

TWO CASES OF DISSEMINATED SCLEROSIS, WITH AUTOPSY.

BY J. A. ORMEROD, M.D.

THIS paper gives an account of two cases of disseminated sclerosis, in which the diagnosis was established *post mortem*. The cases in which this diagnosis is made clinically, and no doubt correctly made, are nowadays very numerous, but completed cases seem to be uncommon. Partly for this reason, I venture to publish these two cases; partly, too, because the clinical symptoms appear to me interesting, and very different in the two patients, though both had the same disease.

In the first case, the disease was not diagnosed during life. There are many cases of disseminated sclerosis admitted to Queen Square Hospital, concerning some of which there can be little doubt as to the correctness of the diagnosis, while others of them present symptoms which are suggestive rather than convincing as to their nature. The bulk of these patients leave the hospital sometimes considerably improved, sometimes as incurable paralytics, and an anatomical proof or disproof of the diagnosis is never obtained. But in the present case this order of things was reversed. Disseminated sclerosis was not suspected during life. Indeed, there were no facts to suggest it. The patient's age, 54, was against it. The symptoms began in a very acute way. Pain was a prominent feature. The leading symptom at the time of admission, *i.e.*, intense flexor spasm of the legs, may certainly be seen in long-standing cases of disseminated sclerosis; but this patient had only been ten months ill. Thus the *post-mortem* discoveries were in every way a surprise. Nor did the localisation of the disease *post mortem* adequately explain the clinical characters of the

case; for in the cord most of the patches were found in the cervical region, yet there was no affection of the upper limbs, at least till a week before death. The dorsal and lumbar regions were less affected, and the descending degeneration of the pyramidal tracts was slight. Yet there was intense contraction of the legs, which had existed in one form or another from the very commencement, and incontinence of urine.

I fear that the rapid termination of the case must be connected in some way with the operation for straightening the legs. This procedure, though necessary and in no way severe, involving indeed no loss of blood, appears to have been too much for the resistance of an enfeebled nervous system.

The second case is remarkable for the hysterical phenomena which occurred in connection with it, and for a peculiar condition of the skin. The association of hysterical symptoms with disseminated sclerosis is, of course, no new observation; but I have seldom seen the diseases so closely interwoven as in this case. The patient (when first seen by me) had an extensive hemianæsthesia, which could be shown by Janet's test to be psychical in character; she had a rigid condition of the fingers, which yielded to forcible passive movement, though at the cost of some pain; she had an inability to walk which must have been largely "functional"; for though she had been bedridden for many months, she was able, after about five weeks' treatment, to get about the ward; and, indeed, had it not been for the presence of Babinski's sign and of optic atrophy, I think the diagnosis of hysteria, pure and simple, might have been justifiable. Yet the *post mortem* showed that she had, as was thought during life, disseminated sclerosis.

The rash was a very striking object when I first saw her, but the principal period of efflorescence was then over. She stated that it began as raised papules, which turned into "blisters," and this seems credible, for one bulla was actually observed. But most of the spots which developed in hospital began either as a small papule, which slowly increased in size and eventually formed a small sore; or as

a raised papule capped with a small excoriation, or as an abrasion or excoriation, generally of an elongated shape. The old healed places showed as oval patches of superficial atrophy, whose white colour was shown up by an edging of dark pigment. The rash only occurred on anæsthetic skin. Various views might be entertained as to the origin of it, *e.g.* :—

(1) That it was the expression of some organic nervous lesion. But no lesion was found *post mortem* with which it could be connected.

(2) That it was like the anæsthesia, hysterical. The existence of hysterical rashes is asserted by some authors, and as flatly denied by others. I myself have never seen anything like the present rash in cases of hysteria.

(3) That it was manufactured by the patient. There was much to be said for this view, but even after prolonged observation I could not prove it.

(4) That the anæsthetic skin had undergone some sort of nutritional change, in virtue of which it reacted in this peculiar way to slight stimuli which would have no effect on a healthy skin. As against this view, it should be noted that there was no rash on the (anæsthetic) right arm.

CASE I.

Onset sudden, ten months before admission. Extensive spasms of legs, with diarrhœa, repeated on four successive days. Further cramps in legs five weeks later, with pain, finally assuming the form of flexor spasms, lasting for some months. Gradual development of flexor contracture of the lower limbs.

Painful tinglings in legs six weeks after onset; incontinence of urine, with anæsthesia of the bladder. Rigidity of abdominal muscles two months before admission. Pain in back and lower limbs.

On admission, lower limbs firmly flexed at hip and knee; much tenderness to touch or manipulation; incontinence of urine. Legs straightened under chloroform two and a half months after admission; patient died of exhaustion some nine days later.

Post mortem.—Viscera normal; plaques of disseminated sclerosis in spinal cord, principally in the cervical region, in the pons and medulla, right optic thalamus, and right optic tract.

Case I.—(The notes of the case were taken by Dr. S. A. K. Wilson.) Charlotte L., aged 54; widow during last twenty-six years. Admitted to Queen Square Hospital under the care of Dr. Ormerod on July 21, 1905. Her father lived to 86, and her mother to 77; she had six brothers and sisters; one of them died of consumption, the rest were healthy. She had three children, two of whom died of consumption (there appears to be consumption in her husband's family).

She had fair health in early life. Eighteen years ago she had rheumatic fever, and scarlet fever at the same time; since this her heart has been "weak." She had had influenza three times, the last attack eleven years ago. There has been no return of rheumatic fever, but she had "rheumatism in the muscles of the back" six years ago.

It appears also that four years ago she had what she calls a stroke on the right side of the body; this lasted for twenty-four hours. Possibly this may have been a premonitory symptom of her present illness.

The present illness began quite suddenly on the afternoon of September 22, 1904. She was in her usual health, and was sitting resting, when, without any warning symptom, her legs became straight and stiff, with the toes spread out. At the same time she had violent diarrhoea. The legs remained in this condition for about two hours, after which time she could again bend them voluntarily. The diarrhoea lasted rather longer. There was then no pain in the back, no tingling or numbness, and no trouble with micturition. She knew of no cause for the attack, which took her completely by surprise.

The next morning, from 7 a.m. till 12 noon, she again had an attack of diarrhoea and extensor spasm of the legs; and on the two following days, during the same time, the attacks occurred again. After this it would appear that the legs, especially the right, began very slowly and gradually to draw up, the knee slowly approaching the trunk.

Six weeks after the first attack (in the beginning of November 1904) she began to have difficulty in holding her urine, and this incontinence has increased. In January 1905 she lost all sensation of the act of micturition, and cannot now tell whether the bladder is emptying itself or not, except by the feeling of warmth on the thighs.

About the same time, *i.e.*, about six weeks after the onset, a painful tingling sensation began in the legs "like a wound-up clock running down." It always seemed to run from the knees

to the toes. It usually came on in the evening and lasted several hours. This lasted for some months, but stopped about February 1905. In November 1904 bedsores began to appear, which lasted through the winter and spring.

To return to the muscular spasms. After the first three days these practically disappeared for five weeks. They then recurred in rather a different form, viz., as violent and painful cramps in which the legs went out straight and then drew up again quickly; these cramps were associated with severe tingling pains in all the muscles from the hips downward. About Christmas, 1904, these cramps were replaced by flexor spasms, i.e., she had attacks of pain over the hips and down the legs, during which the legs were drawn up spasmodically without any previous extension.

All this time the legs had been slowly contracting, and by Good Friday, 1905, the contraction had reached its worst, the right knee being drawn up, she says, under the left axilla, and the left similarly contracted. An operation for straightening them was undertaken, but soon afterwards they began to draw up again till they came to their present position. She cannot move them voluntarily, not even the toes. There has never been any loss of sensation; indeed they have been always, and are now, very sensitive.

She had had, almost from the onset, a tight feeling round the waist, but about two months ago the trunk muscles became more obviously affected, in that the patient had occasional pain across the belly, which became hard like a board. This condition is permanent, not paroxysmal. Constipation has taken the place of the initial diarrhœa.

During the last month she has suffered more definitely from pain in the back; this is severe, usually worst when the pains in the legs come on; it then shoots up into the spine, and she feels as if the whole of the spine were being forcibly doubled backwards.

She does not think the legs are wasted; she says that they were very swollen at first, but that this soon disappeared. There has been no affection whatever of the upper limbs.

CONDITION ON ADMISSION.

Patient is somewhat emaciated; she lies on her back with her legs drawn up and crossed, the right underneath the left. The legs are œdematous. There is an old bed sore on the sacrum. She has pain when the legs are touched or moved, but not otherwise. The abdominal muscles are rigid. Pulse 80; the heart is

somewhat enlarged, the apex being an inch outside and below the nipple, but there is no murmur. There is no evidence of lung disease. The urine contains no albumen.

Examination of Nervous System.

The cerebral functions are natural, though the pain makes her emotional. There is no affection of the special senses, except that she is deaf, especially with the right ear. In the district of the cranial nerves there is no definite abnormality except for a slight facial asymmetry, a slight and occasional ptosis of the right eyelid, and a little difference in the movements of the eyelids when she winks. The optic discs are both rather pale in the temporal half.

Motor Functions.—She cannot stand. The muscles generally and those of the legs in particular are small and flabby; the wasting in the legs is partially concealed by œdema. The legs are contracted; the right is flexed at the hip and knee and strongly adducted, so that it lies right across to the left side; over it is the left leg, similarly flexed at the hip and knee, but not adducted. Any attempt to overcome this contraction causes intense pain. The rigidity seems to be more in the joints than in the muscles, except as regards the hamstrings, the tendons of which one may feel to become taut. The gastrocnemii are peculiarly flabby and toneless. The glutei seem flabby, though not obviously wasted; their condition is more or less masked by œdema.

There are no fibrillary twitchings, no tremors, no reflex spasms, nor involuntary movements. Voluntary movements are normal in the head, neck, and upper limbs; in attempting to sit up the rigid abdominal muscles become somewhat harder than before; the erector spinæ cannot be tested. In the lower limbs the only voluntary movements obtainable are slight flexion and extension at the ankles, better on the right than on the left side. The attempt to move sometimes produces on the right side an incomplete spontaneous ankle-clonus.

Sensory Functions.—There is intense hyperæsthesia to touch and to prick in the legs generally, and particularly over the ankles and the soles of the feet. Stroking the sole of the foot to produce a plantar reflex seems to produce intense pain. Similarly, pressing the feet to estimate the œdema produces much pain. Any attempt at passive movement of the legs, particularly at the hips or knees, causes intense pain. There is pain in the back, about the level of the sixth dorsal area, and again very low down in the

small of the back, and also in the abdomen, coming round from the small of the back.

No definite anæsthesia can be made out, but she seems to feel a pin-prick more acutely over the abdomen than over the pelvis or thorax. There is no loss of the sense of position or of passive movement in the legs or in the toes. The sense of pain when the muscles are pressed is much increased.

Reflexes.—In the upper limbs the deep reflexes are all brisk and equal. The knee-jerks cannot be obtained on either side, probably owing to the position and contraction of the legs. The ankle-jerk seems to be present on both sides, though slight.

No abdominal or epigastric reflex can be obtained on either side.

Plantar reflexes.—Not obtained with certainty; only occasional toe movements follow the stimulation. (Later, however, plantar reflexes of extensor type were obtained.)

There is incontinence of urine as described above.

Vaso-motor and trophic functions.—The legs are oedematous, especially about the ankles. The skin is very thick, coarse, and dry, scaling off in flakes over the toes and feet. The nails are dry and cracking. There is an old bedsore over the sacrum.

October, 1905.—The patient is no better, if anything worse; the left leg is more contracted; she complains constantly of pain. The position of the legs gives rise to much difficulty in the nursing.

Consequently it was decided to straighten them under chloroform. This was done on October 7, at 3.30 p.m.; many adhesions were broken down, the limbs were made much straighter, and the knees separated. She recovered fairly well, but remained somewhat drowsy, and got worse towards the evening; the temperature rose to 100° F., the respirations became quick and shallow, and her colour rather blue.

October 8.—Temperature 100°; pulse and respiration are hurried, but examination of heart and lungs shows nothing abnormal. Drowsy and semi-conscious; right eyelid droops more than usual; swallows with difficulty. The arms are very rigid and resistant to passive movements.

October 15.—During the week the patient has been steadily failing. There has been continuous slight fever (99.5–101°), drowsiness, occasional cough; physical signs of hypostatic congestion at the basis of the lungs. The condition of the legs has remained unchanged; the plantar reflexes are of the extensor type on both sides. There appears to be more definite weakness on the right side of the face.

October 16 (6 a.m.).—The temperature rose to 105·8° F., and the patient died quietly soon afterwards.

The *post mortem* was made seven hours after death by Dr. Gordon Holmes, who has furnished me with the following report:—

AUTOPSY.

Body well nourished, *rigor mortis* present in all limbs, knees contracted and cannot be straightened out. Both feet œdematous.

Visceral organs normal, except that the lower lobes of both lungs are congested.

Vertebral column and skull, normal.

Spinal cord.—Roots and meninges appear normal; cord of fair size, and no discoloration visible on surface. On drawing finger down cord a sensation of ridges is obtained, as though the consistence of the tissues were irregular. On section through it there seems to be irregular patchy disease in the cervical and dorsal segments.

Brain.—Vessels and meninges appear normal. No wasting nor other evident disease of the cortex; no patches of sclerosis visible in it. On cutting sections of the brain, the only patch found is in the posterior part of the right optic thalamus. Small patches of discoloration are visible on the ventral surface of the pons. (When further sections of the brain had been made after hardening, several small foci of disease were seen in the pons, and one in the mid-brain in the neighbourhood of the right external geniculate body.)

Microscopical Examination.

Cord (stained by Weigert-Pal method).—The cervical segments contain the chief evidence of disease. Here there are numerous sclerotic patches in which no myelinated fibres stain; they have, as a rule, a sharp border, though in some regions the sclerosis is more diffuse, and some medullated fibres may persist in the area which appears diseased. The patches do not seem to have any special site of predilection; grey and white matter are equally affected.

In the dorsal and lumbar segments there are fewer definite patches such as are typical of the disease; but foci of diffuse sclerosis, in which some myelinated fibres persist, may be seen in many segments.

There is in addition slight descending degeneration, at least of the myeline sheaths, of both pyramidal tracts; but that of the one side (? right) is considerably more affected than the other.

No definite ascending degeneration is visible in the posterior columns, nor in the spino-cerebellar tracts.

Brain-stem. — A large patch of sclerosis is visible just at the decussation of the pyramids, but no others were discovered in any of the levels of the medulla which were cut.

In the pons is a patch lying in the floor of the fourth ventricle, and at another level a large focus just ventral to the seventh nucleus.

In the mid-brain are a few small foci, situated in the ventro-lateral periphery of the section, at about the level of the posterior quadrigeminal bodies, and two patches are seen in the right thalamus, the larger of which, measuring about $\frac{1}{2}$ cm., lies in the pulvinar.

In sections through the chiasma it is found that one optic nerve had lost all its myelinated sheaths for a considerable distance, and there is some diminution in the myelinated sheaths of the other. The eyes were not examined.

Bielschowski's stain show that the greater number at least of the axis cylinders in the sclerotic patches are normal.

Sections prepared by Weigert's method for demonstrating the neuroglia were not very satisfactory, but they show a dense sclerosis of neuroglia in the foci of disease.

No evidence of vascular disease in the central nervous system can be found.

CASE 2.

Woman of middle age, who had had many illnesses. Lymphangitis with abscesses of right upper limb; splint applied; stiffness of elbow and fingers followed, and paræsthesia in fingers, spreading to right side of trunk. Afterwards abscess right thigh, weakness right lower limb; peculiar rash right thigh and right side of body.

On admission to St. Bartholomew's (January 13, 1905) three or four years after onset she was completely bedridden. Right upper limb was rigid, and particularly the fingers, where there was great pain on passive movement; right hip rigid, but less so than upper limb; wasting of right limbs, especially the upper limb; right hemiansæsthesia, and a rash of peculiar appearance and distribution. Plantar reflexes extensor; optic atrophy (observed later). During next few weeks much improvement in motor condition of right limbs; slight spread of the eruption.

Transferred to Queen Square four and a half months later (May 29). Still extreme rigidity of right fingers; anæsthesia has

spread upwards to right clavicles; rash now stationary. No further change for the next three or four months, then (September-October, 1905) spread of symptoms to left side, viz., anaesthesia left as well as right from neck downwards, weakness of left limbs, jumping movements, and disagreeable sensations in left limbs; further slight spread of rash; general debility and depression.

She left Queen Square at Christmas, 1905, and died of abdominal cancer in November, 1906, i.e., some five or six years after the onset.

Examination of the nervous system showed the presence of disseminated sclerosis.

Case 2.—This patient was an inmate of Tonbridge Infirmary, under the care of Dr. Crawford, before she came under my observation. I must thank Dr. Crawford for sending her to me, and for much valuable information and help. I must also thank Dr. W. T. Storrs for drawing my attention to the case in the first instance.

Alice L., aged 38, single, was admitted to St. Bartholomew's Hospital, January 13, 1905.

In her family history there is nothing that needs mention.

She herself has never been robust. As a child she had "croup," frequent sore throats, and bronchitis. At the age of 13 she had rheumatic fever; at 17, muscular rheumatism in the legs, which laid her up for three or four months; at 19, severe diphtheria; at 22, "congestion of the lungs"; at 31, typhoid fever; at 33, an illness which began with an ulcerated throat, and which led to cough and short breath, with expectoration of blood and offensive pus. She was ill for three months.

During convalescence from this, i.e., between three and four years ago, her present illness began, and in the following way. She scratched her right thumb on a brass bracket, lymphangitis followed, and an axillary abscess which discharged for a month or more; then an abscess appeared at the elbow, lasting several weeks. The elbow was put on a splint; it has remained stiff ever since. The fingers of that hand also became stiff; the limb became wasted and weak. Pricking and tingling sensations began in the fingers of the right hand (not in the thumb apparently) about a month after the accident, associated with a feeling of numbness, and this numbness spread up to the shoulder and on to the right side of the trunk. It appears from statements made to Dr. Wilson at Queen Square that similar sensory pheno-

mena, viz., tingling and numbness, began in the right foot and toes, particularly the great toe, about the same time as in the hand, and extended up to the hip, though the numbness was not always present; the limb felt weak to her, and the ground under the foot "felt soft and funny."

All active symptoms, however, had subsided a year after the injury, but the arm had settled down into its present state of rigidity and uselessness. She remained in this state about nine months.

Then an abscess formed in the front of the right thigh which laid her up again, and while this was still discharging the leg got gradually weaker, and an affection of the skin began. A series of spots, about half a dozen in number, appeared in the neighbourhood of the abscess—raised red papules, about the size of a six-pence, which in twenty-four hours turned into blisters, and finally formed sores which took a long time to heal. This was in February, 1904. In May a fresh crop appeared, and they continued to come out during the rest of the year, spreading down the right thigh and leg, and upwards over the right half of the trunk. Over seventy were counted.

The outbreak of a fresh spot was preceded by local pricking and tingling; they were painful, but did not itch.

She has been in Tonbridge Infirmary more than a year. Dr. Crawford writes me word that, having suspicions as to the origin of the rash, he directed the nurses to watch her carefully, but they never detected her manufacturing it. She had also severe general pains which kept her awake, and for this she had been given, before her admission to the infirmary, morphia subcutaneously, which relieved her insomnia, but gave her severe headache in the morning. Hypodermics of plain water were substituted in the infirmary, and produced exactly the same result. She was given a long course of arsenic, which may have caused the peculiar pigmentation round the rash.

CONDITION ON ADMISSION TO ST. BARTHOLOMEW'S
(January 13, 1905).

(The notes are mainly abridged from those taken by Dr. G. B. D. Adams, then house physician.)

Patient is a woman of rather spare build. Complains of pain in the right hip. She lies quietly in bed with the right arm close to the side, the right elbow (which is stiff) semiflexed, and the forearm laid across the body. The temperature, pulse, and respiration are normal. There is some lateral curvature of the spine, convexity to the left, in the dorso-lumbar region, and some

slight deformity of the chest owing to prominence of the third rib on both sides. Examination of the chest and abdomen shows nothing abnormal. The teeth are bad; her hearing is good; she has bad eyesight, for which she wears spectacles. The pupils are normal, the ocular movements are normal, there is no nystagmus. An examination of the fundus oculi was unfortunately not made, or at least not recorded.

The facial movements are the same on both sides; there is no loss of sensation in the face.

Condition of the Limbs.—The left arm and left leg are natural in appearance, and their movements are natural; but on the right side the upper arm is slightly wasted ($7\frac{1}{2}$ ins. circumference as compared with $8\frac{1}{2}$ ins. on the left); the muscles of the forearm and hand, especially the interossei, are markedly wasted. The movements at the shoulder-joint are good, those at the elbow limited; flexion is possible to an angle of about 45° , extension to an angle of 120° , but the joint is not definitely deformed or thickened. The movements of pronation and supination are free, those at the wrist-joint free but weak. The hand is useless, the metacarpo-phalangeal joints are slightly flexed, and there is a corresponding over-extension of the proximal interphalangeal joints; passive flexion of the fingers is possible, but there is much resistance to it, and it gives her great pain. She cannot voluntarily flex the fingers nor separate them.

The right lower limb looks shortened at first sight, but this is apparent only, and due to tilting of the pelvis and adduction of the limb; measurement shows it to be of the same length as the left. The limb is adducted and very slightly inverted, but not flected. Tapping over the great trochanter or on the sole of the foot causes pain in the hip-joint. The movement of the right hip-joint is very limited; passive flexion is possible only through an angle of 25° , and causes pain. The muscles of the thigh are wasted (circumference $13\frac{1}{2}$ ins. as compared with 14 ins. on the left side); and those of the calf are also rather smaller than the left.

There is no affection of micturition or defæcation. The knee-jerk is more marked on the left side than on the right; there is no ankle-clonus on either side. The plantar reflex is difficult to obtain on the right side, on the left side it is distinctly extensor in type. An extensor plantar reflex was subsequently obtained on the right side also.

There is anæsthesia to touch, prick, and thermal stimuli over the right arm, the right leg, and the right half of the lower part of the trunk. In some places, however, the patient still feels a prick (see fig. 1 made out by Mr. J. G. Watkins, clinical clerk).

The rash, which formed a striking feature of the case, was distributed over the lower half of the right side of the trunk, the right thigh, and part of the right leg. Part of the buttock adjacent to the fundament had no rash upon it. (See fig. 2 by Mr. Watkins, and the photograph.) Thus, though the whole of the anæsthetic area was not affected (the arm, for instance, being free), yet there was no rash on such parts of the skin as retained sensation. On the trunk it was most profuse from the groin to the level

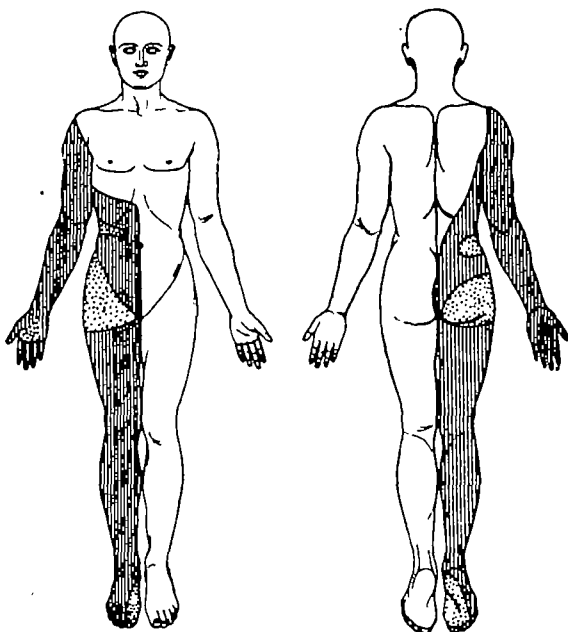


FIG. 1.

Distribution of anæsthesia, January 14, 1905. Anæsthesia to touch, prick, and temperature, over dark area; over dotted areas, a prick is still felt. (Mr. J. G. Watkins.)

of the eighth rib in front, and from the buttock up to the lower angle of the scapula behind; it stopped abruptly at the middle line, both in front and behind, except where some pigment spread across the sacrum to the inner aspect of the left buttock. There was rash all over the thigh, and a few separate patches on the outer side of the calf; none on the shin.

It consisted of spots and patches which were for the most part discrete. The most recent forms appeared to be red, slightly

raised papules, about half an inch in diameter, with a small abrasion at their apex. Other forms were shallow ulcers or abrasions, where the cuticle had been lost, leaving raw red patches of an oval shape for the most part, and from one to two inches long. These oval lesions were placed vertically (in the long axis of the limb) on the thigh, in the groin and neighbourhood obliquely (parallel with Poupart's ligament); on the trunk they were irregularly disposed. Each was surrounded by a zone of brown pigmentation. There were also older lesions, consisting of white

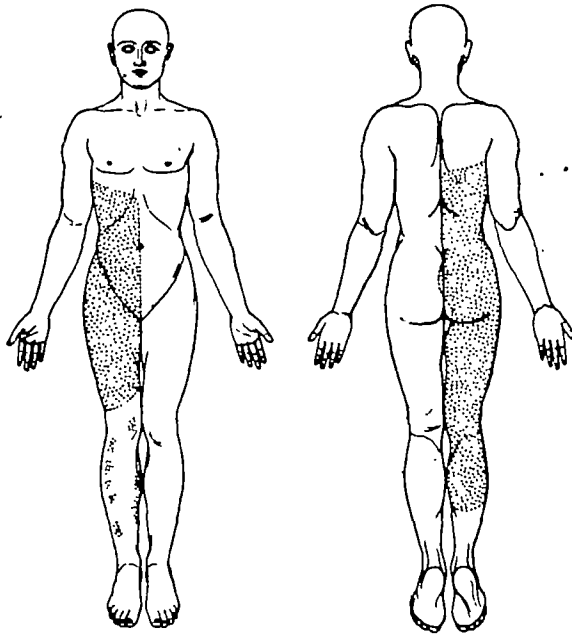


FIG. 2.

Distribution of the rash, indicated by dots. January 14, 1905. (Mr. J. G. Watkins.)

scars, oval or irregular-shaped, each surrounded by a zone of pigmentation. On the front of the thigh the scarring and pigmentation were confluent. There was no indication of the site of the abscess, which was said to have occurred in the front of the thigh.

It will thus be seen that the case presented peculiar symptoms, difficult to interpret; a cursory review of them suggested some such diagnosis as the following:—

That owing to some infection the elbow-joint or the bone of

the upper limb had become diseased, and that the nerve trunks had become involved; hence paralysis with atrophy of the hand muscles, and possibly distortion of the fingers.

That the hip-joint was similarly diseased.

That the unilateral anæsthesia and unilateral rash might be due to some obscure (infective ?) disease of the spinal nerve-roots on the right side.

But these suppositions were disproved by further investigation of the facts. For in the first place, Mr. Lockwood, who kindly examined the joints for me, said that they were not diseased. Secondly, Dr. Lewis Jones reported that the electrical reactions of the wasted muscles were normal; thus the existence of a neuritis, or of disease of the anterior cornual cells, was negatived. Thirdly, the anæsthesia was proved to be psychical in the following way:—

After ensuring that she could not see what was being done, the patient was first touched several times on the sound parts, and told to reply "yes" each time she was touched. She did so correctly. She was next touched on the anæsthetic part, and told to reply "no" each time she was not touched. She did so, equally correctly.

After a few trials, this test (as usual) began to fail, so it was varied. Her attention was drawn to the paralysis of her right hand rather than to the anæsthesia, and she was told first to touch with her right forefinger any part that I touched. I touched a sentient part of her body, and she laboriously touched it with her right forefinger. I then told her to do the same with her left hand, taking care to remind her that this was her good hand. I touched an anæsthetic part, and she followed suit quite readily. This test also began to fail after a time, but in a curious way. When told, as before, to follow with her left hand, she would lift it up to do so and then drop it as if she could not carry out the command, but betraying all the same that she had felt the touch. And yet, all the time, the anæsthetic parts, when tested in the ordinary way, were anæsthetic to stimuli of the severest kind.

I concluded from this examination that the anæsthesia was hysterical, and thought it probable that the paralysis was also largely hysterical. The rash remained a puzzle, but it seemed to me not impossible that the patient had produced it herself. The leg was covered with boric acid ointment and bandaged, and a careful watch was kept for the appearance of any new spot, and the circumstances under which it might arise. Although some of the notes read rather like conviction of a fraud, I think I may say the verdict was "not proven" one way or the other.

I give the notes verbatim as regards the rash.

January 20.—A few new spots have occurred, especially lower down the calf of the right leg.

January 21.—A new blister appeared yesterday, containing clear fluid. It collapsed this morning. There is now a reddened oval area 1 inch long, without pigmentation round it.

January 31.—Two or three new spots have appeared in the right axilla and thigh.

February 7.—Two or three more spots on the back of the leg and in the axilla.

February 21.—One or two new spots.

March 14.—One or two new spots, having the character of abrasions.

March 21.—Scratched left hand with a pin, rubbed it with the fingers of the right hand, and made a sore place, discharging pus.

One or two new spots.

March 28.—Two or three (new) small shallow ulcers on the inner side of the right ankle.

April 4.—One new ulcer since last week on the inner side of the right ankle.

April 11.—Three new lesions transversely placed across right instep, produced by friction with left heel (observed).

May 16.—Two new lesions in groin, and two just below the knee.

The excoriations which had been present on admission were healed by March 8. Of the new spots which developed, some were in points accessible to the patient, some under the bandage. Most of them took the form of red raised patches with an abrasion on their summit, though a definite bulla was once observed (January 21).

The patient was shown to the Dermatological Society of London on March 8. The general view taken by the Society was that the rash was manufactured; but one member, whose opinion should be of great weight, dissented.

The patient was treated with passive movements of the limbs, and the electrical wire brush. The anæsthesia did not clear up, but the motor condition improved remarkably. Thus, whereas she had been bedridden for many months before admission, by February 21 she could walk with very slight assistance, and by March 7 she was walking alone, though she still kept the right leg stiff as she walked. The elbow also soon became flexible, but very little impression was made on the condition of the fingers; they re-

mained in their peculiar position. Passive flexion of these was possible, but caused her intense pain.

A note taken by me on March 7 says that the anæsthesia is still present; the knee-jerk on the left side is easily obtained, on the right it is obtained with reinforcement; there is no ankle-clonus; the abdominal reflex is not obtained; the plantar reflexes are extensor, the left typically so.

It was recognised, in the presence of this distinct Babinski's sign, that the disease was not wholly hysterical.

She was transferred to Queen Square Hospital on May 29, 1905.

The notes in Queen Square Hospital were taken by Dr. S. A. K. Wilson, who took a great interest in the case; I have abbreviated them for the purpose of this paper.

The history corresponded in the main with that just given; one additional point is that the patient stated that for the last seven years she had had to hurry somewhat to pass urine, *i.e.*, had had some degree of "precipitate micturition." She herself acknowledged the improvement in the movements of the lower limb and of the elbow that had taken place while she was at St. Bartholomew's, but in the hand she said (correctly enough) there had been little change.

CONDITION ON ADMISSION TO QUEEN SQUARE (May 29, 1905).

In the district of the cranial nerves there is no abnormality, with the one very important exception, that she has double *optic atrophy*. Mr. Marcus Gunn's note on this point (June 5) says: "Both discs are decidedly pale and atrophic in appearance; the right is the better looking of the two, there being still a pink tinge over the inner half. Vessels of good size, lamina cribrosa well seen. Whole appearance is that of a simple optic atrophy. Eyes nearly emmetropic."

The colour vision (central) is normal on both sides; the field (for white) is reduced to a very small central area.

Motor Functions.—The general muscular development is fair, with the exceptions to be presently noted. There is no kind of involuntary movement.

Of the upper limbs, the left is normal; in the right there is slight general wasting, especially below the elbow; passive movements at the shoulder give rise to some resistance and pain; voluntary movements are also limited here, as if from stiffness of the shoulder-joint; movements at the elbow and wrist are good in range, though somewhat weak; but the fingers are rigid and

fixed in a peculiar position, viz., the interphalangeal joints are fully extended and very rigid, but the terminal phalanges are flexed on the others (particularly in the first, second, and fourth fingers). She cannot close her fist at all; the attempt results in an interosseal position. Any attempt at passive flexion of the rigid joints gives rise to intense pain. The thumb, however, is not affected, and she can move it well.

As to the lower limbs, the left is natural; there is slight wasting and flabbiness of muscles in the right, at the right hip there is some pain and resistance upon passive movement, and the voluntary movements are weak. Similar resistance is noted at the knee, and there is some grating of the patella. Voluntary movements of the ankle are fair in range, but feeble in power; movements of the toes are fair.

There is no incoördination, so far as can be made out in the presence of feeble and defective movement.

Sensory Functions.—Subjectively, there is pain in the right shoulder, knee, and hip upon passive movement, and very great pain when an attempt at passive flexion of the right fingers is made. There is numbness on the right half of the body, from the neck downwards.

Objectively, it will be seen by the chart (fig. 3), as compared with those made in St. Bartholomew's, that the anæsthesia has spread up to the level of the clavicle. From this point downwards the whole right half of the body is completely anæsthetic to touches and to pricks, except for some parts indicated by dots on the chart, viz., the upper edge of the anæsthetic area, the thumb, especially the tip of the thumb, the hypogastrium and region of the anus, and parts of the sole. Here a prick is felt as a touch. Janet's test, however, can no longer be obtained. The perception of heat and cold and the perception of vibration (pallæsthesia) are also lost over the right half of the body from the clavicle downwards, again excepting the thumb. The sense of weight as tested by balls, the muscle pain sense, and the sense of position are all absent in the right limbs. Thus she cannot tell when the limbs (right) are being moved passively, excepting the right thumb: she cannot imitate with her left arm the position into which her right arm has been put; indeed, she does not know where her right arm is till she opens her eyes and looks at it. As to stereognosis, if the patient gets an object between the right thumb and forefinger, she makes a fair attempt at guessing what it is, but in any other part of the hand there is complete astereognosis. It may be, however, that she is hindered from recognising objects by the rigid state of the fingers.

On the left side of the body, and in the left limbs, all forms of sensation are normal. There is no loss of visceral sensation; for instance, she knows when the bowels move and when her urine passes.

Reflexes.—Tendon reflexes are normal in upper and lower limbs (excepting that a contralateral adductor reflex is present, on testing the knee-jerks on both sides). There is no ankle-clonus.

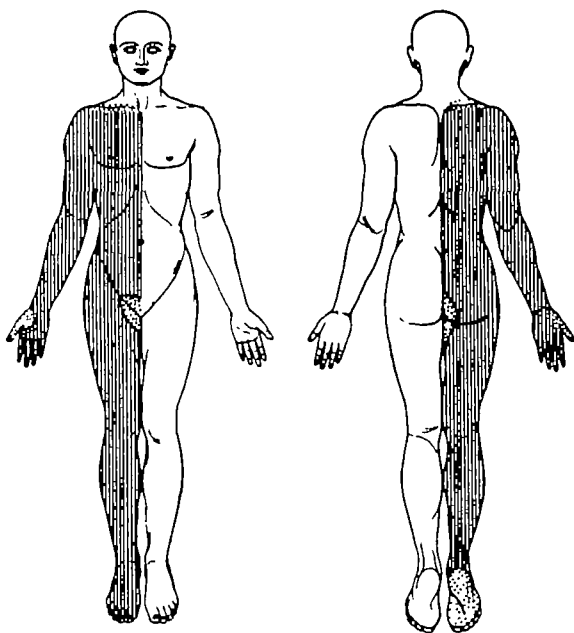


FIG. 8.

(Dr. S. A. K. Wilson.) May 29, 1905. Over dark areas, complete anaesthesia to touch, prick, temperature, vibration. Over dotted areas prick is recognised as a touch. At tip of thumb it is recognised as a prick. Over thumb, too, sense of temperature and of vibration is preserved.

The abdominal and epigastric are present on both sides; the plantar are of the extensor type on both sides, but the left is more marked, or, at least, more easily obtained than the right.

The organic reflexes are normal, except it be for the precipitancy of micturition mentioned above.

Gait.—The patient advances rather slowly, dragging the right foot. The chief step in advance is always taken with the left foot; the right is then slowly brought forward and shuffled along

the ground to a position very little in advance of the left. On the right foot the chief weight is thrown on the heel, the toes are sometimes off the ground. There is no ataxia of gait, and no Romberg's sign.

As to the condition of the skin, it is unnecessary to describe the rash in detail again. There are now only two or three of the spots which have not quite healed; and these healed places present the appearance of pale, non-depressed scars, mostly oval in

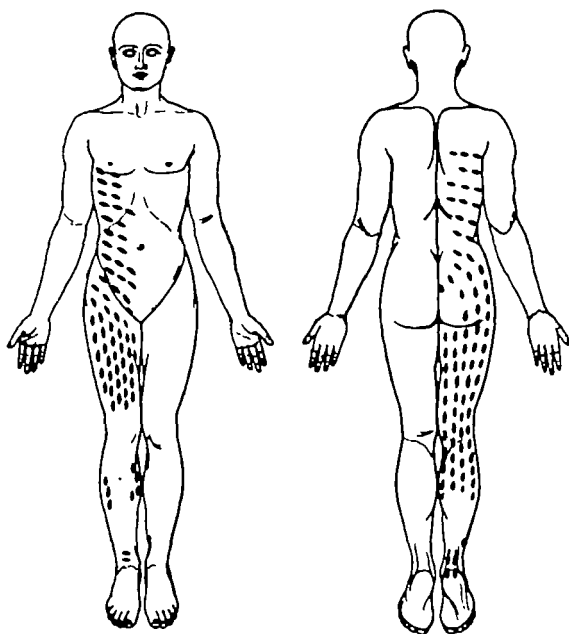


FIG. 4.

(Dr. S. A. K. Wilson.) May 29, 1905. Distribution of the rash, and direction of the long axes of the oval spots.

shape (the long axis having the directions described in the former note), surrounded by a deep zone of pigment, and closely packed.

Electrical Examination.—The muscles on the right side require a stronger faradic current to produce reaction than do those on the left; to galvanism there is no reaction of degeneration. Over the anæsthetic area she bears a very strong current, apparently without feeling anything—a current strong enough to produce firm tetanus of the muscles—and which on the left side is quite unbearable.

Later (July 15) a lumbar puncture was made; 15 cc. of fluid was withdrawn under considerable tension; the fluid was clear, and, when examined histologically, proved to be quite normal.

During the summer no particular changes were noted in the patient's condition; her general condition remained good, her weight increased by a few pounds (May 29, 7 st. 9 lbs.; October 1, 8 st 1 lb.); her temperature was never above 99°; but in the autumn her nervous state, and particularly the sensory condition, began to get worse. Thus a note of October 15 says:—

October 15, 1905. For the last few weeks the patient has had occasional sensations which she tries to describe as follows: "Sometimes I feel as if I weren't anywhere. Yesterday I felt this in the day room; I had to think whether I was there or somewhere else. I feel constantly as if there were no right half to my body, as if it did not belong to the other half."

About six weeks ago she noticed that the left hand and arm were becoming numb. This began with a sensation of pins and needles in the ulnar fingers, spreading up to the elbow. About a week ago it had reached the shoulder. She has felt increasing weakness and awkwardness in the left arm and has been dropping things. Her writing, which used to be wonderfully good with the left hand, is distinctly worse. The sensations in the left arm are distressing her very much because the paralysis of the right arm began, she says, exactly in the same way. Within the last few days this numbness of the left arm has spread and become much more intense. "I feel nothing on that side now; when I take anything in my left hand I must look at it, otherwise I do not know that I have it. I do not know whether my left hand is open or closed unless I look at it."

Within the last week also a sort of irregular spasm has been noticed once or twice in the left arm and leg, consisting in occasional twitches of various muscles and muscular groups. On occasions it is much worse, she says: "Sometimes my leg nearly jumps off."

A careful re-examination of her general state was made (October 15) with the following results:—

There is no particular change in her general appearance. She appears anxious to improve; she does not know why she is getting worse. She has headaches and sleeps badly. She has a cough, obviously neurotic, which increases when attention is paid to it.

Cranial Nerves.—There is no change in the optic discs; there is a slight tendency to nystagmus upon extreme lateral deviations in either direction.

Motor Functions.—The weakness of the right side increases, the fingers are perhaps less stiff, but there is no return of power and they are excessively painful when passively flexed, in spite of the general and complete anæsthesia. There is, in addition, weakness of the left arm; there is no definite limitation of movement, but all movements are very slow. She puts her forefinger to the nose with exceeding slowness, and with irregular to-and-fro movements of the arm. It seems as if she were unable to sustain any muscular effort. There is no wasting of this arm, no rigidity or contracture, but there are occasional irregular muscular jerkings, chiefly in the biceps. There is some incoördination of both arms and legs.

Sensory Functions.—The anæsthesia has spread to the left side, so that the face, scalp, and outer side of the left leg are now the only normal parts (*vide* fig. 5). The anæsthesia and analgesia are absolute; there is complete loss of pallæsthesia over the same areas; there is complete loss of muscular sense in all its forms. She cannot feel the muscular contraction when faradism is applied so strongly as to tetanise the whole limb; she cannot tell in what position the limbs are placed, nor say when they are being moved. This loss of sense of position corresponds to the anæsthesia of the skin, it does not involve the right thumb or the left toes, in which there is no cutaneous anæsthesia. There is also complete astereognosis in both hands. A needle passed through the left supinator longus causes no pain and draws no blood.

Reflexes.—The tendon reflexes are brisk in the left arm, less so in the right; the others remain as before. The plantar reflexes are extensor on both sides.

The skin lesions have not been very obtrusive; occasionally new ones appear; these are invariably preceded by a small papule, which slowly increases during one or two days; it is raised above the general surface of the skin and quite painless. It then breaks open and leaves a little sore. The position of these lesions bears no relation to the bandages which are kept applied.

October 21.—For the second time since admission she has developed a functional aphonia, as a sequel to a follicular tonsillitis. Though she cannot speak, to order, above a whisper, yet she sometimes, as the sister and nurses say, forgets her aphonia and speaks quite naturally. Similar facts have been noticed about her paralysis. Thus she has once or twice either buttoned or unbuttoned her night-dress, which she says she cannot do, and then has gone and asked a neighbouring patient to do it for her.

Twice since the onset of weakness in the left arm, she has involuntarily brought the left hand to her mouth to stifle a cough with quite ordinary briskness, though the same movement, when done voluntarily, is performed, as mentioned above, with great slowness and some tremor.

Passive flexion of the right fingers still causes intense pain, in spite of the absolute anæsthesia to all other stimuli.

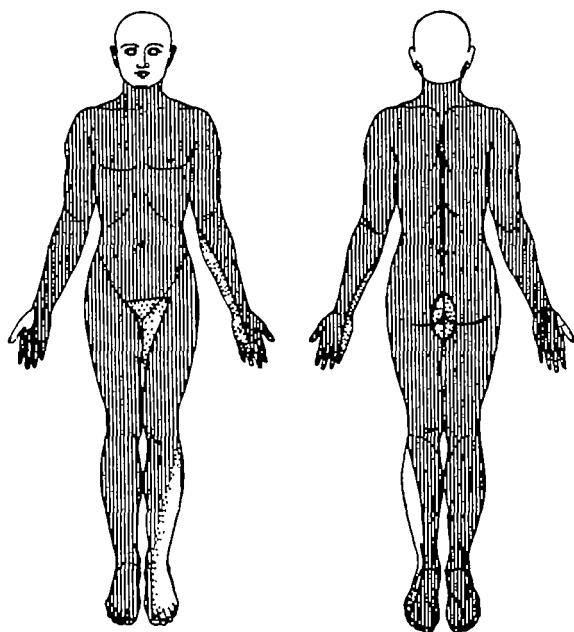


FIG. 5.

(Dr. S. A. K. Wilson.) October 15, 1905. Over dark areas, complete anesthesia to touch, prick, temperature, vibration. Complete loss of muscular sense in same parts. Over dotted areas, a sharp pin-prick is felt as a faint touch.

November 1.—The nurse concealed a hairbrush in the patient's bed, while she was out of the ward. On coming back and getting into bed, she immediately jumped out again and said, "Who put that brush in my bed?" When nurse replied, "I thought you could not feel," the patient said she had seen it, not felt it, though it was hidden so that she could not have seen it.

November 30.—As to the sores on the skin, those on the right hip have healed, and no more have occurred during the last four

or five weeks. Those which began with a vesicle or pimple took rather a long time to heal. One of them was covered with collodion (to prevent any extraneous irritation), but it healed no quicker than the others. But on the right leg sores have reappeared. The bandage which recently has been on the left foot was taken off a few days ago, but the right lower limb has still been kept carefully swathed in lint and bandages, which have been renewed daily, and there has never been any sign that these dressings have been tampered with. In spite of this new sores have appeared within the last few days on the inner side of the right leg, and on the right instep. These are not preceded by a pimple, but are elongated lanceolate areas, about an inch long and a quarter of an inch wide. They look as if the superficial layers of the skin had been scraped off, they usually bleed a little, they heal readily; their edges are usually quite smooth, and do not seem to have been cut or hacked.

December 24, 1905.—During the last three months there has been no marked change. It has been obvious, however, that she has more or less given up hope and endeavour. Once or twice the right arm seems to have improved, but this varies. Sometimes it is possible to flex the rigid fingers with much less pain to her, but sometimes this gives her as much pain as ever. She trails the right leg uselessly behind her, but when it reaches the left foot it is neatly pulled outwards to avoid catching. She denies that she is conscious of this manœuvre. There is some improvement in the power of the left arm.

The loss of sensation remains as on the latest charts. Heat and cold are not recognised on the anæsthetic area. In all other symptoms there is very little change.

She lived about eleven months after leaving the hospital, dying eventually of intercurrent cancer, on November 19, 1906. The nervous symptoms did not alter materially, but in the course of the summer she developed an abdominal tumour. An operation was undertaken for the relief of this, but it was found to be malignant in character and insusceptible of removal.

AUTOPSY.

The *post mortem* was made by Dr. Gordon Holmes and the microscopical examination of the tissues by Dr. Grainger Stewart; and I must express my thanks to them for the trouble taken by them, and for their report, which I append.

The examination had necessarily to be limited to the nervous centres.

In the *brain*, the meninges and superficial vessels were normal, nor was any evidence of disease seen externally on the cerebellum, pons, or bulb. Sections through the brain failed to reveal any definite patches of sclerosis, though there were some doubtful portions which were taken for microscopical examination.

Microscopical examination gave the following results :—

In the *cortex cerebri*, with Weigert-Pal stain, no changes were noted; with Nissl stain, changes were noted in the Betz cells of the motor cortex. These cells showed some chromatolysis, some displacement of the nuclei and pigmentary change. The pigment was arranged usually at one end of the cell, and was of a yellow colour. As a rule, the cell outlines were distinct. There was no evidence of a diminution of the number of Betz cells. Slight pigment changes were also seen in some of the large pyramidal cells, but these were not at all constant. With van Giesen's stain, no changes were noted.

The remaining parts of the brain were examined with Weigert-Pal stain; and (1) in the internal capsule no patch of sclerosis nor pallor was observed; (2) in the mid-brain a small patch was found lying between the pulvinar and the corpus quadrigeminum anterius of the left side; (3) in the pons and in the medulla no patches were seen; nor (4) were there any in the cerebellum. Lastly, the optic tracts showed no pallor, but the peripheral end of one optic nerve which had been removed for examination was unfortunately lost.

The *spinal cord* showed no thickening or other disease of the meninges. The cord was small; and when a finger was drawn lightly along its surface, portions of it felt harder and firmer than the rest. On section through these firm areas, the whole cross section of the cord appeared discoloured. There was no naked-eye evidence of ascending or descending degeneration.

Microscopically, sections stained according to the Weigert-Pal method showed patches of sclerosis, in which the myelinated fibres failed to take the stain scattered throughout the cord. In the cervical, dorsal, lumbar, and sacral regions, these patches involved both the grey and the white matter. In addition to these definite patches of sclerosis, pallor of the ascending and descending tracts could be seen at various places, but of very variable degree. The margins of the sclerosed patches were, as a rule, sharp and distinct, although in some places they tended to shade off into the surrounding tissue.

In detail, the position of these appearances was as follows :—

1 C. Small patch opposite either posterior nerve-root. Pallor

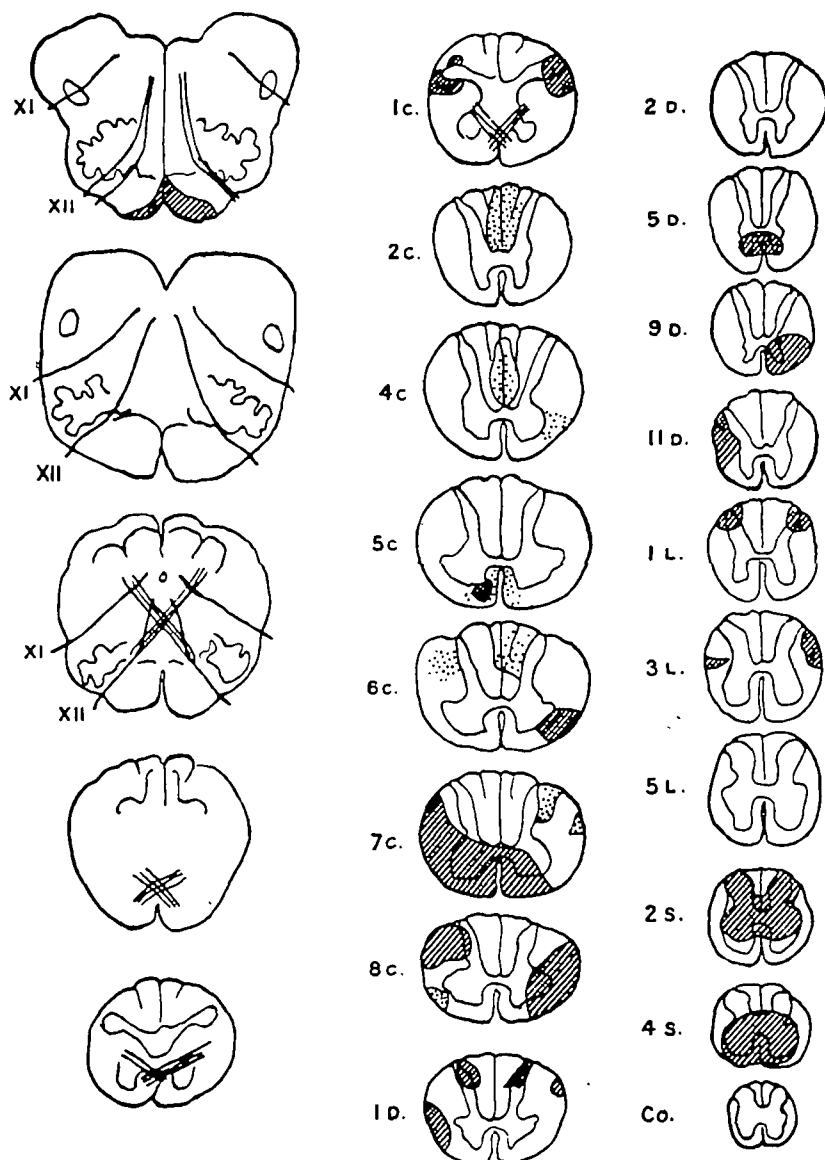


FIG. 6.

To show the distribution of the sclerosed patches in Case 2.

of postero-internal column both sides, and posterior column left side.

2 C. Similar to 1.

3 C. Pallor of posterior medium columns, slight pallor of lateral and of anterior pyramidal tracts.

4 C. Pallor of posterior median columns, of left pyramidal and anterior pyramidal tracts.

5 C. Patch in right anterior pyramidal tract; pallor of lateral and anterior pyramidal tracts.

6 C. Patch extending from external corner of left anterior horn to periphery; slight pallor of both lateral columns and of left postero-external column.

7 C. Large patch involving anterior columns and inner half of left anterior horn, right anterior horn, and anterior two-thirds of right lateral column.

8 C. Large patch involving anterior two-thirds of left lateral column and outer part of left anterior horn; patch at right posterior root; pallor of right lateral column.

There was pallor of the lateral columns throughout the dorsal regions, and, in addition, at the following levels:—

1 D. Patch extending from right anterior horn to periphery; small patch on periphery of left side of cord external to left posterior root.

5 D. Patch lying between anterior horns.

9 D. Patch in left anterior column and left anterior horn.

11 D. Patch involving right lateral column.

1 and 5 L. Pallor of anterior and lateral pyramidal tracts, and of posterior columns on right side.

2 S. Large patch involving posterior parts of anterior horns and the grey matter behind.

4 S. Large patch in same position but involving all the anterior horns.

The condition of the anterior horn cells was examined with Nissl stain, and gave the following results: in the *lumbar* region there were very few normal cells. From many cells the granules had practically disappeared, so that the cell had a homogeneous appearance. In others the granules were collected together at the edges of the cell, while in some they had disappeared, and instead of them there could be seen clumps of yellow pigment occupying part or whole of the cell.

The nuclei were placed as a rule in the centre of the cells, but sometimes eccentrically, and rarely they were hard to distinguish. The nucleoli were as a rule well stained. In the *cervical* and

dorsal regions similar changes were seen, though of a less marked character.

Stained with van Giesen's stain, definite changes were made out only in the sclerosed patches. Here the sections were stained darker, and there was a dense network of neuroglial tissue with very few nuclei. The margins of the patches did not appear so distinct as with Weigert-Pal staining.

The *posterior root ganglia* were examined with Nissl and with van Giesen stains. In certain of the cells some pigmentary change was noted, but this was not general and otherwise nothing abnormal was made out.

Some of the white scars from the *skin* were examined with van Giesen's stain. There appeared to be an increase in the fibrous tissue, but otherwise nothing abnormal was noted.

PLATE I.

To show the position and form assumed by the rash in Case 2 (*cf* p. 349).

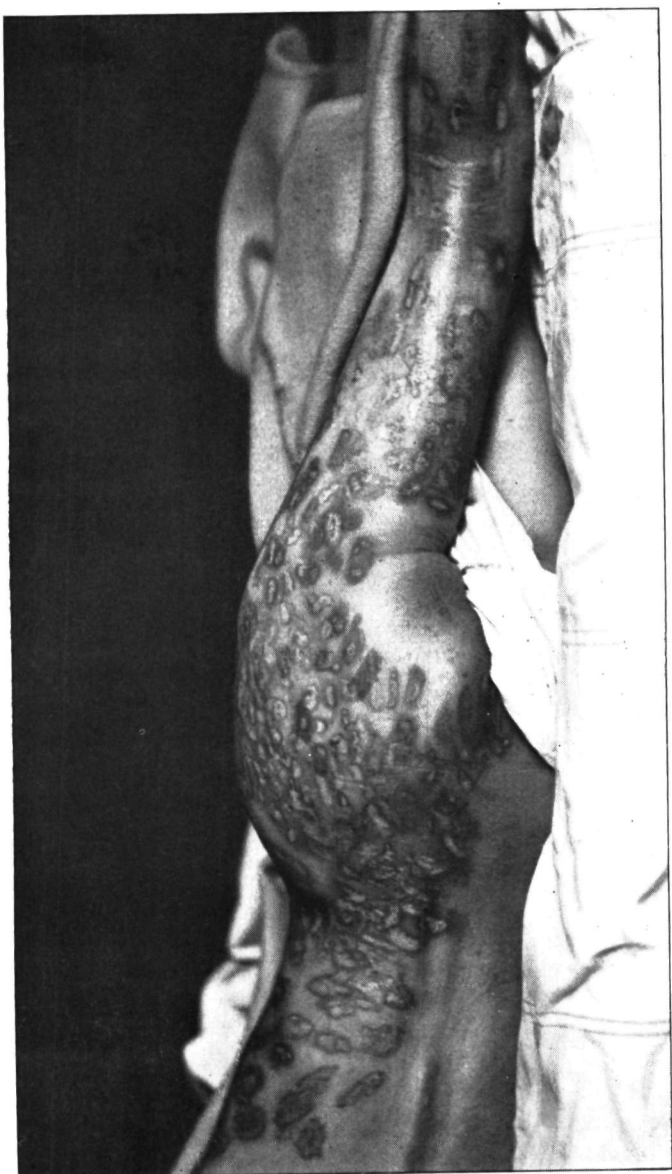


PLATE I.

