fact, however, namely, that while I could readily scrape the epithelium from a large patch for instance, one having a diameter of from two lines to one-third of an inch, I could not do so with the smaller ones of a line and under in diameter, as I then very readily induced bleeding. As distributed in the whole eruption the large brown ones were mostlp situated intermediate from the centre, there being white ones near them while at the periphery the smaller brown spots were principally seen. If the brown patches were pinched between the fingers or the forceps neither thinning or thickening of the integument could be appreciated, indeed it appeared normal as to structure except in color. But these patches were very perceptibly hyperaesthetic, and the patient said that when they began as small spots she experienced a pricking, painful sensation at various times during the day. This

continued, producing considerable discomfort until they reached their full size, when it ceased. It was for the relief of this sensation that the woman sometimes scratched the parts very actively, thus denuding them of their brown covering. I learned when examining the white patches by pinching up the folds that they were somewhat anæsthetic as very considerable manipulation was allowed. Indeed the woman said that there was not much feeling in them and exemplified the fact by pushing a pin into several of them. She said that on the thighs this could not be done as freely, and that the patches were a little more sensitive than on the arms. She also stated that very often she experienced a sensation of numbness in the white patches. As I have said before there were no abnormal nervous sensations in parts beyond the seat of lesion. I learned from the woman that these cutaneous phenomena had first appeared on the fore-arms two years previously and that the affection began on the legs shortly after. During this period she had suffered from general debility, and shortly before that time her menses had begun to grow scanty and had ceased a few months after the evolution of the cutaneous lesion. Since that time she had never felt as well as she had previously, her chief symptoms being weakness, headache, and an unusual nervousness as shown by spells of fainting and tremulousness. The woman's appearance at the time was that of a thin, weak and delicate person whose nutritive processes were performed in a faultly manner and whose assimilation was imperfect.

By questioning her I learned that the brown patches grew very slowly and that fully six months elapsed between the time of the appearance of a small brown spot and its full development as a patch of the largest size mentioned. It is interesting also to know that when they reached maturity, a slight and imperceptible desquamation occurred in many, undoubtedly induced or accelerated by friction, and that they of themselves assumed the white appearance described. Still it was evident to my mind that the superficial layers of epidermis in most instances remained adherent until the atrophic changes beneath had become complete. On some of the brown patches I observed a few minute scales nearly detached but still quite firmly adherent. I also noticed that at the time when patches were denuded of the brown layer they were of a more shining white and characteristic pearly appearance than those from which the epithelium had been sometime before removed, and as the white ones became old, they lost in a slight degree their glossy appearance, yet even in very old patches the lustre was not much dimmed.

It is very evident that the lesion in this case was an atrophy of the whole structure of the skin, in the form of round and oval spots, and that this was essentially the process from the commencement of the affection, rather than a thinning which follows a pre-existing infiltration of the skin, such as we see frequently follow certain forms

of tubercular syphilides, lupus and morphæa. It is, then, as far as our knowledge permits us to state, a tissue degeneration, idiopathic in nature, whereas in the case of the other affections the atrophy is merely symptomatic. My opportunities for its observation were limited to three examinations, but I think that the facts which I gathered from the patient, as well as the appearances which I noted, were sufficient to enable me to form correct ideas, as to the course of the morbid change.

The first appearance of an abnormal condition of the skin consists in the development of a minute brown spot of the size of the head of a pin, which does not seem to be connected with a follicular opening. This spot grows larger very slowly and gradually, and fully six months elapse, and in some instances longer, before it attains a diameter of half an inch, which in my case seemed to be the limit of development, as none of the patches exceeded, and only few reached five-eighths of an inch. Of course no idea could be formed of the condition of the skin as regards thickness when the spots were very small, but when half of their mature size the atrophy could be clearly made out. In general the browned epidermis remained adherent to the patch until the latter reached a diameter of about a quarter of an inch, and then it was shed, leaving a white surface, but still some patches which were fully developed, had the peculiar brown appearance. In the very early stages, this brown covering was not readily removed by scraping, but when about or nearly half the size of full development it could be scraped off readily, and in a mature state there seemed to be a tendency to its desquamation. There could not, however, be said to be present a desquamative process as there were merely patches covered with effete and altered epidermal cells, which were either removed artificially or which were shed in the friction to which the parts were subjected, and when once thrown off, there was no tendency to renewal, but on the contrary, a tissue of exceptional smoothness was left. The woman stated, and was positive of the fact, that the white patches when of half size grew larger, and it is probable that as they increased, the epidermis becoming altered was slowly and imperceptibly cast off. Thus it was that there were no patches seen with a white centre and brown areola. All of them did not reach full development as there were several of half the size of the largest ones. Stated succinctly, the process is as follows: a slight brown spot appears on the integument which gradually increases in size until it reaches a diameter of one-half an inch or rather more. At this time or perhaps before, the most superficial epidermal scales are cast off, and then a very peculiar pearl white spot is seen, which if examined is found to be much thinner than the surrounding skin. The surface of the patches may be studded with minute holes. In fact the atrophy of the deep structures took place synchronously with the increase of the brown spot. As said before, the process of thinning seemed to invade the whole texture as it was evident that the subcutaneous adipose tissue was also much absorbed. The symmetrical development of the eruption

is an interesting fact, and the question as to whether this is the rule or the exception, can only be settled by more extended observation. It was interesting to note that after the cessation of the atrophic process no further change was noted, such as the contraction of the spots or the formation of scar like bands. As to the etiology little can be said except that the accompanying symptoms are referable to some disturbance of innervation. During the progress of the affection, slight pain and hyperæsthesia were experienced, and after its cessation there was marked anæsthesia. These facts are significant as tending to give weight to the suspicion that the process is a tissue degeneration due to some obscure faulty innervation; more cannot be said. It is well to remember that the woman was of delicate, frail build, and that she had experienced suffering in passing that critical period, termed the change of life, not being as well after that as she had been previously. When all of the features of such cases are considered, no mistakes in diagnosis are very liable to occur though the error perhaps might be made of regarding such a case as one of leucoderma; but very slight care and thought would soon suggest the points of distinction.

It may occur to those who have read Mr. Wilson's * descriptions of morphœa and who are in accord with his views that the cutaneous affection of my case was really that form of morphœa which that writer calls morphœa alba atrophica, but I think it will be clearly seen that such a view is erroneous. It certainly is to be very much regretted that Mr. Wilson is not sufficiently definite in establishing the character of his second form of that affection, and this fault is the more apparent as his account of the lardaceous variety is in every particular, excellent. We need not, however, do any more than allude to this form, as it is essentially an infiltration and different in many respects from the affection in my case. It is necessary, however, to a clear understanding of this matter that I should quote the leading passage of this writer's description of the atrophic variety: he says: "Morphœa alba atrophica vel anæsthetica differs from morphœa lardacea not so much in degree of insensibility of the skin as in the total atrophy by which it is attended. The patches are of the same figure and size," (in speaking of the lardaceous form he says that the size varies from that of a lentil to that of an English crown piece, and to the size of one's palm and even larger. T.) "the skin is as white, the lilac erythematous blush and tingling sensation which precede and the lilac border which surround them the same: *but there is no deposition of morbid matter in the texture of the derma and consequently, no marble-like smoothness, polish and hardness: the lardaceous deposit is absent.*" The italics are mine. He further describes the final condition of atrophy of the skin which somewhat resembles that seen in my case. Further on he says: "Just as elephantiasis anæsthetica and elephantiasis tuberculosa being varie-

* On Diseases of the Skin, Page 586 et seq. Am. Edition, 1868.

ties of the same disease may co-exist, so morphœa alba atrophica may be present with morphœa alba lardacea and sometimes the anæsthetic form precedes the deposition which subsequently takes place and becomes a stage of the lardaceous variety." The fact of this matter appears to me to be that Wilson has described correctly one affection, namely, the lardaceous morphœa and that he has classed a totally different affection as a variety of it, because in its course it resembles, in some particulars objective and subjective the final atrophic stages of true morphœa, and that while the latter is essentially a cell infiltration which may go on to complete atrophy, the other is an atrophic or degenerative process from the beginning, not one which follows a pre-existent infiltration. It is evident from Wilson's description that his second variety is really an atrophy from the beginning, evidenced at first by hyperæmia of a deep red or lilac tinge, whereas in my case there was simply a little increase of pigment in the rete malphigii showing a less active hyperæmia. There are certainly cases of morphœa in which the infiltration is very slight, but still in these there is first hyperplasia, then atrophy. This is the opinion of Fox * who in speaking of the atrophic form, the second of Wilson says: "In some cases the deposit of material is not so marked as in others, but there is atrophy together with condensation." Therefore I can but repeat that in order to avoid error and in following nature we must consider morphœa to be essentially an infiltrative process subject to degenerative change and that we must not class as morphœa certain essentially atrophic changes simply because morphœa may be complicated by that process and is attended by similar subjective symptoms. The symptoms of my case of atrophy riesemble very much those of true morphœa, there being in the early stages slightly painful pricking sensations and finally more or less anæsthesia. Summed up, then, morphœa is an infiltration. and is attended with symptoms which resemble those which accompany idiopathic atrophy of the skin, the latter being essentially a degenerative process, the former primarily a cell infiltration and subsequently an atrophy of various grades. As a clinical fact it may be well to remark that the cicatriform appearance left after morphœa is not usually as smooth as that following atrophia cutis idiopathica.

Members of the Society have undoubtedly met with the linear form of atrophy in which, over bony prominences and parts submitted to unusual tension, white, somewhat scar-like streaks, or lines of wavy or perhaps spinal course, or even nearly straight, and variously distributed, either obliquely or in the axis of a limb, are met with. As said before these are not exceedingly rare, and their appearances are well known, and have been fully described. Very little is said, however, as to their development. In the case already referred to, which was also seen by my friend Dr. Duhring, such lines were developed on the outer, and anterior and under surfaces

* Skin Diseases, Page 340, London, 1873.

of the thighs. The patient was a young man, who at the age of 26 began to grow fleshy. At this time, on the sites mentioned, streaks of pale red were seen. The pale red color developed into quite a rosy hue, and these streaks remained in this condition for one year and a half, when they gradually grew pale, until now they are of a pearly white. During the period of this development they gave rise to no symptoms, nor was any preternatural heat perceptible in the parts. When examined it was evident that the integument and subcutaneous structures were very much thinner than normal. During these two years the gentleman had gradually grown stout, having been previously rather thin. It is interesting to note in this connection that he is a man of light complexion, whose integument is of a thin fine texture, which becomes hyperæmic, from slight irritation. The atrophic changes in this case may have been induced by an unnatural tension, caused by the increase of the subcutaneous tissues, while the skin did not proportionately grow. The morbid change was accompanied by slight passive hyperæmia. I have noticed in several other cases, indeed in every case, four of which I distinctly call to mind, in which the form of atrophy occurred that the integument was of delicate texture. Kaposi is the only one who has published any microscopic observations of this condition. In a portion of atrophied skin which he examined he found the epidermic and mucous layer much atrophied, the latter lying flat on the corium. There were upon the corium no papillary prominences, nor in the mucous layer any corresponding depression, but with high powers, very slight, thin and short projections containing no blood vessels, could be made out. The papillæ then were absorbed. The connective tissue and elastic fibres consisted of very thin bundles, between which extremely few and slender blood vessels existed. The fat cells had been absorbed. It is evident from the description that the degeneration of the integument is very extensive.

In strict truth the term idiopathic atrophy is more appropriate to the round and oval spot form than to the linear variety, as in the former there is no abnormal condition of tension of the skin as a probable cause, while in the linear form this condition can generally be determined. Until, however, our knowledge is more extensive it is well to retain the explicative adjective.

125 East 12th Street, Dec., 1875.