

LOCOMOTOR ATAXIA, TERMINATING AS GENERAL PARALYSIS OF THE INSANE.

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AS the relation between locomotor ataxia and general paralysis of the insane has been a problem of interest to neurologists and alienists since the investigations of Westphal in 1863, the following case is, I believe, worthy of permanent record in the proceedings of the American Neurological Association.

P., at the time of first coming under observation was forty-seven years of age. He was a man of good constitution, noted for his strength and endurance, but for three years he had not been well, during most of which period he had been treated by different physicians for "rheumatism." He was addicted to venereal excesses, and used and occasionally abused alcohol. Many years before he had had a chancre, but had not subsequently had any of the ordinary evidences of secondary or tertiary syphilis. He had first suffered from darting or shooting pains in his feet and legs. Soon he experienced sensations of numbness and tingling in his feet, and later in the little and ring fingers of his left hand. For a short time he was troubled with double vision, and his sight had diminished a little in acuteness.

I will give briefly the results of an examination made during the first week he was under observation. No paralysis was made out. Galvanic and faradic irritability were well preserved. He could not walk well after dark. He swayed and tottered on trying

to stand with his heels together, or with his eyes shut, and he could barely manage to stagger a few steps with his eyes closed. He complained of the numbness and tingling in his feet and fingers, to which I have already referred ; and the paroxysms of sharp, sudden pains in his limbs had become more frequent. He was awkward with his hands in dressing. Occasionally a disagreeable vaporous sensation would begin in his fingers, and creep up his arms, and thence spread over his body. A peculiar sense of constriction or drawing in the lower part of the abdomen had annoyed him for several months. Sexual desire had diminished. Within four weeks he had lost seventeen pounds in weight. He suffered from attacks of sleeplessness and from mental anxiety in regard to his physical condition ; but he had no symptoms of aberration of mind. He had no delusions ; and he was fully able to attend to his business, which required a large amount of physical and mental exertion.

At first he was given potassium iodide, but under its use he made no improvement. He was next placed upon a systematic course of treatment with silver nitrate, galvanization of the spine, and faradization of the extremities. A mixture of either strychnia and quinia sulphate, or of the Prussian oil of phosphorus, was occasionally substituted for the silver nitrate. A strong galvanic current was applied with broad rheophores over the entire length of the spinal column twice or three times a week, and faradization of the feet and limbs both with the metallic brush and with sponge electrodes was also employed. The improvement made apparently under this treatment perseveringly continued was remarkable. He gained in weight. The paroxysms of lancinating pains diminished very much in frequency and severity. The sense of constriction about the waist and abdomen disappeared. He ceased to have the sensations of numbness and formication. He became able to walk without trouble in the dark, and even to stand and walk with his eyes shut. After about seven months of treatment, he discontinued the use of electricity ; occasionally, however, taking medicine.

After remaining better for a few months, however, he again relapsed ; and now, in spite of all treatment, including medicines, electricity, rest, and discontinuance of business for a time, he got steadily worse. Occasionally, however, he would temporarily improve. The anæsthesia of his feet and hands deepened ; the staggering gait returned and grew worse ; every two or three weeks he would have frightful attacks of lancinating pains.

Decided mental symptoms first began to make their appearance two years after first coming under treatment. He spent his money very freely upon others as well as upon himself. His friends observed that his ideas were becoming queer and lofty; but the delirium of grandeur did not develop thoroughly until nearly a year later, when he began to talk and act in the most preposterous manner. About the same time a peculiar stagger in his speech, a slight twisting of the mouth to one side, and some tremor of the tongue and lips, became noticeable when he talked. On several occasions at this stage he had attacks of hæmoptysis, and he was troubled with a cough, and now and then with night-sweats. He lost weight steadily.

He made bargains for the purchase of valuable houses, in several instances getting himself into difficulties with persons who at first did not know that he was irresponsible. On one occasion he assembled a number of his boon-companions at a tavern, and gave them an expensive banquet in honor of his fiftieth birthday, which, he said, on account of the good time which he had had in this world, had come for him a year earlier than for ordinary mortals.

Nearly three years after the notes first made by me as to his spinal symptoms, and almost six years after the development of ataxic pains, he was sent to the Insane Department of the Pennsylvania Hospital, where I occasionally visited him.

His delusions became of the wildest character. He became irritable and hard to manage. Anæsthesia, tremor of tongue, etc., increased. On two occasions he had slight apoplectiform attacks, once accompanied by slight spasms. Later he was removed to the State Hospital for the Insane at Danville, where he remained until his death. Through the kindness of Dr. S. S. Schultz, Superintendent of the Hospital, I received some important particulars in regard to his condition about six weeks before his death. His general mental condition was very much enfeebled. He did not seem to understand where he was, or to recognize strangers from those constantly about him. He also did not seem to realize the difference between filth and cleanliness, or between different kinds of food. When given a peach to eat, he said: "Very good cook." His speech was muffled and indistinct, syllables and words often running into each other so as to be unintelligible. While apparently waiting for a word or an idea, his lips were unsteady, would quiver and be drawn together in points. Even when ready to talk

he would seemingly have to make several efforts before he could speak. His movements were weak, hesitating, and awkward. He usually remained in bed, although he was able to get up. His bowels were not paralyzed. A needle could be plunged deeply anywhere into his body or limbs without causing him any apparent discomfort. Of events which occurred previous to his sickness he appeared to retain some correct ideas, but recent occurrences left no impression. His wife visited him and stayed with him several hours, but an hour after her departure he had lost all memory of the visit.

He died exhausted, five years and four months after my first examination of his condition, and about eight years after his first so-called "rheumatic" pains.

His body was sent to Philadelphia and I obtained permission to examine the brain and spinal cord.

An autopsy was held forty-six hours after death, Drs. Eskridge and Massey giving valuable assistance. External examination showed two slight abrasions on the top of the head. The left foot, which looked swollen and enlarged as compared with the right, measured over the instep $9\frac{3}{4}$ inches; the right, $8\frac{1}{2}$ inches.

The skull-cap was found to be about normal in thickness. No adhesions of the dura mater to the skull were present. The dura was, however, adherent to the pia mater for a distance of three inches along the edge of the longitudinal fissure, about the middle of the right hemisphere, and for about one inch along the left hemisphere. A bony formation, an inch in length, and one third of an inch in width and thickness, was found in the falx opposite the middle of the first frontal convolution. A similar formation, one third of an inch long, was discovered opposite the end of the fissure of Rolando.

On the right side, the pia mater of the upper and lateral surfaces presented a deeply congested and opaque appearance over the following convolutions: a small portion of the posterior end of the first frontal, the posterior thirds of the

second and third frontal, the entire ascending frontal and ascending parietal to within an inch of the longitudinal fissure, the lower half of the superior parietal, the inferior parietal, and the first and second temporal. Over the hemisphere, in front, behind, and above this area the pia mater presented a little bluish opacity, and was hyperæmic in points. The appearances over the left hemisphere were exactly similar, except that the area of deep redness and opacity did not involve as large a surface. The convolutions covered by it were as follows: the ascending frontal, ascending parietal, inferior parietal, and upper temporal. A considerable amount of fluid escaped, apparently chiefly from the frontal and occipital regions. The convolutions of the frontal and occipital lobes were flattened, the appearance being most marked over the left frontal region. The entire left hemisphere was a little smaller than the right. The pia mater over the superior vermiform process of the cerebellum was also deeply congested and adherent. The pia mater was adherent at scattered points over both cerebrum and cerebellum. Decortication was marked. Some hyperæmia of the pia mater of both temporo-sphenoidal lobes was noted; it was more marked on the right than on the left side. The membrane across the right Sylvian fissure presented a dark, clotted appearance; a similar, but less marked condition was present on the left side. The frontal convolutions on the right side, basal surface, were more flattened out than is usual. The cerebellum was about normal in size. The medulla oblongata was small and firm.

The cornua of the ventricles were dilated; the posterior horns in particular were much enlarged.

The only notes made as to the gross appearances presented by the spinal cord and its membranes were that the pia mater was thickened, and that the cord presented an irregularly shrunken look.

The spinal cord and membranes, and numerous specimens from the brain, were examined microscopically by Dr. Carl Seiler, H. Formad, and myself. Sections from the membranes, complete transverse sections of the cord at different heights, and large sections from the medulla oblongata, pons, cerebellum, crura cerebri, optic thalami, and convolutions were prepared and examined.

The dura mater of the spinal cord in the lumbar region was found to be slightly thickened. Evidences of leptomeningitis were present; there were some adhesions between leaflets of the pia mater.

The lower lumbar region showed the connective tissue between some of the nerve-fibres much increased. The cortical layer of the posterior columns was sclerosed to the extent of about one fourth the thickness of the substance of these columns. The sclerosis in the columns of Goll and posterior root-zones was of nearly the same extent.

The walls of the vessels appeared thickened throughout the posterior columns. The sclerosis was most marked on the right side. The vessels and perivascular spaces to the right of the central spinal canal were enormously dilated. The central canal was obliterated. A small hemorrhagic infarct was observed in the right lateral column, near the posterior horn and near the periphery. The upper lumbar region showed the same changes, the sclerosis, however, being not quite so marked. The central canal and posterior fissure were obliterated. The dorsal and lower cervical regions also showed the same condition, but in decreasing intensity. The pia mater of the dorsal and lower cervical regions was thickened, and the central canal and posterior fissure obliterated.

Sclerosis in the upper cervical cord was less than in the lower portion, and was more marked in the columns of Goll. The vessels in the deep portions of the columns of Goll, and

in the posterior radical columns, were seen to be much sclerosed. Here for the first time the posterior median fissure was not obliterated.

The ganglion cells and periganglionic spaces showed a granular and atrophied condition.

Corpora amylacea were found in the lateral columns throughout the cord.

The medulla oblongata showed a great number of corpora amylacea in the lateral tracts. The restiform body of one side was much sclerosed, and but slightly so on the other side. Ganglionic cells on each side were highly granular and pigmented. Some of the centres also contained corpora amylacea. In some of the larger vessels were accumulations of white blood corpuscles. The perivascular lymph-spaces appeared widely distended.

The pons Varolii presented corpora amylacea in various places. The vessels contained white blood cells, and their walls were infiltrated with cellular elements.

The left crus cerebri exhibited the ganglionic cells extremely granular and pigmented. The vessels were sclerosed, and their walls infiltrated with cells, and some with pigment. Sclerosis was present along the course of the vessels and corpora amylacea in the white substance. In the right cerebral crus the same conditions were present as in the left side. The corpora amylacea were in greater number.

The left optic thalamus showed the walls of the vessels extremely infiltrated with cellular elements, even in capillary plexuses, and the nuclei in the neuroglia much increased along the walls of the vessels. In the right optic thalamus the changes were the same, but not as well marked.

A section of the convolutions across the fissure of Rolando showed the same state of the vessels as in the optic thalami, but less marked, and also a few corpora amylacea in differ-

ent parts. The pia mater was thickened and its vessels dilated in several portions examined. The corresponding cortical motor region of the right side gave similar appearances, with corpora amylacea more numerous.

The right and left præfrontal lobes showed an increase of nuclei in the fibres of the white substance, the walls of the vessels infiltrated with cellular elements, and numerous corpora amylacea.

Sections from the superior vermiform process of the cerebellum showed both the vessels and the cerebellar substance in a state of sclerosis. Corpora amylacea were also present. The right and the left cerebellar hemispheres presented similar conditions of the vessels and of the brain substance.

Summarizing the results of the examinations, I find that the posterior columns of the spinal cord showed marked sclerosis throughout their whole extent, and that leptomeningitis was present everywhere. The sclerosis was most marked in the lumbar region, decreasing in intensity as the cord was ascended; but was well marked throughout, both in the columns of Goll and in the posterior root-zones. The medulla oblongata, on one side, was much sclerosed, and slightly so on the other side. Sclerosis was also present in the pons, crura, optic thalami, and convolutions examined, and in the cerebellum.

The pathological appearances shown by the microscope corresponded closely to those mentioned by Hitzig, after Westphal (*Ziemssen's Cyclopædia*, vol. xii, p. 853), as occurring in the spinal cord, in dementia paralytica. The first of the two groups presents, clinically and anatomically, the symptoms of tabes dorsalis, or gray degeneration of the posterior columns. On making a transverse section of the hardened cord, the posterior columns show few or no sections of nerve-fibres, and their place is taken by a con-

nective-tissue substance. In the cervical region, Goll's cuneiform columns are especially affected; in the dorsal and lumbar regions, however, the entire area of the posterior columns is involved. In fresh preparations, numerous granular fat-cells and corpora amylacea are found. This change can be followed upward only to the beginning of the fourth ventricle.

In this case the spinal symptoms were the first to appear. Three years before coming under my care, he began to suffer with the lancinating pains of posterior sclerosis. Although, when first seen by me, and until he improved under treatment, he suffered at times from mental anxiety and sleeplessness, apparently the result of the pains and other distressing symptoms of the ataxia, no typical mental symptoms appeared until more than two years after coming under my care, and more than five years after the appearance of the first symptoms of spinal trouble.

According to Westphal, with whom Hammond agrees, no direct relation exists between the morbid process in the cord, in posterior spinal sclerosis, and that in the brain, in general paralysis of the insane. According to these authorities, neither disease is secondary to the other. They simply co-exist as the expression of an excessive proclivity to disease of the nervous system, just as any other two diseases may be present,—one in the brain and the other in the cord, without there being any direct interdependence between them. Locomotor ataxia is by no means uncommon in patients affected with the other forms of insanity. (Hammond: "Treatise on Diseases of the Nervous System," sixth edition, p. 511.)

Hamilton (*New York Medical Record*, July 29, 1876) discusses the relations of these two affections. Leidesdorf has related one case in which general paralysis was preceded by spinal symptoms. Maudsley speaks of other cases.

Calmiel says that in many cases the changes proceed from the cord upward, and Baillarger endorses his views. Charcot has proved very conclusively that disseminated sclerosis can exhibit all the symptoms of general paralysis of the insane. Obersteiner considers that mental symptoms are found in the greater proportion of cases of locomotor ataxia.

He is convinced that the symptoms of general paralysis indicate a progression of the sclerosis upward. He considers the lesions to be identical, and that it is only the seat of the change which has any thing to do with the symptoms expressed. He has also found in general paralytics who have died, a sclerosis of the cord. Tigges considers general paralysis to be a complication. M. Rey has observed nine cases of insanity associated with locomotor ataxia. In three of these the spinal sclerosis preceded the cerebral trouble, and in one the induration had extended from the posterior to the lateral column.

Hamilton details two cases, one from personal observation, and one related by Obersteiner. Obersteiner's case presents many points of similarity to the one which I have just reported.

Plaxton (*Journal of Medical Science*, July, 1878) gives an account of two cases of locomotor ataxia, with mental symptoms simulating those of general paralysis, with a pathological report by W. Bevan Lewis.

In the first case the symptoms of locomotor ataxia began eleven years before death; the mental symptoms only two years and three months before death. "He developed ideas of grandeur, wealth, and power, transcending even the power of the genius of the lamp of Aladdin to fulfil." Microscopic examination showed marked sclerosis of the posterior columns. In the lumbar region the pathological process was most advanced. The columns of Goll formed here a shrunken, fibrous tract crowded with nuclei and amy-

loid bodies. The substantia gelatinosa and posterior root-fibres exhibited intense sclerosis. The cells of the posterior horns were few in number, small, pigmented, and atrophied. In the dorsal and cervical regions the changes were the same, but less in degree than those in the lumbar cord. The antero-lateral regions of the cord were, comparatively speaking, normal. Post-mortem examination of the brain showed some wasting of the convolutions of the vertex in the frontal and parietal regions. The pia mater over these parts was slightly thickened, but presented no trace of adhesion to the subjacent cortex. The brain, as a whole, was firmer than normal. Neither the cortex nor white matter presented any abnormality to the unaided vision, but the arterioles everywhere appeared unduly coarse and prominent. The medulla oblongata was found to be very decidedly firmer than normal. Microscopic examination of the cerebrum detected nothing abnormal in the cortex or medulla of the brain.

In the second case, progressive locomotor ataxia had lasted between five and six years, insanity five months. Delirium of grandeur was here also the leading mental symptom. His fancy ran riot in all possible directions, and his belief kept pace with his fancy. On post-mortem examination the convolutions of the vertex, especially in the frontal and parietal regions, presented great wasting and atrophy. The membranes covering this wasted region were much thickened, but quite free from adhesions. The brain was extremely pale, and its consistence diminished. Serous fluid to the amount of seven ounces (compensatory effusion) escaped. Microscopic examination of the cerebrum, by Bevan Lewis, showed colloid degeneration of the white matter of the cerebrum, and of the pons and medulla oblongata. No morbid change was discovered in any portion of the cortex; and, stranger still, careful examination of the cord in the cervical, dorsal, and lumbar regions,

showed no morbid change in its nervous, connective, or vascular elements.

Dr. W. Julius Mickle (*Lancet*, May 28, 1881) reports a case of general paralysis of the insane consecutive to locomotor ataxia, very similar in most of its features to the case reported in this paper. The locomotor ataxia was of about five years' duration; the general paralysis of nearly four years' duration. The microscopical appearances in the cord were almost identical with those seen in my case.