A case of locomotor ataxy, without diseases of the posterior columns of the spinal cord / by A. Hughes Bennett.

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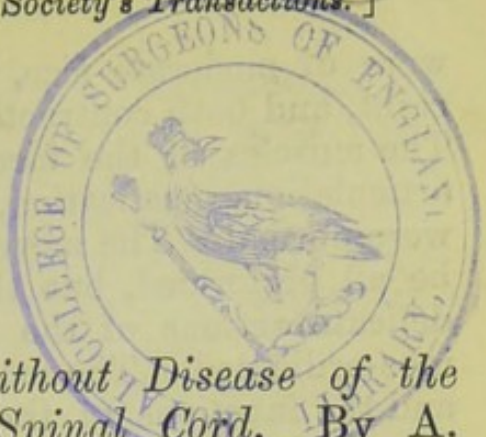

A Case of Locomotor Ataxy, without Disease of the Posterior Columns of the Spinal Cord. By A. Hughes Bennett, M.D. *Read February* 27, 1885.

THE case to which I beg to direct the attention of the Society is specially interesting from the circumstance that, although the patient during life presented all the prominent symptoms of so-called tabes dorsalis, the posterior columns and cornua of the spinal cord were found after death to be without a trace of disease. This fact suggests certain important considerations concerning the physiology and pathology of locomotor ataxy.

The patient was a gamekeeper, aged 48, who stated that he had always been a healthy man, never having suffered from syphilis or other disorder. About fifteen years ago he complained of severe pains in his legs, which continued for three months, from which symptoms he completely recovered. He afterwards remained well till nine months before he came under observation. He then for the first time experienced gradual weakness of the legs, so that he was easily fatigued and was unable to do a full day's work. This slowly increased, without, however, actually incapacitating him from his duties. Some months afterwards he was attacked with severe shooting pains in his lower extremities, occurring in paroxysms, and shifting from place to place. With the advent of these the weakness of the legs augmented, and was accompanied with a sensation of numbness in the feet. He was therefore compelled to give up his situation as a nobleman's gamekeeper.

On examination the patient was found in good general health. There was no evidence that his intelligence was abnormally affected, but he was an uneducated and stupid man from whom it was difficult to obtain a satisfactory account of his illness. There were no headaches or cerebral symptoms except occasional giddiness. Vision in both eyes was good, there was no paralysis of the ocular muscles, and the fundi of both eyes were normal. Voluntary movements of the eyeballs were accompanied with slight nystagmus. The pupils

were of moderate size, the left somewhat smaller than the right, and both contracted well to light and accommodation. The muscles of the face and tongue were as in health, and articulation, mastication, deglutition, and all the special senses were normal. The movements of the upper extremities could be all performed, but the grasp of the hands was weak, and delicate actions of the fingers were unsteady and slightly inco-ordinated. The movements of the trunk seemed normal, and the functions of the bladder, rectum, and sexual organs appeared to be intact. The patient felt his lower extremities to be weak. He walked with a slow, unsteady, hesitating, and markedly ataxic gait. Without assistance he staggered, had difficulty in turning or walking in a straight line. He could not stand on one leg, and all his motor difficulties were increased in the dark. When the eyes were closed the patient swayed and would have fallen if not supported. In bed the movements of the lower limbs were vigorous and displayed considerable force, but they were accompanied by the typical phenomena of inco-ordination. The knee jerk on both sides was totally absent and the plantar reflexes were diminished. There was very slight rigidity of the large joints, which was easily overcome by passive movement. The sensibility of the skin to touch and pain appeared to be everywhere normal, except in the feet and legs, where it was both diminished and retarded. The patient complained of a constant dull aching in his back and lower limbs, and this was almost daily accompanied by attacks of lancinating pains which shifted about from place to place, and which were sometimes very severe. The muscles throughout the body were thin and spare but nowhere specially atrophied; their mechanical irritability and electrical reactions were unimpaired. The other organs and functions of the body were normal.

The patient died after a residence of two and a half months in the hospital. From the beginning the weakness of the legs and insecurity of gait increased, so that in about six weeks he could not stand. The lancinating pains were very constant and severe. For many weeks there were attacks of uncontrollable vomiting. Emaciation ensued and the general health broke up. During the last week the patient was feverish, restless, and prostrate; he became semi-comatose, and finally, after a severe convulsive attack, died. Otherwise no new or special symptoms developed.

Post-mortem examination (30 hours after death).—Permission was given to examine the brain and spinal cord only.

The entire body was thin and emaciated, but nowhere was there any special atrophy. On the right gluteal region there was a superficial bedsore about three inches in diameter, and there was commencing change in the skin on the corresponding left side and over the sacrum. On removing the calvarium the membranes of the brain were found deeply congested; there were considerable œdema and sub-arachnoid effusion, but no adhesions. The convolutions were somewhat flattened, especially on the left side, in the frontal and parietal regions, otherwise they were normal. On removing the brain several ounces of cerebro-spinal fluid escaped. The vascular congestion was found to be universal; in other respects the blood-vessels were healthy. On slicing through the hemispheres the cerebral substance was seen to be deeply injected, the lateral ventricles each contained about half an ounce of clear yellow fluid, and the choroid plexuses were œdematous. In the substance of each frontal lobe, involving the anterior cornua of the lateral ventricle, was a circumscribed patch of softening, on the left side about the size of a hen's egg, on the right somewhat smaller. These extended inwards to the third ventricle and longitudinal fissure, downwards to the base of the brain, and upwards to within an inch of the cortical substance. The under and fore part of the corpus callosum, the lamina cinerea, and the tuber cinereum were involved in the softening; but the fornix, the corpora albicantia, the corpora striata, and optic thalami were apparently normal. There was a similar patch of softening in the cerebellum about the size of a walnut, situated at the left side of the anterior free margin of those lobes forming the upper boundary of the great longitudinal fissure. The middle and inferior peduncles on the left side were softened, but the superior remained of normal consistency. To the naked eye the pons, medulla, corpora quadrigemina, cerebral peduncles, and other parts of the encephalon appeared healthy.

Microscopical examination of the softened cerebral matter showed much granular detritus, broken-down cells, Gluge's corpuscles, numerous oil-globules, and shrivelled blood-corpuscles.

On opening the spinal canal the vessels of the membranes were seen to be deeply congested, especially in the lower dorsal region, where there were found small patches of superficial hæmorrhage. There were no adhesions either to the bone or to the cord, and the latter was enucleated without difficulty. The pia mater was thickened, especially at its posterior aspect, most marked inferiorly, and gradually diminishing from below upwards. Behind, the membrane was studded throughout

with what to the naked eye appeared to be a number of isolated tumours. These were of a round or oval shape, of firm hard consistence, of a white colour, and they varied in size from a mustard seed to a pea. At the medullary end of the cord these were few in number, but on proceeding downwards they gradually augmented, so that in the middle dorsal region they were perhaps from six to eight to the square inch (Pl. 8, fig. 2). Continuing to increase in number towards the lower portion of the cord, they became in the lumbar region quite confluent so as to make the cord in that situation of abnormal thickness and rigidity. These growths were seen to be attached to and to surround the posterior roots. To the naked eye none of these tumours were seen on the anterior or lateral aspects of the cord.

The medulla and cord were first hardened in Muller's fluid and strong methylated spirit. They were then immersed for several days in absolute alcohol, and afterwards embedded in celloidin. Sections were made by Dr. Hebb, Pathologist to the Westminster Hospital, to whom I am indebted for the following description of the microscopical appearances. "There was general thickening of the pia mater most marked at the anterior and posterior aspects. This diminished gradually from below upwards, and at any given level was much more marked behind than in front. In the lumbar region the neoplasm which constituted the thickening of the membranes closely embraced the posterior, and to a slight extent the anterior roots; in the dorsal region the posterior only, and in the cervical region the cell growth had greatly diminished in extent and did not involve any of the nerve-roots. In a limited portion of the lumbar region, one anterior cornu was involved by the disease, the cells of the neoplasm having advanced along the lines of the cornual rootlets and blood-vessels. This seemed to have invaded the normal structures without causing their destruction to any appreciable extent. With this exception the nervous tissues of the cord were normal (Pl. 7, figs. 1 and 2). Some of the anterior spinal roots were involved in the growth, but only to a limited extent in the lower lumbar region. Their structure on section seemed normal. Although the tumour closely surrounded the posterior roots on both sides throughout the dorsal and lumbar regions their histological condition appeared to have suffered little demonstrable change. Examined with a high power (one twelfth of an inch) the only peculiarity that could be detected was unusual swelling of the white substance of Schwann. The

state of the nerve-trunks of the body with their peripheral terminations in the muscles and skin, were not examined.

"The medulla oblongata, which to the naked eye appeared normal, after staining, disclosed in its upper part a morbid mass measuring 0·5 × 0·7 cm., which extended from the floor of the fourth ventricle, into which it projected, to the centre of the bulb, towards the lower portion of which it diminished in size and was limited to its centre, surrounding the central canal, thus being of conical shape, the base upwards (Pl. 8, figs. 3 and 4). The morbid material here also seemed only to infiltrate the otherwise healthy tissues rather than destroy them. In addition to this fairly circumscribed mass there were a number of minute patches of similar nature scattered throughout the substance of the medulla.

"The neoplasm was everywhere composed of round cells, and was sarcomatous in character (Pl. 8, fig. 1)."

Commentary.—In commenting on the facts of this case, special attention is directed to the pathological and clinical phenomena.

1. *Pathological.*—The patches of softening in the frontal lobes, and in the cerebellum and its peduncles, were probably recent, and due to the general cerebral inflammation, which supervened shortly before death.

The sarcomatous mass as seen by the microscope in the centre of the medulla, was in size and shape fairly defined. The morbid process was not strictly limited to this area, but was dotted here and there all over the bulb. Such a pathological condition is extremely rare. The fact that the growth seemed to infiltrate and displace the normal tissues rather than destroy them, may explain the almost total absence of serious bulbar symptoms during life.

Disease of the same nature involving the spinal membranes, appearing to the naked eye as a series of small multiple tumours confined to their posterior aspect, and surrounding the posterior roots, is also an interesting and almost unique pathological condition. Histologically the lesion was found to leave the cord itself absolutely intact, except at one anterior cornu, in a limited portion of the lumbar region, where the tissues, as in the case of the medulla, were displaced but not extensively destroyed. This also accounts for the absence of symptoms due to serious implication of that locality. The anterior roots were seen to be only slightly implicated, which harmonises with the fact that there was no paralysis, wasting of the muscles, or abnormal electrical reactions during life.

Finally, the posterior roots were extensively involved, and although the anatomical changes in their structure were slight and indeed doubtful, they were sufficient to induce marked abnormalities in the functions of the sensory tracts.

2. *Clinical.*—The points of clinical interest may be considered under two heads, namely the phenomena connected with disease of the medulla, and of the spinal cord. From the position and extent of the growth in the former it might have been supposed that important bulbar centres would have been involved and corresponding symptoms produced. Yet these during life were conspicuous by their absence. There was no visual disturbance and no facial paralysis or spasm; the special senses were intact; articulation, vocalisation, deglutition, respiration, and circulation were normal, and there was no optic neuritis or glycosuria. The only phenomena attributable to derangement of the medulla were giddiness, slight nystagmus, and during the last few weeks of life persistent vomiting. The explanation of this absence of symptoms has already been suggested.

The clinical point illustrated by this case, to which it is the object of this paper specially to direct attention, is the relation which exists between the spinal symptoms and the lesion of the posterior roots of the cord discovered after death. The patient had been examined by many physicians, all of whom had pronounced him to be suffering from tabes dorsalis, and his condition in most respects justified that diagnosis. There was inco-ordination in the movements of the limbs and chiefly in the lower extremities, with a typical ataxic gait, increased in the dark. There was at the same time no appreciable failure in the gross power of the muscles, nor were these atrophied. The Romberg symptom was present, the patient swaying and losing his balance when his eyes were closed. There were paroxysms of characteristic lancinating pains, and deficient and retarded sensibility in the skin of the lower limbs. The plantar reflexes were diminished, and the knee-jerk phenomena totally abolished. This sequence of events, although not constituting a complete clinical picture of tabes, warranted, it must be admitted, the conclusion during life that the patient was suffering from that disease. It is true that there was no affection of vision, or trace of optic atrophy. The pupils, though unequal in size, were not paralysed to the stimulus of light or accommodation. There was no abnormality of the vesical or sexual organs, and the disease had only been in existence for nine months. But clinical

experience has shown that none of these circumstances preclude the presence of those central changes which are usually associated with tabes dorsalis, or that the absence of these symptoms is incompatible with a pronounced specimen of that affection as we now recognise it at the bedside. Now, it is universally assumed that the lesion which causes this train of phenomena is situated in the posterior columns of the spinal cord, and our standard text-book on nervous diseases thus expresses the general belief:—"Sclerosis of the posterior root zones for a considerable portion of their longitudinal extent, is the essential morbid alteration in locomotor ataxia."*

In the case before us there were all the prominent symptoms of that affection, without a trace of degeneration of any portion of the posterior columns, the posterior cornua, or the posterior roots within the circle of the spinal membranes. Evidence is thus adduced to show that, in the production of that group of symptoms comprised under the term tabes dorsalis, disease of the posterior columns of the cord is not an essential factor, and that this may be fully developed, that centre being in a perfectly normal condition. This fact had already been observed by M. Dejerine, who has placed at least two cases on record presenting features during life almost exactly the same as the case at present under consideration, in which after death the cord, spinal roots, and nerve-trunks were found healthy.† In these, however, he discovered that there was parenchymatous neuritis of the nerve-endings, in the skin, and, to a lesser degree, in the muscles of the limbs affected. He concluded from this that the ataxic symptoms were due to disease of the sentient nerves in the skin and muscles, producing anæsthesia of the former, and loss of the so-called muscular sense in the latter. Afferent paths were thus interrupted at the periphery which alone was capable of producing the typical condition of ataxy without central change. This form of the disease he proposed to call "*nervo-tabes périphérique*," in contradistinction to the classical tabes dorsalis. In the present case it is a matter of regret that the cutaneous and muscular nerve-endings were not histologically examined; at the same time facts show that the primary disease did not originate there, the post-mortem revelations proving the lesion to have begun in another portion of the nervous system. There can be no doubt that

* *A Treatise on Diseases of the Nervous System*, by James Ross, M.D., second edition, vol. ii, p. 57.

† "Études sur le Nervo-tabes périphérique," par J. Dejerine, *Arch. de Physiologie*, 3me série, vol. iii, p. 231.

locomotor ataxy, as generally met with in practice, is usually associated with degeneration of the posterior root zones. The observations of M. Dejerine demonstrate that symptoms indistinguishable from this may occur without any appreciable lesion of the cord, and may be due to degeneration of the peripheral nerves only. This case makes it evident that precisely similar results may follow interruption of the co-ordinating paths in a third locality, namely, in the posterior nerve-roots. Van Deen and Claud Bernard experimentally demonstrated that the section of these in animals induced total inability to steadily utilise and co-ordinate movement, without causing loss of motor power; in short, a condition of extreme and typical ataxia. We have here before us a repetition of this experiment, produced by disease, and followed by precisely the same physiological phenomena. The posterior roots throughout the dorsal and lumbar regions were embedded in a sarcomatous mass. Pressure on their fibres was the consequence, accompanied by the symptoms of tabes dorsalis. Thus may the anæsthesia, analgesia, and retardation of sensibility be explained. Irritation of the sensory filaments caused the neuralgia and attacks of lancinating pains. Interruption at this portion of the reflex arc accounts for the alteration of the tendon and other phenomena, as well as the production of ataxy and the Romberg symptom. So far then this case supports the view that those elements which convey the impulses regulating co-ordinate movement, are situated in the afferent paths of the nervous system. The disease in this case being limited to the posterior roots, constitutes a more conclusive fact in favour of that theory even than those in which degeneration of the peripheral nerve-endings alone was demonstrated.

It is admitted that in this observation there are two possible flaws against absolute proof of the preceding proposition, namely the existence of implication by disease of the medulla, and anterior roots. Reasons, however, have already been given for the opinion that these complications, for practical purposes, need not militate against the argument.

Whether Dejerine's lesion was primary or secondary to undemonstrable trophic influences in the nerve-centres, need not be here discussed. But, whatever theory be advanced to explain the physiology of locomotor ataxy, and the prominent symptoms of so-called tabes dorsalis, this case serves to show that the point at which, in the causation of the phenomena, the nervous path is interrupted, need not of necessity, as is

generally asserted, be primarily situated in the posterior root zones of the spinal cord.

Reflection on this fact suggests that we have not yet fathomed the anatomical substratum of that Protean disorder which we at present recognise under the term tabes dorsalis. This assemblage of symptoms probably consists of a combination of different pathological conditions, many of which are represented by phenomena common to all, and each of which in time may be differentiated. It is possible that a case such as the present may be a link in the chain of discovery.

Such considerations are of practical utility, as the tendency of the present day is to assume that certain isolated symptoms of necessity indicate the existence of serious and progressive central disease. In practice we see persons who happen to suffer from pains in their extremities, peculiarities of their pupils, alteration in their reflexes, or from other special nervous symptoms, often pronounced as the subjects of tabes dorsalis. These are either made the victims of prolonged and unnecessary treatment on that hypothesis, or doomed as hopelessly incurable. The case under consideration, although it ultimately ended fatally, proves that our present conceptions of the pathology of locomotor ataxy must be modified, and that the generally believed classical lesion for its production is not without exception. It further suggests that there may be other more innocent or removable circumstances which may give rise to the same series of symptoms.

DESCRIPTION OF PLATE VIII, ILLUSTRATING DR. BENNETT'S CASE OF LOCOMOTOR ATAXY.

FIG. 1.—Transverse section of the spinal cord through the dorsal region, showing the posterior roots involved in sarcomatous growth.

FIG. 2.—Transverse section of the spinal cord through the lumbar region, showing the posterior roots, and, in a lesser degree, the anterior roots and one anterior cornu involved in sarcomatous growth.

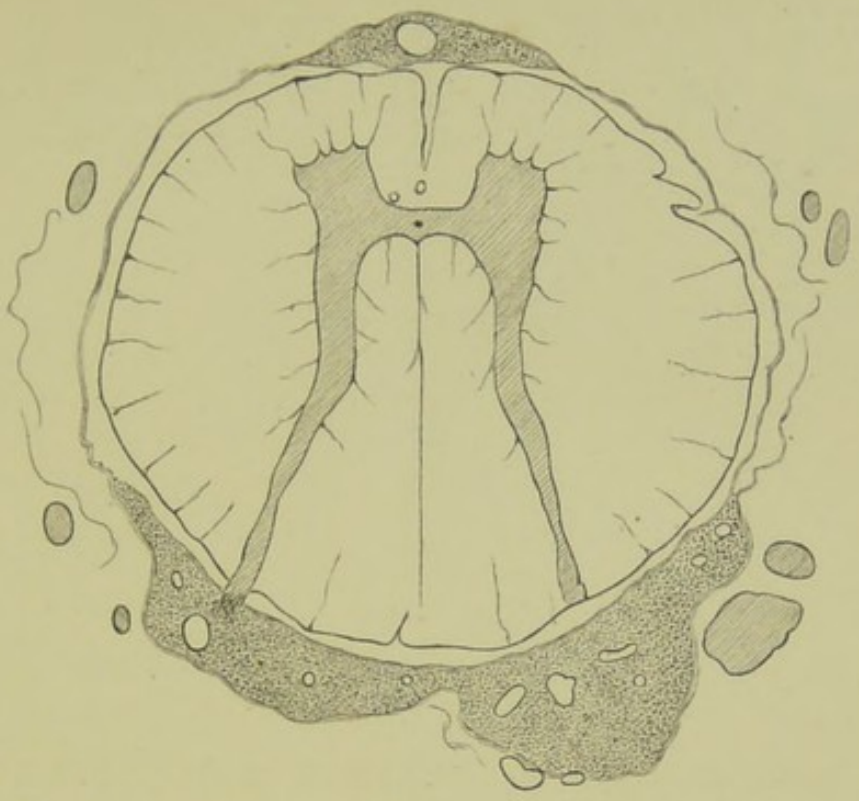

Fig. 1.

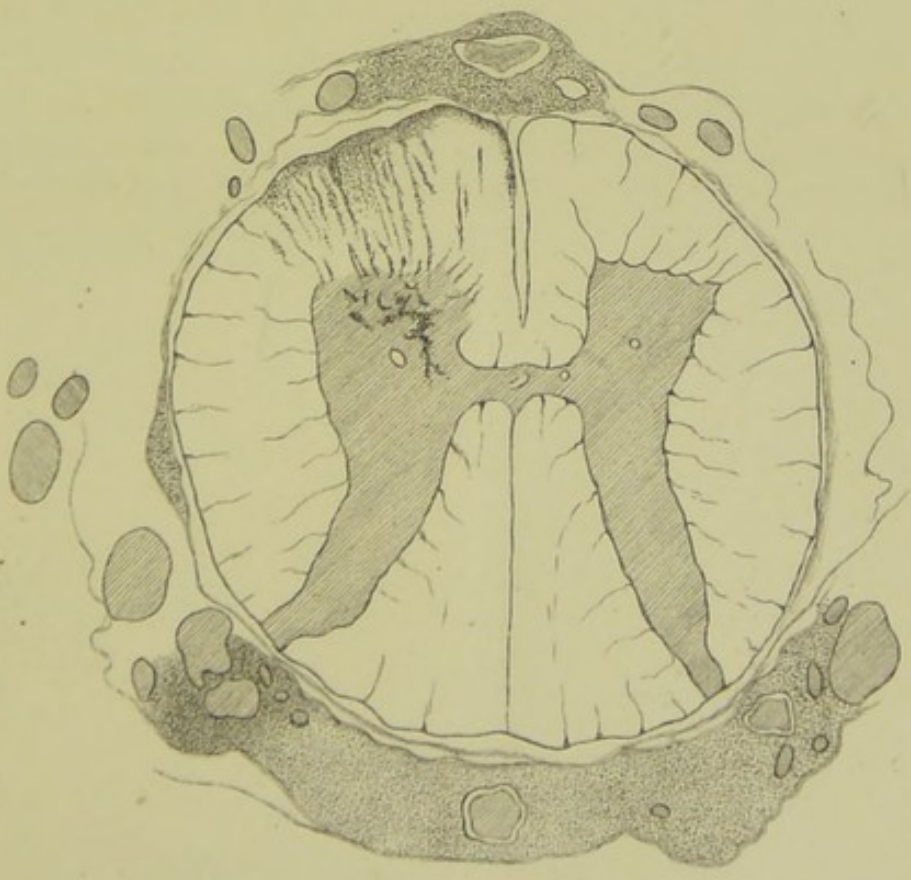

Fig. 2.

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Dr Hughes Bennett's case of Locomotor Ataxy

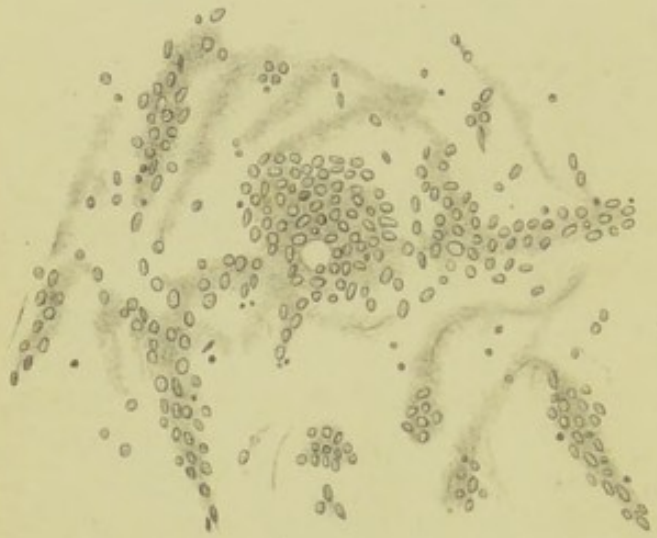

Fig. 1.

Fig. 2.

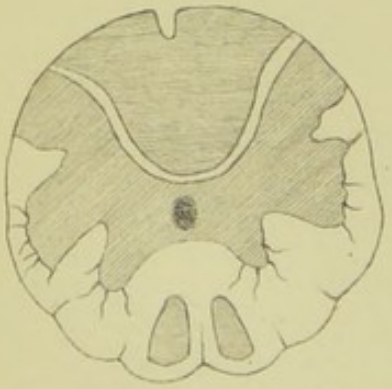

Fig. 3.

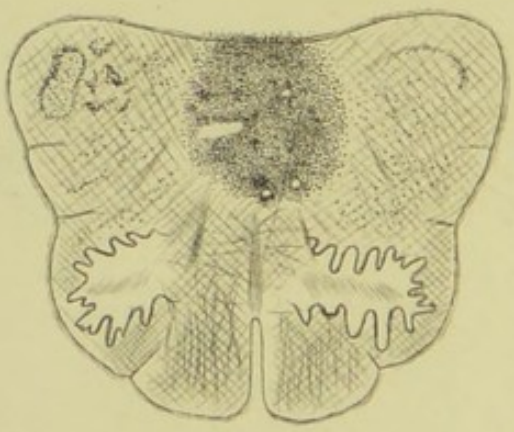

Fig. 4.

Dr Hughes Bennett's case of Locomotor Ataxy.

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DESCRIPTION OF PLATE VII, ILLUSTRATING DR. BENNETT'S CASE OF LOCOMOTOR ATAXY.

FIG. 1.—Section of morbid growth, 200 diam., showing cellular structure of sarcoma.

FIG. 2.—Naked-eye appearance of the middle dorsal region of the spinal cord, showing multiple sarcomatous tumours scattered over the posterior aspect of the pia mater.

FIG. 3.—Transverse section at the lower part of the medulla oblongata, showing the central canal surrounded by sarcomatous growth.

FIG. 4.—Transverse section at the upper part of the medulla oblongata, showing the floor of the fourth ventricle involved by sarcomatous growth.