Two cases of morphoea : with remarks on the disease and its differential diagnosis / by L. Duncan Bulkley.

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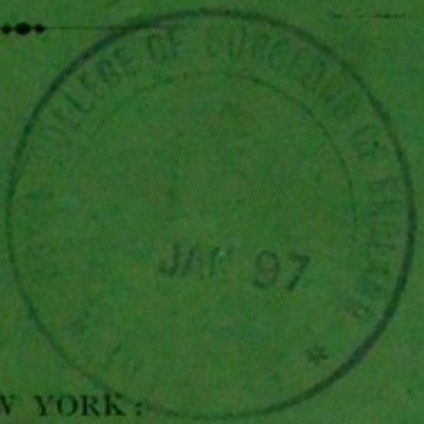
(6)

WITH REMARKS ON THE DISEASE AND ITS DIFFERENTAL DIAGNOSIS.

BY

L. DUNCAN BULKLEY, A.M., M.D.

Physician to the Skin Department, Demilt Dispensary, New York, etc.



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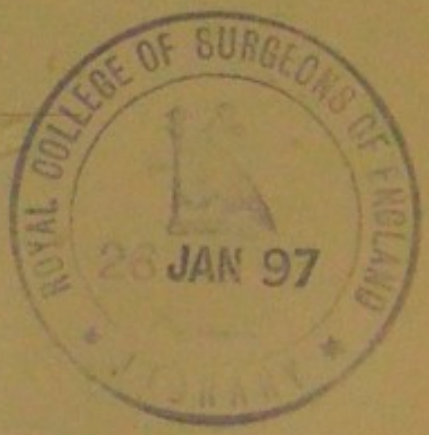

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TWO CASES OF MORPHŒA,

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MORPHŒA is a disease of which so little is known, and which has so recently acquired a definite and well recognized place in Dermatological literature, that each well observed case adds to our knowledge of it and enables us to make a more full study of the characters of this strange affection. The two following cases are, as far as I am aware, the first which have been described by an American writer, although I know that a number of cases have been observed in this country and the disease recognized by others.

By morphœa is understood the affection now thus designated by Wilson and Tilbury Fox, also by Fagge, Hutchinson and others, and which corresponds to or rather is the disease described by Addison as true keloid, although the latter from his description evidently includes the now well-recognized scleroderma under the same name. Fox thinks that morphæa bears certain relations to scleroderma, that indeed it may be a phase of it or it may exist alone. I cannot see any connection between the two, the morbid processes appear to be entirely distinct, as will appear later. Wilson declares that morphœa is related to elephantiasis Græcorum, true leprosy, that it is indeed the remnant of the disease handed down from former generations ; this view is but little borne out either by the clinical history or the phenomena of the disease, and, from the recent study made by the direction of the English government into the skin diseases of India and hot climates generally, it is found that morphœa is not known where the leprosy abounds, although a few of the reporters evidently committed the error of confounding spots of morphœa with the white spots of macular and anæsthetic

leprosy. Neumann and Kaposi have done the same, and fail to describe any disease corresponding to the cases given by Wilson and others, and those about to be presented, or to the description by Fox; the former, Neumann and Kaposi, make morphœa a synonym for the leprosy, elephantiasis Græcorum. We cannot here enter into the discussion of the differences between morphœa on the one hand and scleroderma and leprosy on the other, but after having studied a number of cases of both the latter diseases as well as the literature pertaining thereto, the conviction is fixed upon my mind that very essential differences exist between them.

With this much of introduction I will detail somewhat fully the two cases of morphœa which form the subject of this paper; the first of the cases has been under observation for more than two years, the second for nearly nine months, both of them have been seen repeatedly and watched and noted from time to time.

Case I. B. M., a tolerably healthy and well developed, but very nervous girl of 10 years, was recommended to my care by Dr. James R. Leaming of New York, on April 20th, 1874, for the treatment of her skin affection. She is the oldest of three children, born in the U. S. of American parents, and living in the central part of New York State; her younger sister and brother are perfectly healthy, as also an infant born since. Her mother is a large healthy lady, but very subject to sick headaches, father a delicate and very nervous man whose family had all died of consumption.

About fifteen months previous to her first visit, a spot was noticed on the right hip, just above the trochanter, which was thought to be caused by a bruise acquired in sliding down hill. Soon after a similar one appeared on a corresponding part on the left side, and very shortly one was noticed on the sacrum. These, when first observed by the parents, presented much the same characters as at her first visit to me. When first seen the following appearances were recorded: On both hips, just below the crests of the ilia, are seen patches of diseased tissue presenting similar appearances, they are about two inches by one in diameter, lying horizontally, of a dirty white or yellow color, with a very distinct pinkish margin. The diseased skin is dense and stiff, cannot be pinched up, and moves in a mass; the transition from healthy to diseased tissue is well marked and readily perceived by the touch, the mottled character of the spots stands in striking contrast to the healthy blush of the child's general integument. The surface of the spots is smooth, without desquamation, and is on a level with the adjoining skin. A very similar spot is seen over the sacrum, about one and a half by one inch in diameter. *On close examination the right thigh, which was thought to be healthy, is found to be of a mottled, purplish color, irregularly so, with here and there a small spot of the same yellowish, almost cadaver-like color, recognizable also by the touch; from these the red or purplish color seems to fade away, their margins being still considerably reddened; all of this redness is erythematous, disappearing momentarily on pressure, but the discolored patches undergo no

change on pressure. There are no other diseased portions on the body save those mentioned, nor does the preliminary erythema exist elsewhere.

During the time the case has been under observation I have seen in a number of places the same process gone through with, that is, the lilac or purplish congestion of the skin, mottled as though from the cold, upon which isolated spots, presenting the same whitish-yellow, cadaverous color and feel have developed. When they are formed their outline is very distinct to the sight and touch. There has never undergone in any of them any amount of contraction, the skin of the affected portions has been hard and dense, almost as though a piece of leather had been set in the skin, but when fully developed there seems to be little or no tendency to change, certainly very slight if any contraction of tissue such as occurs in scleroderma. Both thighs, both arms, and to a slight extent the forearms, the chest, and the face have been moderately invaded, the patches however keep, in the main, distinct and isolated, and at last accounts no impairment of function of any part had occurred in consequence of the disease. There was in some portions a slight depression of surface, but not at all marked; there were never any tubercular elevations.

Certain of the spots have seemed to improve very considerably under treatment, and for a while the disease seemed to be checked, but no permanent or very encouraging results have been obtained.

Case II.* Ann B., a well developed and apparently healthy woman of 30 years, unmarried, was first seen by me at Demilt Dispensary, February 29, 1876. About twelve months previously she noticed a red spot near the popliteal space, which gave but little sensation save occasional itching. The red spot gradually increased in size and soon became pale, and of the color and appearance now present. The disease has increased up to the time of first observation, but has been almost entirely checked during the last six months, since under treatment, or has spread very slowly upward while the lower portions have gradually resumed the normal state.

When first seen, the following appearances were recorded. On the right leg, commencing at a point about four inches above the center of the popliteal space, and extending downward to within about two inches of the internal malleolus, the skin is seen to be diseased, in a patch, irregular in shape and outline, from three inches at its broadest diameter above, to about an inch at other portions lower down, and turning round from the popliteal space toward the internal malleolus. The diseased surface presents the following characters: the margins are well defined and do not shade off into the healthy skin, as is shown by drawing the finger with moderate pressure, from the healthy on to the diseased tissue; the entire outline of the disease can be thus traced on care-

* This case was exhibited at the New York Dermatological Society, October 17, 1876. For discussion thereon see Archives of Dermatology, Vol. III., No. II., page 138.

ful palpation, with the eyes shut, as was demonstrated to and performed by a number of physicians present at the clinic. The affected skin has a hardened, leathery feel, or rather like that of pork-rind which has been cooked, and although it does not pit upon pressure, a sensation is given as though the tissue was infiltrated with some lardaceous or waxy substance ; it cannot be pinched up, but the resistance to this seems rather to be in the doughy thickening of the skin than in any fibrous, bound-down condition as in scleroderma ; pressure on it downwards and sideways moves a considerable extent of skin. The color of the diseased portion is of a brownish or dirty yellow (billiard-ball) hue, more or less mottled with light and dark patches of the same color, which surface contrasts very markedly with the slightly but distinctly reddened margin around its entire extent. This margin is erythematous, disappearing momentarily on pressure, and shades off insensibly into the normal coloration of the adjoining healthy skin, with an abrupt margin on the side of the diseased skin. Some portions of the diseased surface present a marble-like whiteness, especially toward the edge of the hardened skin, the central portions being darker. Where single lines of disease exist, as at the margin and near the ankle, there is a slight depression of surface, but in general the diseased portions are on about the level of the surrounding healthy skin, neither above or below.

Three weeks later she complained of pain running down the internal surface of the limb, and on deep examination a hard cord-like mass was felt deeper in the leg ; the diseased patches have changed in appearance, being now mottled and much of the marbly white color of portions is lost, they are also much softer and more movable. The line of demaraction is still red, but not so well defined as before.

On October 7, 1876, it was recorded that the patient was very much better ; that the lower portions of the diseased leg had lost a great deal of the thickening, and much of the marbly hue, the skin becoming natural, or a little shrunken. The upper part, just above the knee, posteriorly, was painful to the touch.

When last seen a week or so ago, some of the patches at the lowest portion of the limb had nearly disappeared ; there has been some extension of the disease upward since the patient has been under observation, but the progress, she thinks, has been much slower than previously. There has been no contraction in the diseased tissue as in scleroderma, and locomotion is not at all impeded, although almost the entire popliteal space is the seat of the morbid process.

The disease whose usual clinical history is very perfectly portrayed in the preceding notes of the two cases given, which were recorded at the time, (those of the latter case being dictated to my assistant, Dr. Robert Campbell, which case was also observed at the same time by six or eight physicians attending my clinic at Demilt,) presents several striking features, which should render its

diagnosis comparatively easy and should preclude the possibility of mistaking it for any other affection. The peculiarities of morphœa are briefly these ; the isolated or conjoined patches of dirty-white, or yellowish-brown, or mottled skin, of a firmness and density contrasting very strongly with the neighboring healthy tissue, and reminding one very much of a piece of moistened sole leather set into the skin, the erythematous halo around every patch of disease, and the almost entire absence of sensations in the part, except under certain circumstances, when a portion of the body subject to much movement is affected. These spots of hardened tissue are preceded by an erythematous redness, which contrasts with the history of *scleroderma*, which attacks fresh surfaces without preceding congestion, and does not present the pink or purplish border. Morphœa has little if any tendency to contract, whereas scleroderma sooner or later causes atrophy of the skin, and subcutaneous tissues and even muscles by the irresistible pressure from its steady contractile powers, and may even cause death by impeding or altogether checking the movements of the chest.

Morphœa is very chronic in its course, and the patches of disease may even appear to remain entirely stationary for a long time, and then slowly retrograde, and may disappear without leaving any trace of its former existence, though for a time there may be some cicatricial appearance and slight depression of surface.

Morphœa has certain very slight resemblances to *leucoderma*, from which, however, it is to be clearly distinguished. The marbly whiteness of the leucopathic patches might be taken for the white spots of morphæa, but there is no thickening or condensation of skin in the former. The halo around the patches of leucoderma is of a light brown, and does not alter on pressure as does the congestive halo around the spots of morphœa.

As already mentioned, morphœa has been confounded by many with the elephantiasis Græcorum, *anæsthetic leprosy*, and by some the two diseases are not at all differentiated. The patches of leprosy have not the hard, waxy feel of morphœa ; those of the former are very commonly, if not always, round or oval, those of morphœa irregular, and in leprosy other symptoms will very certainly appear, whereas patients with morphœa generally exhibit most perfect health. Moreover, leprosy, especially the anæsthetic and macular form, is a most uncommon and rare disease in this country, while morphœa, I am inclined to think, is not nearly so uncommon as formerly imagined. In morphœa the patches of cutaneous disease are fewer in number than leprosy, and generally slower in development.

Morphœa should never be mistaken for *syphilis*, as there is no lesion of this latter disease which can simulate it, even in a moderate degree. Single patches of morphœa may resemble *alopecia areata*, but in the latter there is no alteration perceptible in the tissue of the skin, no thickening or hardening, nothing but the falling of the hair, leaving a perfectly smooth, generally circular spot of marbly whiteness and smoothness, and there is none of the

congestive, purplish halo so peculiar to morphœa. Certain *cicatrices* resemble morphœa to a greater or less degree, but the antecedent history and a careful study of the surface should enable every one to recognize a cicatrix from a present and definite lesion of the skin.

In regard to the treatment of this interesting disease there is but little to add. Wilson advises to use local stimulation, Fox advises to avoid it. Both of my cases did fairly under a mild mercurial ointment gently and well rubbed in to the diseased patches. Both counsel tonic remedies; my first case, the child, did not seem any better with than without them; the second patient took none. It has occurred to me that iodine internally would promise more than any other remedy, as the clinical features of the disease suggest a lymphatic disturbance.*

Nothing is yet known in reference to the etiology or pathology of morphœa, and treatment must therefore be entirely empirical. It is hoped that further contributions of cases and careful studies of them, as also investigations into the microscopic anatomy of the disease may lead to more definite results in the future.

* I find since writing the above that Addison also suggests the use of iodine in this affection.