Clinical Lectures

ON

CASES CALCULATED TO ILLUSTRATE

THE

DIAGNOSIS & TREATMENT OF PARALYSIS.

By C. B. RADCLIFFE, M.D., F.R.C.P.,

PHYSICIAN TO THE WESTMINSTER HOSPITAL,
AND TO THE NATIONAL HOSPITAL FOR PARALYSIS AND EPILEPSY, ETC.

No. IV.

ON PROGRESSIVE LOCOMOTOR ATAXY.

GENTLEMEN,—The malady to which I am about to direct your attention was discovered seven or eight years ago by M. Duchenne (de Boulogne), and described under the name of *ataxie locomotrice progressive*. In it there is no deficient action of the will upon the muscles individually, and no loss of muscular force or change of muscular structure; but there is deficient action in that involuntary instinctive co-ordinating power by which various groups of muscles are made to work harmoniously together in such acts as standing, walking, or handling. The term ataxy (α , privative, and $\tau\alpha\chi\iota\varsigma$, order), which has reference to the deficiency in proper co-ordinating power, is not very fortunate, for the adjective *ataxic* is very commonly used in a different sense, as in ataxic fever, where what is meant is low fever in which nervous exhaustion is the predominant condition. To meet this difficulty other names have been suggested; but the satisfactory name has yet to be found, and for the present, therefore, we must be content to use the one employed originally, and call the disease in question *ataxie locomotrice progressive*, or *progressive locomotor ataxy*, or *ataxy* simply.

Until M. Duchenne (de Boulogne) drew attention to it, ataxy was confounded with spinal paralysis in general, and especially with that vague form of this paralysis which is known under the name of *tabes dorsalis*. The disorderly movements of the muscles were regarded as symptoms of paralysis. It was not perceived that the muscles in which these disorderly movements were manifested were healthy, and behaved well in all respects except when they were called upon to act in concert. Moreover, it was not known that this loss of co-ordinating muscular power simply was associated with other symptoms in a definite category. In a word, to M. Duchenne (de Boulogne) must undoubtedly be ascribed the honour which belongs to him who discovers a disease, sufficiently definite in its characters, which had been confounded previously with other and very dissimilar diseases.

The case which serves me for a text is now in the wards. It is that of J. C—, a sailor, thirty-four years of age, living at Islington, and admitted into the National Hospital for Paralysis and Epilepsy on the 3rd of April, 1865. The notes I have run thus:—

Present condition.—J. C— is a man somewhat under the average height and weight, well-proportioned, well-developed as to muscle, especially in the upper parts of the body, with scarcely any subcutaneous fat, with abundance of brown hair on the head and face, and with a complexion much dried and browned by long exposure to sun and sea.

He is capable of walking without a stick, but his gait is peculiar—staggering, precipitate, the legs thrown about vaguely and spasmodically, and the heel brought down with force at each step. With his eyes shut, or in the dark, he reels over at once, and would fall helplessly unless prevented from doing so. In the sitting posture he can lift either leg steadily into any position, and keep it there, and when the leg is put out in this manner, he can keep it extended in spite of a very strong effort on my part to flex it. In order to this, however, he must see what he is doing, for if his eyes are shut the leg at once becomes unsteady, and little force comparatively is required to bend it. The right leg is a little weaker than the left, but not to any very considerable degree. He finds it very difficult to come down-stairs, or to quicken his pace much, and he is speedily fatigued by the acts of standing or walking. On being told to shut his eyes, and touch his nose with the fore-

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finger of each hand in turn, he did so with tolerable accuracy, especially with the forefinger of the left hand. On being told to stretch out his arms, and keep them out, he did so quite well, but only so long as he was allowed to see what he was doing, for on holding a book before his eyes, his arms, shoulders, neck and head—the upper part of his body generally—at once become affected with convulsive titubation. When the book was taken away these movements speedily came to an end, but not before they had issued in a fit of crying and sobbing, which was not a little distressing to witness. This fit took the patient quite by surprise, and it could not be accounted for by the examination having been conducted roughly, or carried on for an undue length of time; indeed, the holding of the book before the eyes, which was its immediate cause, did not occupy more than a minute at the most.

The muscles of the lower limbs generally are less developed than those of the upper limbs and trunk, but there is no appearance of wasting in them. Their electro-contraction is perfect: they stand out firm and hard when made to contract by the will, and the contraction seems every whit as strong as it ought to be. Indeed, as has been stated already, when the leg is put out and stiffened, it is not in my power to bend it against the will of the patient, except by the exercise of great force. There is no tremulousness anywhere; and no marked reflex movements are produced by tickling the soles of the feet.

What the patient complains of chiefly are excruciating, stabbing, boring, nipping pangs of pain, flitting from one spot to another in a very erratic manner, in paroxysms lasting from a few minutes to twelve, twenty-four, or forty-eight hours; and generally fixing upon one spot only in any single paroxysm. These pangs are most frequently felt in the two feet, especially about the outer side of the metatarsal bone of the little toe; they are also felt not unfrequently at the back of the thighs, in the nates, and in the upper arm about the lower part of the belly of the biceps. They are scarcely ever absent, especially at night; at night, too, there is often a sensation of great coldness, with some degree of constriction, in the painful parts.

Measured by the compasses, tactile sensibility is found to be much impaired in the feet (especially in the soles), in the lower halves of both legs, and to some degree also in the back of the thighs, in the nates, and in the palms of the hands. The ground is felt very obscurely, but so far as it is felt the sensations are accurate—that is to say, it does not seem to the patient as if there were elastic cushions, pebbles, or other imaginary bodies, or as if there was nothing at all, under the feet.

Very rough pinching is scarcely at all felt in the benumbed parts, but elsewhere the sensibility to painful impressions is keen enough.

In the benumbed parts also, and in a lesser degree in some other parts, muscular sensibility (and with it probably the proper sensibility of the joints and bones) is manifestly diminished. In bed, for example, when there is no light, J. C— says that he cannot clearly tell where his feet are, and that not unfrequently his legs get quite out of bed without his knowledge. There is also very little feeling of pressure when the muscles of the feet and legs are squeezed with considerable force; and in the arms, as well as in the legs, the faculty of measuring and adjusting the amount of muscular action necessary for any given act is evidently not as much alive as it should be. For instance, when two weights are placed in his hand one after the other, one of them being at least a pound heavier than the other, he finds it difficult to say with certainty which is the heavier of the two.

The sight of each eye is defective, and glasses afford no relief. The pupils are equal in size, and respond fairly to the light. The conjunctivæ are injected. There is no arcus senilis. There is no squinting or ptosis.

The hearing of each ear is so dull as to make it necessary to speak in a very loud tone in order to be heard. There are also constant singing and booming noises in the ears—"almost like the wind in the shrouds," he says.

The memory is bad, the spirits are very despondent, the head generally "feels very heavy," and of late (this statement is volunteered by the patient) there has been a frequent temptation to commit suicide.

The pulse is feeble, and about 70 in the minute. The appetite is good. The bowels are somewhat constipated. In any case a long time is spent over a stool: "it wont come out," is the patient's own description of the trouble in this quarter. The urine is also voided slowly and with difficulty, although the urethra is free from stricture; and now and then it escapes in

bed at night. Sexually, the state may be spoken of as approaching to, if not as amounting to, spermatorrhœa.

Previous history.—Five years ago J. C—, it appears, began to suffer from pains in the legs and back, and to be unsteady in his gait; about the same time, also, his sight and hearing began to fail; and from that time to this he has continued to get gradually worse and worse. Four years ago he had a sun-stroke in the West Indies, of which the immediate symptoms were violent agitation and shaking, without loss of consciousness, and for which he was taken into an hospital and bled; but this accident was twelve months after his present malady had commenced, and therefore, it is not possible to look upon it as a cause of this malady.

C— was at sea seventeen years in all, chiefly in hot climates, as the West Indies and the West Coast of Africa; and he continued at sea three years after he had begun to suffer from unsteadiness of gait and from the other symptoms which have just been mentioned. Once during the time he was at sea he had chancres, without secondary symptoms; and repeatedly he had diarrhœa: but, with these exceptions, his health on all occasions appears to have been very good. He says that he was always very careless, often sleeping almost without clothes upon the bare deck or upon the ground; and that he was always "too much given to drink and women." For the last two years the sexual power has been much damped; but before this time, according to his own showing, he appears to have been little better than a very satyr. Two years ago, when obliged to abandon his calling as a sailor, he was for a while treated in the hospital at Quebec for rheumatism. Afterwards he found his way to this country, and became an out-patient first at one hospital and then at another. During this time he appears to have been frequently blistered along the spine, and on one occasion to have been salivated. For the rest, I have only to add that his father died early in life of consumption; that his mother died young from some unknown chronic disease; and that a brother, the only member of the family besides himself, is now dying of the disease which proved fatal to his father.

M. Duchenne, whose description is the best as well as the first, marks out three stages in the course of the malady of which this case is an example. In the first stage, the patient suffers from paralysis, often temporary only, of one or other of the motor nerves of the eye, from some degree of amaurosis, usually accompanied by unequal pupils, and from the peculiar boring erratic pangs of pains from which C— suffered. In the second stage, in addition to the symptoms of the first stage, the characteristic unsteadiness of gait begins to show itself, together with diminished sensibility to touch and pain in the skin of the lower parts of the legs, in the skin of the soles of the feet especially, the interval between the first stage and the second varying in length from a few months to several years. In the third stage the malady becomes more profound and general, the disorder in involuntary co-ordinate movement increasing in the legs, and extending to the upper parts of the body, the anæsthesia increasing in the parts first affected, and extending to other parts, not of the skin only, but even to the muscles, joints, and bones. M. Duchenne does not consider the affections of the bladder, the rectum, and the genital apparatus as essential symptoms of the disease in any one of these three stages: he speaks of them as "epiphénomènes" only.

So far as it goes, the history of C— does not tally with this division of the disease into stages; for in it the failure of sight, the pains, and the irregularity of instinctive co-ordinate movement are seen to have made their appearance together. Nor is this history altogether exceptional in this respect; and therefore the only conclusion to be drawn is that there are some cases, perhaps the majority, in which the symptoms admit of being grouped in the three stages indicated by M. Duchenne.

In the majority of cases, undoubtedly, the principal symptoms of ataxy agree with those which are met with in C—'s case. There is the same irregularity of involuntary co-ordinate movement in muscles perfectly healthy in other respects, the same peculiar pains, and the same anæsthesia. Still there are other cases in which the pains or the anæsthesia are either not very conspicuous phenomena or else absent altogether. Thus, I have met with four cases out of seven, not very advanced in degree it is true, in which there were no pains to speak of, and with two cases (out of the same seven) in which there was no anæsthesia. In the majority of cases, perhaps, the eyesight fails at an early period, and in many cases this failure is associated with some paralysis, often transitory,

of the motor nerves, one or other, of the eye, or of some branches of the motor nerves of the face; but there are other cases in which none of these phenomena are present. In a word, the affections of the bladder, rectum, and genital apparatus are not the only features which must be looked upon as accidental—as "epiphénomènes"; and the simple truth is, that the idea of the disease becomes clearer just in proportion as this fact is clearly realised.

Ataxy, it is said, may be confounded with several diseases, especially with simple loss of muscular sensibility, with disease of the cerebellum, with general paralysis of the insane, with general spinal paralysis and common paraplegia, with saturnine paralysis, with Cruveilhier's disease, with paralysis agitans, and with chorea and some other affections of the kind; but fortunately, as a rule, very little attention will serve to prevent such confusion.

Simple loss of "muscular sense" has been supposed to be the cause of ataxy, and undoubtedly this is a malady frequently associated with ataxy, and most easily confounded with it. In simple loss of muscular sense, however, the sight can supply what is lost; and thus when the eye is open and the attention alive, the involuntary co-ordinate movements as well as the voluntary movements of the affected muscles are all executed regularly.

In disease of the cerebellum there appears, at first sight, to be the same disorder in muscular movements as in ataxy, but this similarity is deceptive. There is now, curiously, in the same ward, and also under my care, a boy in whom there appears to be some congenital defect in the cerebellum—want of due development probably. This boy has a very peculiar gait: he reels and rolls about in walking, as if he were giddy or drunk: there is nothing peculiar in the way in which he plants his feet: and this way of walking is precisely that which appears to be, more or less, characteristic of disease in the cerebellum. In ataxy, on the other hand, the gait, as has been said, is staggering, precipitate, the legs thrown about vaguely and spasmodically, and the heels brought down with force at each step. There is something very peculiar in the way in which the feet are planted: the gait does not give the idea of giddiness; there appears to be some want of balance between the flexors and extensors in each leg, the flexors having the advantage; and, in addition, the muscles, when they act, seem to contract with a sort of jerk—spasmodically. In certain diseases of the cerebellum, also, other symptoms are likely to be present which will assist in the formation of a correct diagnosis, especially violent pains, augmented by movement, in one or other part of the head, and frequent and obstinate vomiting. Moreover, in these diseases the onset is generally sudden, and the progress uncomplicated with the peculiar pains and anæsthesia of ataxy proper.

In general paralysis of the insane, the hesitation in speech, the tremulousness of the lips and tongue, the general tremulousness, the true paralytic weakness of the muscles as to voluntary movement, and the mental condition of the patient, must readily serve to prevent the unsteadiness of gait and other evidences of disordered co-ordinate movement from being confounded with those which occur in ataxy; and in other respects also the features of the two diseases are sufficiently distinct.

In general spinal paralysis and in common paraplegia there is true paralysis, more or less complete, of the muscles as to voluntary power, and the muscles moreover are much damaged as to their nutrition and contractility, and generally as to their sensibility too. In the majority of cases, also, formication is associated with numbness, and, what has never yet been noticed in ataxy, the sensibility to differences of temperature is much impaired or altogether lost in the paralysed parts. Very generally there is tenderness in some part of the spine, and not unfrequently pain in the same region or in the paralysed parts; and if there be pain, it is, as a rule, more like dull rheumatic aching than like the peculiar excruciating, stabbing, boring, nipping pangs of pain, flitting from one spot to another in a very erratic manner, and occurring in paroxysms of varying duration, which are present in C—'s case, and which are rarely absent in cases of the kind. And in those cases where progression is possible the gait is almost always sufficiently characteristic—not staggering, precipitate, the legs thrown about vaguely and spasmodically, and the heel brought down with force at each step, as in C—'s case and in other cases of the kind, but hampered and slow, partly from want of voluntary power over the legs, and partly from the muscles becoming stiff in moving, each leg being brought forward with evident difficulty, even with the help of an upward hitch of the whole side of the body belonging to it, and the part of the

foot first brought in contact with the ground being, as a rule, not the heel, but the toes.

In saturnine paralysis it is the voluntary power over certain muscles which is impaired and gone, and the muscles are atrophied and deprived of electric contractility when the malady has reached its height. Indeed, it is difficult to understand how this affection can be confounded with ataxy.

In Cruveilhier's disease, as I showed in a former lecture (THE LANCET, Jan. 16th, 1865), the wasted muscles are changed in great measure into fat, and, as it were, dissected away, and any errors in movement are such as may be accounted for by this atrophy and absence; whereas in ataxy the muscles are plump and to all appearance perfectly healthy, and the errors of movement are those which refer to want of proper involuntary co-ordinating power. Moreover, in Cruveilhier's disease the pains and anaesthesia of ataxy are not met with.

In chorea there is great want of co-ordinating power in muscular movement, but the rest of the history is quite different from that of ataxy—so different and so well known as to make it altogether unnecessary to recall it to your memory. And so likewise in regard to those other affections which come into the same category as chorea.

And lastly, in paralysis agitans a mistake cannot well be made, for the general features of the disease are more akin to those which are present in general paralysis than to those which are characteristic of ataxy.

In some cases Duchenne's disease has been associated with other diseases of the nervous system, as with Cruveilhier's disease, or with general spinal paralysis or common paraplegia, and in these cases the distinctive characters of the disorder may be somewhat masked; but in ordinary cases there can be but little difficulty in arriving at a correct diagnosis.

The pathology of ataxy is yet in its infancy. The most marked change detected after death as yet is degeneration and disappearance, more or less complete, of the posterior columns and of the posterior roots of the nerves of the spinal cord. Mr. Lockhart Clarke has shown me some beautiful microscopic slides, which furnish most conclusive proof of this fact. But I am not prepared to think that these are the only changes to be met with. On the contrary, I fully believe that corresponding changes will eventually be found in those parts of the cerebellum and cerebrum, wherever they are, which have specially to do with the co-ordination of muscular movements.

The prognosis is full of gloom. The progress from bad to worse may be slow, very slow, but as yet there is too much reason to believe that the term "progressive" in the sense of going on from bad to worse is not misapplied. Still I know of two cases which justify a much more hopeful opinion.

The treatment in C—'s case consists in good food and rest, in hypophosphite of soda and cod-liver oil, and in the electrifying of the region of the cord with free positive electricity, and we shall see in time whether any good will come of it. I am not very sanguine; at the same time I have this to encourage me—that this is the kind of treatment which has done and is doing good in the two cases to which I have just referred.

ON SYPHILIZATION.

By PROFESSOR BOECK, Christiania.

No. I.

BEFORE commencing to employ syphilization I used to think it as great a paradox as any of the readers of THE LANCET do to this day; but I had to yield to what were soon proved to be indisputable facts. I only hope I may be able to give clear evidence of what I have experienced, in order to convince my professional brethren that in this method of cure we possess something more and something better than belonged to any formerly known remedy for a disease that has proved so distressing and so destructive a scourge to mankind.

By syphilization I understand the curative method, by which it is possible, by continued inoculations of syphilitic virus taken from primary ulcers, to bring the body into such a state that it is no longer susceptible of the inoculation of the syphilitic virus.

It is well known, I believe, to the readers of THE LANCET that M. Auzias Turenne is the author of this method, and that he found by making inoculations upon the lower animals that

they gradually acquired immunity from the effect of the syphilitic virus, or became no longer inoculable. M. Sperino and M. Auzias Turenne began about the same time to try inoculation on men suffering from constitutional syphilis, and found that the disease was gradually removed under the continued inoculation.

I shall not enter into particulars concerning the history of syphilization. Like every other new idea it has had innumerable difficulties to overcome. These, however, may be forgotten now that we may hope the subject displayed has been carried so far forward as to be discussed without passionate resistance. The phenomena are certainly of so considerable importance that they deserve to be examined most closely by the physiologist as well as by the physician, even irrespective of the fact that syphilis may be cured by continued inoculations of syphilitic matter. I will therefore, on this occasion, keep exclusively to the physiological side of the question, if I may so express myself.

I have already mentioned that M. Auzias Turenne had observed animals acquiring immunity after continued inoculation of syphilitic virus taken from primary ulcers. The same fact was ascertained by the experiments that M. Sperino and M. Auzias Turenne almost at the same time instituted on men, and the same fact has also been observed by myself in nearly 800 cases in which I have practised syphilization. This fact is so strongly proved that I declare it with the greatest certainty to be infallible; and further, I maintain that the accounts which have appeared now and then, to the effect that immunity was not always obtained, have been founded on careless observations. Immunity occurs gradually, pustules and ulcers diminishing by degrees, until at length none at all result from the inoculation of syphilitic virus; in fact, the virus becomes as innocuous as water.

It is a great and important fact, that the human organism, after a series of inoculations, ceases to be susceptible to an animal virus of such intensity; but it is not without analogy. We know how, after inoculation with variola, the organism acquires immunity from that virus, and how, after inoculation of pleuro-pneumonia, cattle become proof against that disease. One attack of any of the acute exanthemata protects from a second attack. I might quote many more facts that show us how the organism, as a rule, has a certain cyclus to go through; that this cyclus will very rarely be repeated, and that if it be, the symptoms will always be far less developed. It is with respect to the acute diseases that we have hitherto observed this cyclus, for in the chronic diseases it has been less palpable, although syphilis itself certainly gives us an idea of it. Ricord has pronounced this dictum: "Only once constitutional syphilis"—a rule to which there have lately been too frivolously brought forward a great many exceptions.

Immunity from variola is obtained by one single inoculation, whereas immunity from the syphilitic virus is only obtained by a long series of inoculations. The number required differs, and depends partly and chiefly upon the intensity of the virus that is being inoculated; partly also upon the constitution of the patient. As a general rule, the virus from the soft chancre and from the suppurating bubo is far more intense than that from the indurated chancre,* and may be transmitted through many more successive inoculations (always taking the matter for each new inoculation from the last pustules). The first-named virus may be carried on through thirty generations or more, whereas the latter will, perhaps, scarcely go through twenty.

When the first-employed matter will take no longer, that by no means proves that immunity is obtained. New matter will act with certainty, not from being more intense, but simply because it is taken from a new individual. This new matter is, however, capable of transmission through a smaller number of successive inoculations than the first, and when it has ceased to take it will be found that a third and a fourth will still act through a few successive inoculations. In order to obtain the necessary immunity the inoculations must be persevered in as long as any matter will take.

There is another fact with respect to immunity that seems very strange—namely, that immunity is by no means general, although a negative result may have been obtained from the inoculations on the part where they were first begun. I always commence on both sides of the chest. When I get a negative result there, I go on to the arms, and at last to the

* My observations for a number of years, especially those I have made on individuals whom I have treated by syphilization, have convinced me that there does not exist more than one kind of syphilitic virus. Dr. Bidentkap has, by a great many experiments made in the hospital in Christiania, proved the same. These experiments he has described in "Wiener Medicinische Wochenschrift," Nos. 31—34, 1865.