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A Unique Case of Bi-Lateral Athetosis.

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IN 1871, for the first time in the history of neurological medicine, the term Athetosis appeared in the literature. It was employed by Hammond in the first edition of his classical "Treatise on Diseases of the Nervous System," to designate a peculiar form of movement and spasm of the voluntary muscles.

Charcot and others have disputed the claim made by Hammond, but confirmatory cases have been recorded by Allbutt, Gairdner, Eulenberg, Ringer, Ritchie, Shaw, Landouzy, Grasset, Oulment, Bernhardt and Brousse.

In athetosis the muscular movements are similar to, but not identical with, those of chorea, being involuntary but more rythmical and decidedly more deliberate and continuous, and without the characteristic lightning-like suddenness of accession, peculiar to chorea, while they do not cease during sleep, and the position of the fingers and toes in the spastic state often being such as they could not normally be voluntarily placed in by the individual, and by an inability to retain them long in one position without external aid. The facial muscles are usually not implicated, though they were somewhat in the present case, unless it be by intercurrent paralytic distortion, and the muscles involved are strong and hard and large, sometimes markedly hypertrophied in contrast with the unaffected portions of the body, as appears in the accurate drawing of the boy in the accompanying illustration, made true to life, by Mr. Lowry, a skillful and well-known portrait painter and landscape artist, of St. Louis.

In the diagram the boy's arms are placed in the best position to bring out the characteristic athetosic spasm and temporarily arrest the rhythmical movement, a position of complete and extreme extension, and this posture of the muscles involved, I have found to be the best diagnostic attitude.

The boy could not maintain the position more than a few seconds at a time, or any other in which the upper limbs were in a fixed position, so that the artist had to have a number of sittings in order to secure the outline drawings, two of which are here presented.

This boy had not complete voluntary control over the movements of his muscles; that is, he could not by direct effort of the will, along the regular channels of nerve conduction, restrain either the rhythmical movements or the spasmodic attitude assumed by the fingers; but he could, by strategy, modify both attitude and movement, by bringing one limb to bear upon another and assuming for the affected limbs, flex positions; but no matter how much he succeeded in momentarily managing these movements, grotesque attitudes would always be assumed by one or more of the fingers. His affliction unfits him for any occupation requiring manual dexterity. He has tried a number of things, but had to give them up because of physical incompetency.

He could put his limbs as a whole in any attitude. For example,—the arms akimbo, the hands on top of head, behind back, straight out or down at side when requested to do so, and swing arms to and fro and rotate them.

In attempting to write (which he could not succeed in doing any better than an ataxic) he would double himself down into a peculiar position, grasping his left hand entirely over the thumb and all the fingers of the right, and after ten minutes of effort, the chirographic results were mostly an unintelligible, tremulous, irregular scrawl, not nearly as good as the chirography of diffused cerebral sclerosis, only one or two efforts being approximating intelligible to one who knew how his name ought

to be written, and in these approximatory successful attempts at a signature the small letters were made four or five times the usual size in height and diameter.

This young man has no history of epilepsy as it existed in most of Hammond's cases; never had convulsions nor chorea in infancy or childhood, nor is there any evidence or history of genuine hemiplegia. The case appears to me to be in every way a distinctive symptomatic one, even more fully sustaining Hammond's claim for this disease as a distinct pathological entity, than this author's own recorded cases.

This case has contributed more than any other that has come under my observation to dissipate all skepticism as to its claim to separate nosological and pathological consideration. M. Charcot and others who have confounded athetosis with post-hemiplegic chorea will not confound this case in like manner, as its antecedent history will not justify such confusion.

I am now satisfied that the cases hitherto reported by me under the designation of persistent rythmical clonic spasm when I was unwilling to accept, with the full meaning of its author, the term athetosis, as descriptive of a distinctive neuropathological grouping, were cases of true athetosis. (*Vide* ALIENIST AND NEUROLOGIST, Vol. II., p. 662, 1881.) These cases were neither post-epileptic or post-hemiplegic, the movements were deliberate and the irregular contractions were characteristic, though not so marked as in the present case. They were designated and regarded as athetosic at the time, but not as athetosis, pure and simple.

In the literature on this subject as contributed and collected by Hammond in the last edition of his work, there are now enough correctly recorded cases to satisfy the most skeptical as to the right of the term athetosis, I think, to be retained in the distinctive nomenclature of diseases, and the line is there clearly drawn, between the real and its resembling symptomatic states, and to this author the reader is therefore referred for a fuller discussion

of this subject, which it were manifestly unnecessary to here prolong.

Following is briefly the record of the history of this case, as entered in my case-book, April 19, 1887, the patient having at that time been several weeks under observation, though an active attempt at treatment was not begun before the date of beginning of the record.

CASE.—Geo. E. M., age twenty years, American born, about nine years ago, met with an accident on the Cairo and St. Louis Narrow Gauge Railroad (now the Mobile and Ohio) causing injury by concussion and direct violence. The train on which he was sleeping at the time of the accident broke through a bridge and was thrown off the track. He was taken from the wreck in an unconscious state, but remained so for a few minutes only. He had two ribs broken and was hurt in the lower dorsal region, but not seriously enough to affect function of bladder, kidneys or bowels, or power of moving lower limbs. He was able to go out within six weeks after the accident, but had an incomplete brachial menoplegia.

About one year after the accident contracture of the left forearm on the breast appeared and embarrassed movement of the right arm set in, so that he could not throw a stone easily or project it far from him. He "threw like a woman," he says, and could not make a full rotary movement.

About four years ago, long after he had fully recovered the general use of the upper extremities, irregular contractions first appeared in fingers and flexors and extensors of arms and forearms. About one year ago the left arm grew worse and the right arm became useless for three months, and had to be carried in a sling.

He can hold nothing requiring strong coördinate movement in left hand. Has dropped things from both hands, and cannot cut meat with left hand, nor hold a fork.

He can cut with the right hand by clinching the knife

in the hand as a dagger is held. Can put his left hand in a side pocket of coat but cannot easily get it out without assistance from the other, on account of the irregular spasmodic position of the fingers. Cannot trim finger-nails of the left hand without great difficulty, because of the irregular spasmodic attitude assumed by the fingers.

Was tongue-tied when an infant, but could never talk plain though frænum was cut by Dr. Boothe, of Sparta, Ill. About two years ago speech was much embarrassed. He cannot now say what he wants to fluently.

He cannot whistle. He has some slight rhythmical spasmodic movements of the obicularis oris, and levator and depressor anguli oris muscles, and some apparent remains of former slight paralysis of right side of face, apparently in his expression.

Movements of tongue are only restricted by cicatrized and contracted frænum linguæ.

Can handle pen and pencil, and makes out to do a sort of writing, by holding right fingers and thumb with left hand and steadying right arm against chest. Eye balls move normally.

He was a bright boy before the accident, but became feeble-minded after the concussion of the accident, and was with Dr. Wilbur, at the Illinois State Institution for the Feeble-minded for several years, though his mother thinks was never dull enough in mind to have been sent there.

His intellect is now quite good and his intelligence up to the common average, even above it probably, considering how much and how long he has been afflicted and how little education has been given him.

Has a markedly left splayfoot resembling talipes, from injury to instep and displacement of tibia on astragalus, caused by fracture of external malleolus, though both feet are turned out abnormally.

Tendon reflex normal; both knees. No anæsthesia or hyperæsthesia anywhere.

Can button and unbutton shoes and dress and undress himself, though clumsily and tediously.

Has rhythmical movements of toes. Never had chorea or convulsions.

This case is unique in that, it is double or bi-lateral, not associated with epilepsia, and resulted from cerebral concussion and shock.

It is unique in the fact, that though feeble-mindedness was associated with it from the common cause, that has disappeared. It is unique in having some facial movements.

When he is sitting with hands on lap, the left hand will involuntarily assume the position of a pointing hand, the index finger standing straight out and stiff, while the rhythmical movement in the other fingers and the thumb continue.

It is unique from the further fact that it appears, after nine years, to be recovering, the spastic conditions having now almost disappeared, while only the deliberate contraction and extension remains, and this symptom is certainly much better.

The case is still under treatment, and should a recovery or death occur while under our care, it will be made the subject of a further communication.

This case tends further to establish what Hammond has stated, and Oulmont and Brousse's cases confirm, that the disease is not necessarily confined to one side.

It shows that athetosis is not a necessary choreac sequence of hemiplegia, and that it certainly is not a post-hemiplegic chorea. It shows what other cases have shown, that a cause sufficient to produce paralysis may cause it, while, at the same time, it establishes the fact that like cases previously reported, it may come on after the paralysis is gone.

It shows, also, how tardy a sequence it is to the original injury, and tends to confirm the conjecture that it is due to chronic degenerative change, rather than any sudden pressure, such as results from hemiplegic clot, emboli, or

thrombi, though the improvement in this case might be thought to militate against the conjecture of chronic degenerative atrophy as was found by Ewart in one of Ringer's cases.

The original causative injury in this, as in preceding cases of record, was in the head.

It is not probable that in this patient any degree of softening about the corpus striatum, as was found in Landouzy's and Sturges' cases, exists. None of the other symptoms justify it, unless we assume the establishment of separation of cerebral tissue or vicarious neural function, neither of which however are organic impossibilities.

My own opinion, for what little it may be worth, is that degenerative tissue changes in the motor area of the cortex and subjacent striate bodies have set in, as the chronic sequence of the railway concussion.

This patient is and has been from the beginning of treatment on iodide and bromide of potassium, hypophosphites compound, arsenic, strychnia, and when malarial symptoms have intervened, courses of quinine. The bowels have been kept open regularly with rhubarb.

The accompanying cuts are illustrations of two attitudes taken at different times in the progress of the case, by the artist. They make the boy appear rather more fleshy in the chest and stouter in the legs and thighs than he actually is.