IDIOPATHIC LATERAL SCLEROSIS.*

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WITH A DESCRIPTION OF THE MORBID CHANGES OBSERVED IN THE SPINAL CORD IN A CASE OF THIS DISEASE.

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I DESIRE this evening to direct the attention of the Medical Society to a disease which, in this country at any rate, has been but little studied, and has not received that attention which its well-marked symptoms richly deserve. I consider it the more incumbent upon me to make known what I have seen regarding this characteristic disorder, because, so far as I can discover, almost every case which has been admitted into the Manchester Infirmary has fallen under my care. The disease to which I refer has received several more or less distinctive names from those who have investigated its nature and symptoms. I propose to-night to discuss it under the title of "idiopathic lateral sclerosis, or symmetrical sclerosis of the lateral columns". This name was approved of by Erb in his latest writings on this subject, in which he expresses the opinion that the anatomical basis of the group of symptoms here observed consists in a sclerosis of the lateral columns, especially of their posterior divisions, gradually advancing from below upwards. Still, at the time when he wrote, he did not consider himself justified in adopting this name, as his own views and those of Charcot on this subject had not then been confirmed by satisfactory *post mortem* examinations. Erb, therefore, discusses the disease under the title of "spasmodic spinal paralysis"; Charcot having selected the term "tabes dorsal spasmodique". In the remarks I am about to offer, I shall hope to show that one of the cases which has come under my observation goes far to confirm the opinion of these eminent investigators of nervous diseases. This case terminated fatally, and my friend and colleague Dr. Dreschfeld, who has carefully examined the spinal cord, will describe to you the morbid changes which he has observed in its structure. He will, I believe, be able to satisfy you that the characteristic symptoms of the disease are due to a symmetrical lesion of the posterior portions of the two lateral columns.

I would remind my hearers at the outset that idiopathic lateral sclerosis is essentially a primary degeneration. It is altogether distinct from that variety of secondary degeneration which is the result of cerebral disease. It differs also from the secondary sclerosis of the lateral columns, often observed in grave lesions of the spinal cord. Both these forms of secondary spinal degeneration have long been recognised. Indeed, the careful study of the symptoms and morbid changes observed in these two forms of sclerosis, has not a little assisted in unravelling the characteristic features of the idiopathic form of the disease, on which *post mortem* examinations have hitherto shed little light.

During the last eight years, I have had under my care in the infirmary seven cases of idiopathic lateral sclerosis; six of my patients were men, only one a woman. All were in the prime of life, between twenty-five and fifty years of age. I do not at present propose to allude to my earlier cases; for, although I well remember the characteristic gait, I failed to comprehend the full import and significance of the symptoms. I will confine my attention to three cases, all of which have occupied my beds in the infirmary during the last twelve months. Two of these sufferers I propose to introduce to the meeting this evening; they are still under treatment as in-patients.

The first case is that of P. A., aged 46, by occupation a twister. He has always resided in a healthy country district. He has lived a regular and temperate life, never indulged in alcohol or tobacco, and his circumstances have been comfortable. There is no history of syphilis, or of any other grave constitutional disorder. He enjoyed excellent health till about twelve years ago; he then noticed a weakness in his feet, legs, and knees, soon followed by an aching pain in the lumbar region of his spine, which troubled him more especially in going down hill. At such times, he was compelled to lean backwards in order to maintain his balance, otherwise he would have fallen forwards. Soon afterwards, he experienced tremors, and irregular jerky movements in the muscles of his lower extremities. He also noticed that his legs seemed drawn together. He had great difficulty in separating them. He did

not think he could have ridden on horseback without experiencing considerable pain. He continued in much the same state for about eight or nine years, during which he was able to get over the ground with a more or less shuffling gait, without having recourse to the support of a stick. Latterly, however, the weakness in his limbs increased. He found it necessary to use first one stick, and then two; while lately he was compelled to rest the greater portion of his weight on crutches. This patient stated that he had a brother, nine or ten years older than himself, affected with symptoms exactly similar to his own (to the case of the brother I shall again refer). His parents appeared to have been healthy persons. The patient lay on his back in bed. The lower extremities were extended. The feet assumed the position observed in talipes varo-equinus. The great toes were turned upwards, owing to the contraction of the extensor longus pollicis in each foot. The knees were closely drawn together, so that it was difficult to separate them without using considerable force; the adductors of the thighs being rigid and contracted. There were tenderness and stiffness over the spine in the lower lumbar region, and he could not raise himself from a recumbent to a sitting posture without assistance. When his arms were placed at the back of his head, he was able to flex his legs, a movement he could not execute so long as they occupied their normal position. The patellar reflex and ankle-clonus are exaggerated. There is no alteration in either tactile or pathetic sensibility; no retardation of motor conduction; no alteration of the muscular sense. The reflex action of the skin is normal. When he is placed in a sitting posture on the bed, the legs and thighs protruding beyond the side, a smart tap over the lumbar region causes a slight but perceptible contraction of the adductors of the thigh, due to increased reflex action in their tendons. In attempting to walk, he uses two crutches, which he places on the ground considerably in advance of his body. [As soon as he rested the extremity of his foot on the ground, his heel became elevated about three or four inches from the floor. In advancing, the leg in motion was slightly flexed, the knee turned inwards, and the foot drawn in a semicircular manner outwards and forwards. It was then jerked down in front of the stationary limb. As it passed over the ground, the nail of the great toe scraped along the floor.] The functions of the bladder and rectum are unimpaired. The brain and cerebral nerves seem healthy.

The next case I propose to bring before the Society is that of J. H., aged 36. For twenty years he has followed the employment of a spindle-maker, worked in draughty rooms, and indulged freely in beer and in spirits. His circumstances are comfortable. Though married for several years, he has no children. He has never suffered from syphilis or from any other constitutional disease. The body is well nourished, and the man looks healthy and cheerful.

Two years and six months ago, he was camping out at Fleetwood with the Volunteers. The weather at the time being wet and cold, the straw bed on which he slept became soaked with water. The following morning, he experienced stiffness in the calves of his legs, especially on attempting to "double". Though it was wringing wet, he was compelled to occupy his straw bed for three nights. The stiffness continued, and in the course of a few weeks extended to the muscles of his thighs, and of his back, where he experienced severe pain, particularly in the lumbar region of the spine. One day, in attempting to rise from a chair, he found considerable difficulty in doing so. Some twelve months after his camping-out discomforts, he noticed that, in walking, he was unable to raise his feet from the ground; they seemed to hug the floor. His toes especially were liable to be tripped up by any obstacle or inequality of the ground. He often stumbled over small stones, and walked with so unsteady a gait that his companions thought he must be intoxicated. About this time, he first observed trembling and jerking movements in his legs, which he had no power to control. Often, on waking in the morning, he found his legs drawn up, and had great difficulty in straightening them. In the early part of his disease, the right leg suffered most severely; but after a time the left became similarly affected. He likewise felt a sensation of tightness in the abdomen, and severe gnawing pains in his thighs and in his legs, chiefly at night-time. Eight months ago, a feeling of weakness came on in his arms, so that he experienced difficulty in grasping his tools, more especially with his left hand. This weakness in the arms has gone on increasing. His father and one of his sisters are said to have died of phthisis; otherwise the family history is unimportant.

The patient lies in bed on his back, his legs and thighs drawn close together. The centre of the abdomen, from the epigastrium to the symphysis pubis, is somewhat drawn in. This retraction is due to the rigid contraction of the recti muscles, on each side of which the abdominal parietes more or less protrude. There is a distinct groove likewise extending from the anterior superior spinous process of the ilium

* Read before the Medical Society of Manchester.

on one side to that of the other. Several of the tendon-reflexes are exaggerated, notably those of the tendo patellæ and tendo Achillis; while the reflex activity of the tendons of the adductors is readily excited by tapping over the lumbar vertebrae in the region of the spine. Tendon-reflexes also may be elicited in the tibialis anticus and posticus, in the biceps femoris, in the sartorius and the gracilis. The back is stiff, so that he is unable to raise himself in bed and sit upright without assistance. In the recumbent posture, he can flex and extend both legs, though in an awkward and constrained manner. When, however, one leg is held down, he is unable to abduct the other. The measurement of the left thigh is 21 inches; of the right, 19; of the left calf, 12½ inches; of the right, 11½. But there is no atrophy, the enlargement of one limb depending on well-marked changes in the muscles and interstitial connective tissue. They feel lumpy and knotted. There is no anæsthesia or paresthesia. When he is placed in a sitting posture on the side of the bed, and directed to stand, the moment the extremities of his feet rest firmly on the ground, and he attempts to walk, the feet, legs, and thighs tremble spasmodically, until the whole frame is agitated and convulsed. In progression, he places his two arms on the chair, and rests the greater portion of his weight upon it. He cannot plant his heels upon the ground; they are elevated three or four inches above it. At the same time, the foot, in advancing, is moved forward and outwards in a semicircular manner, and the toe drags along the ground.

The third and last case to which I shall allude is that of J. N., a spinner, aged 45. He was admitted into the infirmary on May 5th, 1880, and died on July 7th. This patient was a healthy and intelligent-looking man. He attributed his illness to standing up to his neck in water for the greater portion of the night, during a poaching expedition, in the river Ribble. This occurred two years before his admission. A day or two after his immersion, he observed weakness and numbness in his right leg. Several months elapsed before the left was similarly affected. After a time, the well-marked gait characteristic of spastic paraplegia supervened. There were paresis, muscular tension, contractures, and heightened reflex action; no disturbance of sensibility, no weakness of the bladder or rectum, no trophic changes of the skin, no bed-sores, no ataxy, and no symptoms referable to the brain. In this case, as in the two preceding ones, there was tenderness on percussion over the spine in the lumbar region; and the patient, while in bed, could not raise himself from the recumbent to the sitting posture. He died on July 7th, from thrombosis of the femoral vein. Immediately after death, he was placed in the prone posture, and ice applied along the spine. The *post mortem* examination was held ten hours after death. Dr. Dreschfeld (who has made numerous sections of the cord) has explained in the appendix to this paper the morbid changes he observed in its structure.

Such, then, is the clinical history of these three cases of idiopathic lateral sclerosis. In one of them there seems strong grounds for believing that the disease was hereditary. Thus P. A. asserts that he enjoyed excellent health till about twelve years ago; when, without any apparent exciting cause, his legs and back became weaker. He accounts for his ailment by assuring us that a brother nine or ten years older than himself, became similarly affected at a corresponding period of his life. For the particulars of this brother's case I am indebted to Mr. E. Llewellyn Luckman, who kindly visited the patient at his own home in Eastwood, Yorkshire. He informed Mr. Luckman that his present ailment commenced about twenty years ago with a feeling of numbness in the legs, and gradually increasing difficulty in walking, the legs being drawn inwards. About twelve months later on he was compelled to walk with one stick, and two years afterwards with two sticks, which he still continues to use. As the patient reclined on his couch, Mr. Luckman noticed that his thighs were strongly adducted, and that his feet were inverted; he was unable to separate the thighs. The patellar reflex was considerably increased. There was slight diminution in the strength of his hands. In walking he shuffled along with the characteristic gait of this illness. This patient asserted that a sister, now dead, suffered from the same form of paralysis in the legs, and that another, aged thirty-seven, is similarly affected. Of these cases, however, I have not been able to learn further particulars. In P. A.'s case then, I think we are justified in affirming (though this has been denied by some authorities) that the disease is hereditary. In the two remaining cases, the earliest symptoms seem to have followed exposure to long-continued and extreme cold. Thus J. A. dates the failure of his health to three nights spent on a wet straw bed while camping out with his volunteer corps. J. N. also referred the commencement of his attack to a chill contracted while poaching for salmon in the river Ribble, up to his neck in water. In none of these patients did the affection commence with an acute attack. All spoke of their disorders as having come on gradually and insidiously. Their legs felt

weak. They could not raise their feet from the ground; it seemed as though lead were attached to their soles. In these and similar terms the sufferers severally described their sensations during the early stages of the disease. After a time, manifestations of motor irritation set in. Twitching and jerking of the legs early supervened, especially on dropping off to sleep at night. A feeling of tension also took possession of the muscles, so that voluntary movements were more or less retarded, requiring the expenditure of an unusual amount of force. In one of the cases, the contractions are so marked that knotty lumps can be felt in some of the muscles. After a longer or shorter interval the characteristic walk, termed by Erb the *spastic gait*, supervened. Under these circumstances, the act of progression is strikingly peculiar. The feet being rigidly extended, the patient, in attempting to stand, naturally throws a portion of his weight on the ball of the great toe. No sooner is pressure exerted on this part of the body, and the foot brought into dorsal flexion by pressure upon the sole, than a rhythmic jerking of the foot (the well known ankle-clonus) commences. This jerking is communicated to the whole body, and the patients may be seen resting on their toes, rising up and down as though they were swinging on a spring board. In carrying their feet forward, a peculiar hopping movement is communicated to the frame. In progression the legs adhere closely to one another, and so violent is the contraction of the adductor muscles of the thighs that each leg, after rubbing forcibly against the knee of its fellow, suddenly springs across its path as though prepared to oppose its onward progress. The upper part of the body is bent forward as in paralysis agitans festinans. In this manner, with a swinging hopping movement, and with a vast expenditure of muscular energy, the patients slowly and painfully advance, breathless and agitated by their exertions. If, however, they be directed to retrace their steps, and to walk backwards, they seem utterly unable to make the least retrograde movement. This inability to walk backwards is a feature in the spastic gait to which I have not seen any reference in those authors who have more especially studied this subject.

In idiopathic lateral sclerosis, I have experienced considerable difficulty in testing the effects of electricity; for, whether the induced or continuous current be used, no sooner are the electrodes brought into contact with the cutaneous surface than violent clonic spasms are at once induced. It has further appeared that when the patient is placed in a prone posture, and the so-called spinal epilepsy set up by pressure on the sole of the foot, the agitation of the limbs has continued, even though a strong electric current was passed along the nerves or muscles. Neither the faradaic nor the galvanic current, when applied percutaneously, seemed able to check the clonic spasms which convulsed the limbs. When, however, needles were inserted into the muscles, and electricity passed into the deeper tissues, even a weak current instantly produced a tonic tetanic spasm. This spasm ceased as soon as the electrodes were removed from the needles, and with it the clonic spasm was likewise arrested, and the disorderly movements in the muscles at once checked. In a paper read before this society in the year 1879, and afterwards published in the *Lancet* of the 27th September and 4th October in that year, I expressed the opinion that, in "electric acupuncture we possess a remedy likely to prove valuable in certain forms of abnormal irritability of the spinal cord and medulla oblongata". In lateral sclerosis, we may observe these effects. We set up a tonic spasm which we are able to control; and, on its cessation, we find that the clonic spasms, over which we had before no power, have likewise passed away under the influence of electricity. In these and similar cases, I have, for several years been in the habit of using a mixed current generated by a faradaic and galvanic battery connected together by means of a conducting wire. By this arrangement, a modified interrupted current appears to be induced. In cases in which the cutaneous reflex irritability is increased, this galvano-faradaic current is followed by more favourable results than either the continuous or induced current, when used separately.

But, assuming that the posterior divisions of the lateral columns are the portions of the spinal cord more especially involved in idiopathic lateral sclerosis (as I believe we shall to-night learn to be the case from Dr. Dreschfeld), have recent physiological researches thrown any light upon the functions of these regions of the spine which would assist us in explaining such symptoms as those I have endeavoured to describe in this paper? This question may, I think, be answered in the affirmative. Experiments performed on rabbits by Miescher seem to show that certain afferent influences, in passing up the cord, travel almost exclusively along the lateral columns. Miescher's experiments have been repeated and confirmed by Woroschiloff, who has arrived at the conclusion that both afferent impulses, which originate reflex action, and efferent impulses, which produce movements, pass almost exclusively along the lateral columns, which form (according to these

authorities) the chief channels for the conduction of impulses of all kinds. We can readily believe that, if these channels be invaded by a chronic inflammatory process, such as occurs in sclerosis, the centrifugal nerves will be more or less affected, and paralysis and tension are likely to supervene. We can further understand that a lesion occupying the same region of the spine would prove a source of irritation to the centripetal nerves in their passage upwards. In this manner, the increased reflex movements we observe in percussing over the tendons of certain muscles may be partially explained. But the investigations of Woroschiloff do not rest here. They go to prove that not only do afferent and efferent impulses travel along the lateral columns, but that here also (in part at least) lie the tracts which control reflex movements. If this be true, the sclerotic change may, by destroying these tracts, cut off the inhibitory influence exercised by the brain on the reflex arcs of the spine; releasing these centres from the salutary control to which they are normally subjected, and leaving the muscles a prey to every passing stimulus.

DR. DRESCHFELD'S DESCRIPTION OF THE SPINAL CORD IN
J. N.'S CASE.

The cord, when freshly examined, showed to the naked eye no abnormality except softening in the lowest dorsal region. After hardening in bichromate of ammonium, sections of the cord showed already to the naked eye one lighter coloured patch in each lateral column. This patch was particularly well marked in the dorsal region, and became less distinct in the cervical and lumbar regions. Sections stained in carmine showed the following histological changes. In the cervical region, there was a small patch of sclerosis in the lateral columns; internally, this patch was in close relation to the grey matter; anteriorly, it did not extend up to the anterior grey horns; and externally it did not reach to the external border of the white matter. The grey matter, the ganglion-cells in the anterior and posterior horns, and both anterior and posterior columns, were perfectly intact. In the dorsal region, the lesion was much better expressed. It occupied the same position, but extended a little further anteriorly. In the softened part of the dorsal region, the cord seemed a little out of shape; but the same sclerotic patches were easily recognised. In the lumbar region, the sclerotic portion formed a small triangular patch, close to the outer side of the posterior horns, extending almost up to the outer border of the lateral columns posteriorly, but extending but little anteriorly. As in the cervical, so also in the dorsal and in the lumbar regions, the other parts of the spinal cord (the anterior and posterior horns, and the anterior and posterior columns) were perfectly healthy.

M. Charcot, to whom I sent sections from the different regions, and who had the kindness to examine them, finds the lesions very characteristic. He likewise states that, as far as he knows, this is the first case recorded in which *post mortem* examination has demonstrated lesions confined to the lateral columns of the spinal cord without participation of either the grey matter or the posterior column.

In a second letter, after a further examination of the cord, M. Charcot thus writes: "Je crois en effet votre cas de sclérose latérale *unique* dans l'espèce, pour le moment."

LITHOTOMY IN CHILDREN, AND THE ADVANTAGES
OF A LONG-BEAKED STAFF.*

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I BEG to offer you to-day a few practical suggestions on the manner of performing lateral lithotomy; and to exhibit a special form of staff, which I have used successfully for some years.

From the day of Cheselden to the present time, this operation has been regarded as one of the most successful in the art of surgery; and, during the last fifty years, the details have undergone very little modification, although they are surrounded by many debatable questions. It is performed upon well-established rules, with great success, by surgeons in all parts of the world; yet it is a fact that most practical men have some hobby about the operation. Some expatiate upon the importance of some little manœuvre of their own; while others attribute a great deal of their success to a special knife, or other instrument; and upon their respective advantages they are ever ready to dilate. After all, it is only natural for us to regard with preference the instruments we have often used with success, and to be somewhat conservative when asked to replace them by others of which we have had no experience.

I know that I have the honour to address many to-day who have repeatedly performed the lateral operation; and I feel that I should,

indeed, be a bold man if I asked them to adopt my practice, or even give my long-beaked staff a trial. On the other hand, there may be some present who are about to perform their first lithotomy, and who may be willing to test the value of my experience. I do not pretend to offer any great innovation, but merely to point out that, by the steady observance of a few simple directions, and the use of a special modification of the ordinary staff, lateral lithotomy in children can be made easier, and the common difficulties of young operators avoided.

Now, it is evident that an operator must possess a sound knowledge of the anatomy of the perineum and pelvis; yet this is only a part of the equipment of the skilful lithotomist. The whole anatomy, so to speak, is within the field of mental vision, during the performance of the operation; but the mind is not merely occupied with anatomical details. Moreover, the eye cannot completely direct the hand, as it often does, when the structures to be divided are in a region exposed to view. The surgeon has mainly to rely upon the sensibility of the hand, and the sense of touch. His chief instrument is the left forefinger; and his concern is, first, to reach the staff at the right point; and then, directed by his finger, to cling to it as his only guide into the bladder.

The staff, which I have used with great confidence for some years, is a modified rectangular staff, with a rounded-off angle, a long beak, and the point slightly turned upwards. The lateral groove commences half an inch above the angle, and terminates about the same distance from the point. The main peculiarity is the length of the beak, and this varies regularly with the size of the staff. (Fig. 1.)

I am aware that some of the best lithotomists do not recommend a long beak. Sir Henry Thompson says, in his elaborate treatise on *Practical Lithotomy and Lithotripsy*, page 15: "The curved portion is not to be so long as is usually made, since a small or contracted bladder is sometimes exposed to the danger of perforation when that portion of the staff protrudes far into the cavity." Again: he remarks (page 70), when referring to perforation of a diseased and softened bladder as an occasional cause of death after operation: "I have been frequently struck with the unnecessarily long curve which some staffs have; and have felt, when seeing such an one in use, how dangerous its point must be in a bladder affected in the manner described. I am strongly inclined to think, that, in those cases of rapidly fatal peritonitis, which are sometimes met with after lithotomy, if the search were carefully made, a minute rupture at the upper and back part of the bladder would be sometimes found. It may be very small, lying between the rugæ of the organ, and easily escape observation, yet give issue to a small quantity of urine into the peritoneal cavity; for it is not very easy to explain otherwise some of those attacks which carry off a patient in thirty-six hours or so after operation..... Let me say, injury to such bladders may as readily be inflicted by the forceps as by the staff; but I do not think the knife has anything to do with it." In these words, Sir Henry Thompson clearly points out the possibility of doing a serious injury to a weak and softened bladder with the staff; and that a long curve, under such conditions, increases the risk of perforation; still, I venture to remark that he does not mention a single case in which the accident has undoubtedly occurred; and, therefore, we may fairly conclude that, although such an unfortunate event is sometimes possible, yet, under ordinary circumstances, the danger is very remote. He remarks, moreover, that an injury to a weakened viscus may as readily be inflicted by the forceps as by the staff; and it must always be a very difficult matter to decide in any case the exact part of the proceeding in which the injury took place. It appears to me that the energetic use of the forceps is more likely to result in rupture of a diseased bladder, than the steady pressure of the blunt point of the staff. On the other hand, allow me to add, that I have never seen the slightest injury, or even irritation, follow the use of the long-beaked staff in any case in which I have used it, or have seen it used by my

* Read before the Southern Branch.