

VAN DER POEL (S. O.)

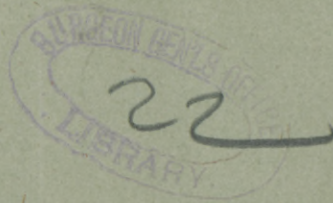
from the author.

PROGRESSIVE LOCOMOTOR ATAXY.

BY

S. O. VANDERPOEL, M. D.,

ALBANY.



Reprinted from the Transactions of the New York State Medical Society, 1865.

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In October of last year, Mr. V—, a gentleman engaged in business in the city of New York, entered my office; his step was jerking and irregular, and his countenance indicated prolonged suffering. He came, he stated, to seek relief from severe pain which he was suffering at irregular intervals, principally confined to the lower extremities; though at times encircling the lower part of the abdomen as a constricted band; also, that the lower extremities were partially paralyzed, as evinced by an almost total loss of sensation, and irregularity of motion of those parts.

As my acquaintance with him was limited, inquiry was made as to his previous history, and the opinion expressed of his case by his physician in New York—a gentleman of some eminence in the profession. His replies, as to the early period of his affection, are condensed as follows: The first indication of any trouble in the feet was about two years since, when he found himself daily taking a seat in an omnibus or car, instead of the usual walk to business, but thought little of it at the time; soon he could not sit comfortably with the feet resting on the ground, having an inclination to raise them; this feeling continued until he always sat with the feet on a chair before him, and the knees raised. About this time he began to feel a heaviness in the soles of the feet, and a want of sensation, which was followed by a prickling, tingling feeling. In walking he invariably stopped at a street crossing, and hesitated which foot to use in stepping off or on the curb; and if a hydrant or lamp-post were near, always put out his hand to assist himself. From that time he always used a heavy cane. The heavy numb feeling now extended to the legs, and soon after entering the house he found the sofa or bed the most convenient place. He experienced trouble in washing, as when leaning over, and happening for the moment to close the eyes, found himself staggering backward and forward, not being able to keep his balance. He also had trouble in going about in the dark, and suffered from pain in the legs and joints, always wanting to see his feet when he had occasion to use them. The opinion expressed by his physician in New York, he stated, was, that it was considered the commencement of paralysis, which must be gradually progressive. Recognizing that the tableau of symptoms presented, in no way answered to this diagnosis, I instituted a closer scrutiny. The intellect showed no impairment, nor were there any indications which induced me to suppose this condition imminent, as would be were it progressive paralysis. His sleep was sometimes broken from sudden and sometimes prolonged pains in different parts of the body, chiefly, however, in the lower extremities. Vision had not been affected, either from temporary paralysis of the muscles of the eye or of the retina itself. The other bodily functions were normal or nearly so.

In order to test whether the supposed paralysis were truly such, or an anæsthesia of the parts merely, I requested him to be seated, and taking hold of the foot directed him to resist flexion and extension of the limb. There was no diminution of the muscular force. I then bade him stand upright, with the feet placed closely together, and close the eyes; the instant he did so the equilibrium was lost, and he would fall unless the eyes were promptly re-opened.

I told him I considered his malady "Progressive locomotor ataxy," or more definitely a lack of muscular co-ordination.

While I gave him a change in the signification of his malady, it was not accompanied with any more encouraging views as to the result of any treatment thus far suggested for its cure. Pathologists placing it in the family of neuroses such as chorea and epilepsy, have administered the treatment recognized as proper in those affections. Prominent among the remedies for which, has been the prolonged administration of nitrate of silver. Little encouragement having thus far followed these efforts, I prescribed a scruple of bromide of potassium in two drachms of comp. tinct. bark, three times daily. Its prolonged use was favorable in alleviating to a great degree the migratory pains, and restoring somewhat the co-ordination of the muscles.

A severe attack of pneumonia which has awakened the softening of latent tubercles has suspended all treatment adapted to the ataxy proper.

Remarks.—Progressive locomotor ataxy, though by no means a new malady, has been but recently described as a distinct and recognizable affection. True, many of its symptoms have been often noticed, under the denomination of tabes dorsalis, but with this were included many other conditions and diseases of the spinal cord. Dr. Todd, in the encyclopedia of anatomy and physiology (article "nervous system,") refers to the *two* varieties of paralysis of the lower extremities; the one consisting simply in the impairment or loss of voluntary motion; the other distinguished by a diminution or total absence of the power of co-ordinating movements, while considerable muscular power remained; the patient experiencing great difficulty in walking, and his gait being so tottering and uncertain, that his center of gravity was easily displaced. Though this description is faithful as far as it goes, he still fails to recognize the true character of the malady, classing it as a paralysis, and ignorant of its history and distinctive progress.

The credit is due Dr. Duchénne of Boulogne, in 1858, of first calling the attention of the profession to it as a distinct malady. As Bright in acute desquamative nephritis, so Duchénne in this malady, has presented so full and accurate a picture, that later descriptions merely portray the fuller shadings. And as in the former, the malady is even yet known chiefly as "Bright's disease," so enthusiastic admirers have applied to the latter the name "Maladie du Duchénne."

Dr. Duchenne defines it "a malady essentially chronic, characterized specially by progressive abolition of the co-ordination of voluntary movements, simulating a paralysis with which it contrasts by the integrity of the *muscular force*."

From the limited information which must still exist of this interesting

malady, it has seemed proper to give as brief a resume as possible of some of its most prominent features, at the same time that it may easily be recognizable.

The *pains* are peculiar, as they are also one of the most constant precursors. They are usually intense, coming and going with lightning rapidity; again they may be more prolonged, lasting half a minute, and repeating themselves ten or twenty times an hour.

They return by accessions several times a year, without any appreciable cause, or again without any save variations in temperature. Authors, under the titles of "General Neuralgias" or "Neuralgic Rheumatism," describe pains which are but the precursors of Progressive Locomotor Ataxy.

Among nervous accidents, observed about this period, are nocturnal incontinence of urine, and especially spermatorrhea—indeed half the cases thus far noted have been subject since puberty to spermatorrhea. Strange as it may appear, the opposite condition, which is sometimes observed to precede the progressive paralysis of the insane, also occurs, viz: The singular faculty of being able to repeat the sexual act, a great number of times within a short period. Yet cases occur where its repetition eight and ten times in the twenty-four hours is noted; probably this false appearance of exaggerated virility is a morbid state which is allied to the medullary excitation, the cause of the involuntary seminal emissions, and incontinence of urine.

In this first stage of the malady, and before the want of co-ordination in the muscles is apparent, there generally occur paralysees; some transitory in character, others more persistent. Among the first is *facial* paralysis—or again a *hemiplegia*, which comes on without precursory symptom, without any trouble of the intelligence, or even momentary loss of consciousness, and yields in a few days, which excludes all idea of cerebral hemorrhage, or cerebral softening. Also paralysis of the tongue, which often disappears as rapidly as it came.

Paralysees more persistent, are also precursors. The more common are those which affect vision. Thus a paralysis of the nerve of the external motor oculi, induces convergent strabismus—or again the nerve of the common motor oculi, inducing divergent strabismus, and falling of the eye-lid. The duration of these forms of paralysis is very variable, with some remaining through life, others only a few days, weeks or months. Various other forms of local paralysis are noted, but not of sufficient frequency to call for notice in this paper.

Another train of precursory symptoms, is a sensation of constriction in different parts of the body. The individual complains that his chest is constricted as by an India-rubber band; sometimes their arms and legs, seem bound. They think their clothes too tight; or a sensation as a girdle bound about the abdomen.

Soon after the foregoing symptoms, those pathognomic of the affection present themselves. This defect of co-ordination in the voluntary movements, is little marked in the beginning, but augments progressively as the malady increases; at first almost exclusively confined to the inferior members—the superior are implicated only in the last stage of the ataxy.

In the lower extremities, the want of co-ordination in the voluntary

movements, is shown at first by light oscillations of the body, of which the individual is scarcely conscious. We can establish however that these oscillations are due to the fact that certain muscles of the lower extremities contract strongly or quickly in rapid succession. As the malady becomes more confirmed, the defect in co-ordination is shown by the difficulty shown to preserve an equilibrium when standing, or walking. This difficulty may extend to an entire impossibility to do either, unless the sense of sight comes to aid the disordered movements.

When those affected rise to walk, especially if they have been for some time seated, they do so with difficulty and a sensation of stiffness, accompanied by a pain more or less severe in the limbs, extending to the lumbar region. In order to rise they are obliged to lean upon a cane, or upon the arm of a chair. Once standing, they make great efforts to maintain themselves; they totter, and perceiving the muscles do not obey the will, they fear falling, and if they have nothing to sustain themselves, their arms fly instinctively from the body, a little forward, oscillating much as a tight rope walker without his balance pole. This want of equilibrium, more marked in the first steps, is just as apparent when the individual desires to change the direction of his course, or especially to return on his footsteps. It is very sensibly diminished if he uses a cane, or leans upon the arm of another.

Once started upon a course, their step, at first slow, becomes involuntarily accelerated; it is uncertain, hesitating, and as one intoxicated. Instead of the ordinary measured flexion of the knee, it is jerked, with a forced extension. The feet thrown out right and left, are pushed forward as if sent by a spring, and fall back, the *heel* first striking. In noticing this difficult and uncertain walk, it would seem the individual would not progress far without exhaustion—yet upon level ground they may walk miles, and fatigue walkers with whom there is no nervous affection. In later stages, exercise soon exhausts the forces, and after a short distance the individual is out of breath, and bathed in perspiration.

The ataxic difficulty rarely extends to the upper extremities; when it does it is usually at a late period of the malady, and develops gradually as in the lower. The aid of vision is even more necessary to guide the irregular action of the muscles than in the lower extremities.

Spasmodic contraction of the muscles, at first scarcely noticeable, increases in severity with the progress of the disease. They appear not only when the will commands a regular movement, but also in a state of repose. These muscular spasms in the latter case consist in energetic shocks of the affected limbs, and have a great symptomatic value. The sick persons say that in walking, and even when standing quietly, the ground seems suddenly to sink under them. When lying we see their limbs tremble, sometimes violently. If the thigh is seized the muscles tremble under the restraint, while the foot moves with remarkable celerity, even against the will of the individual.

As the disease arrives at its extreme period, the anæsthesia becomes more and more pronounced, and more generalized in different parts of the body. The feet scarcely recognize the ground on which they travel, and when to the cutaneous insensibility is added the muscular osseous and

articular anæsthesia, the individual has no consciousness of standing, or of the resistance of the ground upon which he may stand with contracted muscles—closing the eyes, he feels as if suspended in mid-air. This anæsthesia may even extend to the mucous membranes, losing taste and smell, or the sensation of heat and cold.

And yet this anæsthesia, marked as it is, and present in the majority of cases, can only be regarded as an epi-phenomenon of the malady. It can not be regarded as essential to the disease, for many cases have been noticed where it was wholly absent.

In the latter stages of the malady the temporary paralysis affecting the different muscles of the eye usually returns and becomes permanent.

Despite the grave symptoms thus portrayed, individuals affected with progressive locomotor ataxy preserve the integrity of their mental faculties. Their digestive functions are also regularly accomplished; the assimilative functions suffer little. Finally, however, they become somewhat emaciated, and, as with paraplegics, bed-sores form where parts are submitted to continual pressure, on account of this pressure, and the modification which the troubles of innervation cause to the parts.

Diagnosis.—Locomotor ataxy, as will be seen by the previous narrative, is not difficult to diagnose when fully established. Still there are certain symptoms which have a peculiar significance in the early stages. Thus the transitory paralysis of the apparatus of vision, and the singular character of the pains, are phenomena of the highest signification. These phenomena, taken separately, have a certain nature; taken together they are almost pathognomic. Thus, when an individual complains of experiencing at intervals lightning flashes of pain, erratic, manifesting themselves sometimes in one part and sometimes in another; when also with this individual exists a paralysis of the third or sixth pair of cranial nerves, inducing a convergent or divergent strabismus, complicated with amblyopiæ or complete amaurosis; when especially these accidents appearing without appreciable cause, have yielded of themselves, after a longer or shorter time, resisting in the mean time any and all treatment, we have a right to suspect that progressive locomotor ataxy is commencing.

Disease of the cerebellum, arising from some congenital defect, is accompanied by disorder of the muscular movement; but in this disease there is a peculiar gait; they rest and roll in walking as if giddy or drunk; there is nothing peculiar in the way in which the feet are planted. In ataxy, on the other hand, the gait is staggering and precipitate; the legs thrown about vaguely and spasmodically, and the heels brought down with force at each step.

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.

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 too are damaged as to their nutrition and contractility, and generally as to their sensibility. In the majority of cases formication is associated with numbness, and what has yet never been noticed with ataxy, the sensibility to difference 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 rarely absent in ataxy. And in those cases where progression is possible, the gait is almost always sufficiently characteristic, not as has been shown to occur in ataxy 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*.

Chorea, considered in relation to the troubles in movements, which accompany it, is of all convulsive affections, that which approaches nearest to progressive locomotor ataxy. For when one affected with chorea wishes to execute determined movements, the perturbation of the locomotor functions are most energetically manifested; the will, still sufficiently powerful to command muscular action, cannot control it, once that the impulse is given. The difference however is readily apparent. While the individual affected with progressive locomotor ataxy, no matter at what stage of the malady, can guard as long as he wishes absolute repose; this repose is never as complete in chorea. The troubles in muscular action are shown by convulsive movement, more or less limited to certain parts of the body, more or less violent, and which really never cease save during sleep. It is these spasmodic contractions of the muscles of the lower extremities, which renders standing upright difficult—phenomena which take place independent of the will, and even in despite of it.

The duration of locomotor ataxy is usually long; the mean being from four to five years—it may be extended to ten, fifteen or twenty years.

However long may be its duration the prognosis is none the less unfavorable; the fatal termination is most frequently hurried by inter-current affections, when the perturbation of the nervous system, in taking from the system the power of resistance to new morbid impressions, give to them peculiar gravity.

Romberg, a German pathologist, and Luys, of Paris, who ranks as the leading microscopist of the nervous system, have carefully investigated the pathological anatomy. The dorso-lumbar region of the spinal cord, is the seat of the material lesions in muscular ataxy. These lesions occur in the posterior columns of the cord, and the roots which emerge from them. On opening the canal of the spinal cord, the *dura mater* presents a reddish tint, more or less injected, depending upon an augmentation of its normal vascularization, and this vascular injection extends to the most profound

ramifications of the vessels. The dura mater is also somewhat thickened and ardematous.

The pia mater participates in this anomalous vascularization, which becomes more pronounced as it approaches more the inferior part of the cord, and the posterior roots of the nerves, to which it adheres, so that in removing it, portions of the nervous substance are dragged away.

The lesions of the cord itself consist sometimes in a sort of grey degeneration, sometimes to a translucent and gelatinous state ; in a diminution of consistence, or on the other hand an induration, called sclerosis. Ordinarily there is a sensible reduction in the volume of the posterior columns. In the posterior roots the same alterations march parallel to those in the cord. Topineul suggests, "that this alteration of the roots is if not a consequence, at least a consecutive condition ; in no case anterior to disease of the cord, and that the alteration in the cord is the only constant lesion.

And here arises a question of exceeding interest. If it is incontestable that the cerebellum is the seat of the faculty of co-ordinating movements ; if as corollary, certain affections of this organ have for result a lack of this co-ordination ; on the other hand we have the clinical fact, that between the ataxy arising from disease of the cerebellum, and progressive locomotor ataxy, there are remarkable points of dissimilarity.

It is then in the cord, and a single portion of it that the lesion exists, and we seek therefore the reasons for the characteristic symptoms of the malady. But between these symptoms and the lesions there are at first sight singular contradictions. Locomotor ataxy being essentially characterized by troubles of motion, and loss of sensibility, entering into it as a secondary element, we should expect to find, after what we know of the distinct functions attributed to the different columns of the cord, the material lesions, not in the posterior columns, which according to the doctrine of Sir Charles Bell are the source of sensibility, but rather in the anterior columns, which are considered the place of origin of the motor nerves. It is the inverse which we really see.

This contradiction, however, is not as absolute as it appears, at least as to absence of lesion of the anterior columns. If these columns are intact, there is no paralysis, a phenomenon which is the consequence of their greater or less alteration. The troubles of motion are altogether a different order, and however exaggerated they may be, the muscular force in progressive locomotor ataxy (and this is the capital fact) is *preserved*.

Treatment.—The multiplicity of remedies by turns employed, witnesses the infidelity and powerlessness of medicine in the malady. Nitrate of silver has probably been more extensively employed than any other remedy. The records of its use give no encouragement of success. In the case we have narrated, the salts of bromine have acted favorably in controlling the pains, and to a certain extent regulating the co-ordination of movement.

