ON FUNCTIONAL ATHETOSIS AND INCOORDINATION OF MOVEMENT.

BY LEWIS SHAPTER, M.D. CANTAB.

Physician to the Devon and Exeter Hospital.

J. P., aged 7 years, thin and delicate, but intelligent in appearance, was admitted into the Devon and Exeter Hospital on the 20th of February, 1879. There appeared to be a general tendency to tonic spasm of the flexor muscles of the left side of the body, as evinced by distortion of the hand and foot, on movement and at rest; an inability to keep the left side of the body in any one position for any length of time; and a strained incoordinate movement of the arm and leg when seeking to attain any special object. When at rest the left arm appeared to be retained awkwardly or with difficulty at the left side, with the fingers of the hand extended, or the palmar surface of the fingers pressed firmly against the hip. On moving the arm for any definite purpose the fingers at once became abducted from the middle line, from probable spasm of the dorsal interossei, and the palm of the hand would be drawn inwards, so as to form an arch with the lower part of the forearm from spasm of the flexors. On being told to touch his nose, e. g. with the forefinger, the fingers at once become abducted from the median line, and tend to close inward on the palm; the wrist becomes rounded; the palm with the lower part of the forearm becomes arched; and the hand, spasmodically moving, is first placed on the chest, with the view of pressing the fingers firmly against the body to prevent their closing: the whole hand is then at length sprawled over the face in a vain endeavour to accomplish the task imposed. With the eyes shut, all these movements become more strained, complex, and difficult. On examining the left foot it is clear that it is similarly affected to the hand, and the normal antagonising force between the flexors and extensors is lost. On standing erect he rests the weight of his body on his right foot, and bending over slightly to that side advances his left leg straight outwards from the hip, the knee being unbent, the os calcis drawn up, and the toes

A Case of Athetosis.

DR. LEWIS SHAPTER.

extended with their tips presented to the floor. The condition simulates talipes equinus, with dislocation of the hip into the thyroid cavity. By a strong effort of the will he can restore the balance in muscular action, and the sole of the foot is thus brought flat upon the ground, but this is only for a time, until the opposing muscles are wearied by their overwork.

Beyond this spastic condition of the flexor muscles there appears to be no disorder indicative of nerve lesion; sensibility to touch, pain, heat, and cold appear normal; there is neither numbness nor pain; galvanism of the flexors and extensors affords no reliable results; but the arm and leg are perhaps somewhat wasted in certain parts, such, e. g., as the calf of the left leg. The abnormal muscular action, it is most important to observe, was continuous rather than jerking, and rapidly recovering or spasmodic. The movements resulting from the tonic muscular spasm, might, indeed, be described as slow in attaining their object, deliberate, and forcible.

The history of the case, according to the account of the mother, was as follows:—When three months old the child was vaccinated on the *left* arm, after which an abscess formed on the outer side of the same arm, which broke and subsequently healed. About this time it was noticed that his hand was constantly moving in a peculiar manner, and when he began to walk he walked on the tips of the toes of the left foot. These symptoms have since increased. The child was always weakly, and the three brothers and three sisters are also delicate; but, as far as can be ascertained, there is no defined disease in any of them.

The treatment consisted in the use of the bromide, of conium, and of the extractum physostigmatis; both of which latter drugs were pushed to the production of physiological effects. On one occasion he was placed under chloroform, with the view of observing how much of the acquired distortion depended upon muscular spasm, and with the further view of getting the foot into such a position as to render a boot adaptable. As results, spasm was temporarily relaxed, deformity removed, and a boot adapted; but this latter, after some days, was obliged to be again removed. Special treatment directed to the existence of spasm was, on the whole, unsuccessful; but under tonics and attention to general nutrition some benefit was derived.

Under the name of Athetosis (*Aθēros*, without fixed position) Dr. Hammond first described an affection which is mainly characterised by an inability to retain the fingers and toes in any position in which they may be placed, and by their continual motion. Dr. Hammond illustrated the disease by two cases, one an intemperate epileptic who after an attack

of delirium tremens, followed by unconsciousness for six weeks, complained of numbness in the whole of the right upper extremity and in the toes of the same side, severe pain in these parts, and complex movements, which were to some extent under the control of the will, especially when this was strongly excited and assisted by the eyesight: coincidently with the development of this condition the memory became impaired, and the intellect manifestly weakened. The second case was also an epileptic, who, after an attack of aphasia, developed numbness on the affected side, incoordination and loss of voluntary muscular motion of the right forearm and leg, loss of antagonising force between the flexors and extensors both of the fingers and toes, and slow moving pains from the hand and foot up to the body. Both cases, therefore, came on with epileptic paroxysms; both were characterised by complex movements of the fingers and toes, with a tendency to distortion; both were also associated with similar head symptoms, tremulousness of the tongue, numbness on the affected side, and pains in the spasmodically affected muscles; but in neither case was there any paralysis. Hammond's view as to the pathology of the disease was purely hypothetical; but he associated the disorder with the intra-cranial ganglia and the upper part of the spinal cord, and named the corpus striatum as one probable seat of the morbid process. Since Dr. Hammond described such a special form of unilateral motor disturbance, under the name of Athetosis, in 1871, other illustrative cases have, from time to time, been recorded by Dr. Gowers, Professor Gairdner, Charcot, Dr. Munro, Dr. Claye Shaw, Dr. Sydney Ringer, and Dr. Clifford Allbutt. Some few of these recorded cases have involved general bilateral irregularity of movement, whilst others have been post-hemiplegic and unilateral; but all appear to have been connected with, or preceded by, other disease of the nervous system, and thereby afford evidence that the pathological change (whatever it might be) causing the disordered movement has been secondary to, or dependent on, a permanent lesion of the higher encephalic centres.

In some few cases pathological research has come to our aid. Charcot found in three cases of post-hemiplegic irregular choreoid spasm occurring on movement, lesions involving the posterior extremity of the optic thalamus, the posterior part of the caudate nucleus, and the most posterior part of the corona radiata; and in these posterior fibres of the corona radiata he assumes there is a special motor function, alteration in which causes choreoid spasm. Dr. Gowers ('Medico-Chir. Transactions,' vol. lix.) reports a case of post-hemiplegic incoordination affecting the arm

only without spontaneous mobile spasm: the brain revealed but one lesion, a cicatricial induration of the optic thalamus extending across its centre beneath the upper surface, but not involving the ascending white fibres from the crus. Dr. Sydney Ringer ('Practitioner,' September 1879) in a case of post-hemiplegic athetosis, found the disease on examination to be limited to the optic thalamus, corpus striatum, and the parts just external. The corpus striatum was the part which suffered most, but the left optic thalamus was also wasted. Dr. Ringer remarks that it is obvious that the cavity in the brain could not be the immediate cause of the incoordinate and involuntary movement of athetosis, for this breach of continuity could only cause paralysis. "The athetosis, therefore, must be attributed to the damaged tissues in the neighbourhood of this cavity, and must, in this case, be due to the damage in the optic thalamus or the corpus striatum or both." It is beyond my present purpose to discuss how far the special phenomena of athetosis are to be dissociated from the commoner characteristics of fixed and mobile spasm so often occurring in partly paralysed limbs, and consequently I may be debarred from considering how far atrophy and degeneration of the basal ganglia may be looked to as the sources which give rise to the phenomena peculiar to post-hemiplegic athetosis. Considering the observations of Crichton-Browne and Meynert on the optic thalamus as a reflex centre, we can with Dr. Gowers readily understand how lesion of this organ alone may account for the disease. My desire rather is to look apart from the basal ganglia for a point from which to ground an explanation of the causation of the phenomena peculiar to athetosis as a functional disorder.

According to the observations of Tcheschechin, the centres of innervation of the movements of locomotion are found at the protuberance, and as high up as the cerebral peduncles, and these centres are in communication with the different encephalic centres that are attached to the protuberance by the middle cerebellar peduncles and cerebral peduncles. Lesions of these centres, or peduncles, occasion a disturbance in the co-ordination of movements, and also unilateral lesions to the peculiar movements of rotation, such as whirling; motion on a pivot, rolling, or somersaults. Although, therefore, we cannot physiologically regard the protuberance as the seat of transition between the spinal cord and the higher perceptive or volitional cerebral centres, we can at least accord to a "centre" which is one of the principal seats of the reception of sensations, a function presiding over the movements of locomotion, and liable therefore as a "centre," especially in the young with growing nerve elements, to receive partial or

permanent damage or alteration of function from injury or even shock, direct or indirect, or reflex.

The case of athetosis under consideration is one unassociated with other nerve lesion either past or present, and presumably it is dependent on atrophic changes in nerve elements of spinal centres, and primarily due to shock indirect or reflex.

The clinical features characteristic of the disease are briefly (1) tonic, slow, more or less designed, and perfected muscular spasm, especially of the flexors; (2) motor incoordination; (3) distortion, the result of unopposed muscular contraction; (4) a want of controlling volitional power, whether such want is really dependent upon a loss of power, or only upon a local ataxia of the agent to be controlled.

Now the point naturally at issue to every clinical investigator is the cause of the incoordinate movement; or, in other words, is this incoordination a result of an affection of the co-ordinating spinal centre, or is it merely a mechanical result of flexor spasm which would necessarily give rise to an awkwardness and irregularity of movement?

In locomotor ataxy we have incoordination of movement as a pathognomonic feature, and although this incoordination is, in the majority of cases, intimately connected with spinal lesion, preventing conductivity of motor force, it is not necessarily dependent upon it. As Trousseau observes, it would be wrong to regard the disease as dependent on such conditions as the palpable lesions found, since they are only a consequence and an effect. Incoordination of movement, indeed, when it exists as a purely functional disorder in a class of cases free from any other implication of nervous function, speaks against the *necessary* dependency of co-ordination on the evolution of force from nerve centres. An example of this class of case I ventured to publish, in 1872 ('Med. Times and Gazette'), and I then drew attention to the fact that incoordination, as occurring without the supposed necessary lesion involved in posterior spinal sclerosis, could not be regarded as necessarily dependent upon it; but that we had rather to view, with Dr. Lockhart Clarke, a disorder of that physiological state (*viz.* tonicity) of the muscles, which, as dependent on reflex action, maintains the constant state of partial contraction, and keeps the antagonistic muscles in equilibrium or static tension. Incoordination of movement may then, clinically and physiologically, be a local disorder in the antagonising muscles themselves, and is the result of a loss of the physiological state of tonicity. In posterior spinal sclerosis, again, there is found a sufficient lesion, not only to interrupt the ordinary cycle of events and to disturb the necessary reflex act upon which co-ordination is dependent;

but also to produce an irritation to react upon the cord as a centre, and so occasion an evolution of motor force upon which harmony of action must be directly due. Incoordination then being, from the nature of the case, a disorder of motor force, and the cord being a generator as well as a transmitter of nervous force, we find in posterior spinal sclerosis motor action, irregularly distributed through the proper nerve channels from a nerve centre, and connected with a morbid condition of the sensory system, or that part which goes to complete the cycle necessary for the production of a reflex act. In athetosis we have only to go a step further, for we have to deal with unilateral incoordination of movement, presumably the result of disorder of a special incoordinating centre, encephalic in location. It follows also that in all cases of post-hemiplegic athetosis there must necessarily be a continuity of function between the motor path and the grey matter of the centres, and it must be assumed that the lesion causing the associative hemiplegia is only partial as regards the grey matter, and consequently permits of that "central irradiation of voluntary impulse" insisted upon by Nothnagel.

Finally, we are asked by many investigators to see a connection between athetosis and chorea, paralysis agitans, writer's cramp, spastic paraplegia, &c. Hemichorea is thought to bear the closest analogy, but if the pathognomonic feature of athetosis is incoordination of movement, and the pathological element disorder of the co-ordinating centre at the protuberance, the disease is sufficiently well-defined to render comparisons of minor clinical details dangerous and inconvenient. Chorea has its own clinical landmarks. In the so-called "insanity of the muscles," in the morbid restlessness, degree of variety, and combination of the involuntary movements; in the rapidly recurring and alternating flexion and extension, pronation and supination, abduction and adduction, occurring in the upper and lower extremities; and finally, in the "dreams of movement" which may occur during sleep, we have surely clinical characteristics that need not be confused or even dwelt upon. In athetosis we find little or nothing in common with chorea; the incoordination of movement, the flexor spasm leading to distortion, and the slow tonic designed character of the spasm, are phenomena distinctive and pathognomonic. It has been with the hope of rendering these signs more generally accepted, that I have thought a case of "functional" athetosis, uncomplicated with any other nerve disorder, worthy of consideration.