Case of progressive atrophy of the muscles of the hands : enlargement of the ventricle of the cord in the cervical region, with atrophy of the gray matter: (hydromyelus) / by William Gull, M.D.

CASE

OF

PROGRESSIVE ATROPHY

OF THE

MUSCLES OF THE HANDS:

ENLARGEMENT OF THE VENTRICLE OF THE CORD IN THE CERVICAL REGION, WITH ATROPHY OF THE GRAY MATTER: (HYDROMYELUS).

By WILLIAM GULL, M.D.

The following case is given as a contribution to the pathology of "progressive muscular atrophy."

Three classes of cases are as yet confused together under this term, and its synonym, "wasting palsy."

Progressive, or perhaps better, "excessive" muscular atrophy, may arise from primary lesion of the muscular elements—or from lesion of the trunks or branches of the nerves—or from morbid changes in the gray matter of the cord. It is the difficulty of distinguishing the primary seat of disease in each of these classes of cases, which has led to exclusive, and therefore erroneous views of their pathology, one observer maintaining that progressive muscular atrophy is always a peripheral affection, whilst another asserts that it has constantly a centric or spinal origin.

Besides the forms here enumerated, there is a fourth class, in which muscular nutrition fails from a morbid diathesis,

allied to rickets. It has nothing in common with the pathology of the former cases but the want of muscular power. Dr. Meryon's[1] are good examples. I have seen the disease in two girls of one family. It is a malady beginning in, and limited to childhood, and peculiar to it. The muscles are not diminished in bulk at the commencement of the disease, nor is there necessarily any paralysis during the whole course of the case, but only a lentor and feebleness of movement. This cursory mention of such cases is sufficient to show with what little practical value they can be included in the first enumeration. No doubt much of the obscurity which at present besets this subject is favoured by the assumption, that where disease was not discovered in the cord, it did not exist; happily, however, positive assertions from negative evidence are at this day considered of less weight. We cannot peruse recorded cases of progressive muscular atrophy, without feeling how unsatisfactory, in most of them, is the post-mortem examination of the cord, and must hesitate to accept the conclusion that it was free from lesion, because none was discovered. Dr. Beale's 'Archives' (No. 9), contain an almost critical case in illustration of these remarks, and certainly, but for the rare skill of Mr. Lockhart Clarke, it might have been recorded as one of muscular atrophy, the cord being healthy. Thanks, however, to the means we possess of investigating ultimate structure, Mr. Clarke was able to show that there were certain areas of the gray matter which had undergone marked change of a morbid character, although the cord had an entirely healthy appearance. During the life of the patient referred to, there was a difference of opinion as to the primary seat of the malady, and but for such an exhaustive examination as it received after death, instead of its being a contribution to our knowledge, the record of the case would have served only to give strength to false assumptions, and fortify us in the error of assuming that all is sound where imperfect examination detects no weakness.

The following case, on its entrance into the hospital, gave rise to the same question; whether the muscular atrophy had a peripheral or central cause. The patient was a journeyman tailor, working hard at his business in London, and, therefore,

[1] 'Med.-Chir. Trans.,' vol. xxxv, p. 73.

of necessity using the muscles of his hands in an extreme degree. Here, therefore, was a possible, and not an improbable cause of a primary affection of the muscles themselves, but, as was remarked in a clinical lecture given on the case, it was to be remembered that the will does not directly act upon the muscles in voluntary movement, but upon the gray matter of the cord to which the nervous filaments are connected, and therefore, that it is as reasonable to infer a lesion of the gray matter from over-work, as of the muscles.

The first symptom, in this case, was inability to extend the little and ring fingers of the right hand, with a sense of coldness and numbness in the part. This was the only complaint for eleven months, and, no doubt, if the patient had been seen during that time, it might have been thought more probable by some that the disease lay in the muscular tissue than in the centre of the cord. It was not until after eleven months, that the adjoining middle finger began to fail in a similar way. A perusal of the case will show, that the centre of the cord had by this time undergone extensive changes, yet it was not until the left hand became affected, that the central character of the lesion began to appear, and even then it might have been contended, that the symmetry of the muscular affection was owing to the same conditions of over-wear in both hands. Had it not been for typhus fever, which then prevailed with unusual virulence in the hospital, we should not have known how great morbid changes the central gray matter of the cord may undergo, with but slight and limited, and only slowly progressive peripheral effects. The lesion discovered after death was evidently in no way connected with the attack of fever, nor in any way affected by it. The tissues at fault showed no traces of recent activity about them. This case is therefore another instance of atrophy progressing from muscle to muscle in the slowest way, and unattended by any of the common proofs of central disease, though depending upon it.

It also suggests some doubt respecting the validity of the present theories of the function of the gray matter of the cord. A glance at the annexed plate of a transverse section of the cord in this case, will show how large a part of the gray matter may be slowly removed, without affecting sensation to any corresponding extent, and without disturbing the general

functions of the cord, or the influence of the brain upon the parts below.

Although there were no other remains of the gray matter in certain parts of the cervical region but the anterior cornua, the patient was still able to walk perfectly well, and to move the arms freely in all directions, and the sphincters were good, nor was there any affection of sensation in any part, except a feeling of numbness in the right hand.

What the nature of the change in the cord was, may be a matter for speculation. So far as it affected the gray matter, it seemed to be no more than atrophy from distension of the ventricle of the cord, by an accumulation of fluid in it. A chronic cervical *hydromyelus*, comparable to a chronic *hydrocephalus*.

It is noticeable that normal epithelium still lined this extremely dilated ventricle.

The appearance of a distinct membrane to the cavity was produced by condensation of the normal textures pressed outwards, and not by any new plasma.

The extent to which the disease reached in a longitudinal direction, is shown in the woodcut, the dotted outline in the middle of the cord indicating the extent and form of the cavity in the interior.

The greatest diameter of the cavity was opposite the origin of the seventh or last cervical nerve, and hence the peripheral effects were chiefly marked in the branches of the ulnar nerve, which here has its principal origin.

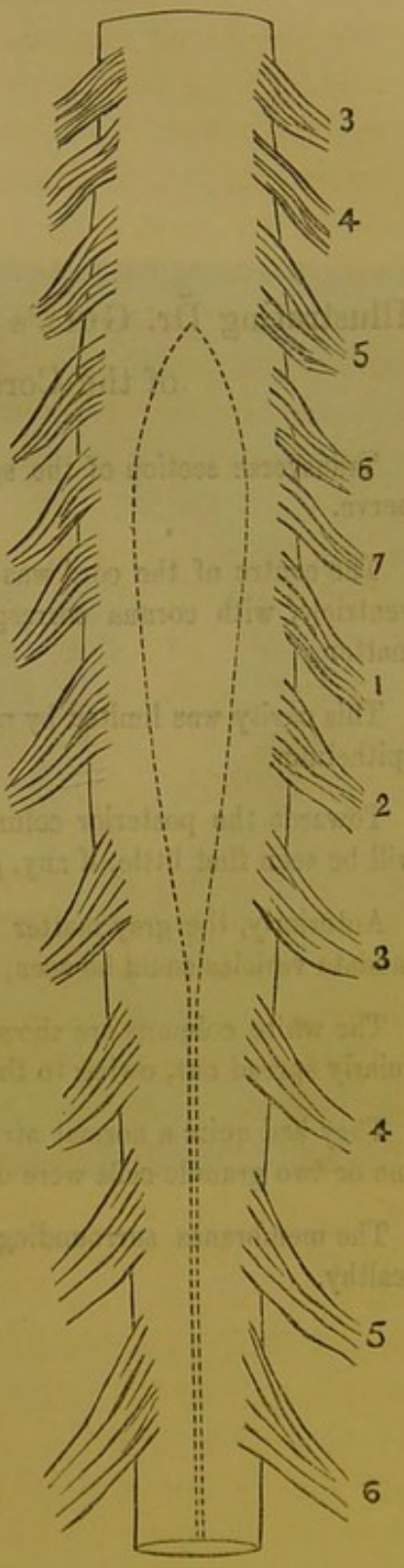
The form of the cavity, on a transverse section, is shown in the plate. It will be observed that it was not a simple circular dilatation, but corresponded to the general disposition of the gray matter and its cornua.

G. B—, æt. 44, a journeyman tailor, of sober habits, was admitted into the clinical ward, February 5th, 1862. States that he has always been healthy and strong. Never had any injury to his back. Thirteen months ago, when working in London, the fourth and little finger of the right hand became weak and flexed, without any assignable cause. The hand was cold, and there was a feeling of numbness in the fingers, but no pain. This give rise to a good deal of inconvenience, but he was able to continue working at his trade. Two months ago, the middle

finger of the same hand became suddenly affected, and three weeks ago the three inner fingers of the left hand became weak and flexed in the same way, but without any numbness. The hands gradually wasted. The arms are not affected. Seven weeks ago he had pains through his chest, and a feeling of tightness across the upper part. He is pale, complains only of wasting and weakness of the hands; has no pain in them, but the right is cold, with a feeling of numbness. The left hand is not so cold, and the sensation in it is perfect. He can move both thumbs and index fingers freely; he can also extend the first phalanges of the other fingers of both hands, but not in the least degree the second and third phalanges, which are gently flexed towards the palm. The interosseous spaces on the backs of the hands are sunken from the wasting of the muscles. The palms of the hands are hollow, and the flexor tendons very prominent. The thenar eminences are wasted, and the hypothenar almost entirely gone, particularly on the right side. The motion of the wrist joints is unaffected. He can move the arms freely in all directions. Can walk perfectly well.

At the upper part of the dorsal region there is slight flattening of the natural curve of the spine, from the long muscles of the back being at this part wasted. Pressure on the fourth dorsal spinous process causes a sharp, pricking pain, as of a knife running into the part, but when the part is not touched he has no pain. No pain on pressing the other spinous processes; no affection of sensation in any part, except the feeling of numbness in the right hand; sphincters good; urine normal; appetite and digestion good. He was put upon a full diet, and the wasted muscles were daily galvanized by an intermittent current. A fortnight after admission he had gained power in the hands. He said he felt them stronger and more pliable after each application of the galvanism. It was noted that, with a moderate current, the contractility of the muscles of both hands was good, but more particularly in the short muscles of the thumb, which were least wasted. Sensibility not so acute in the right hand as in the left, but no marked anæsthesia of either. Both hands were rather cold. A few days after this report the patient sickened with typhus fever, and died on the 8th of March.

A post-mortem examination was not permitted at the hospital, and it was only after much difficulty that the cord could be obtained. The bones and ligaments of the spine were healthy; the membranes of the cord healthy; the exterior of the cord presented nothing abnormal, except that the cervical enlargement appeared broader and somewhat flattened. On making transverse sections, the white columns had their normal consistence and texture, but the centre of the cord had a large cavity, beginning at the fifth cervical, enlarging downwards to the seventh, and from thence tapering as in the accompanying woodcut. The appearance of the cord on a transverse section at the origin of the seventh cervical nerve is shown in the annexed plate. It will be seen that the only remains of the gray matter are at the anterior part of the cavity behind the anterior columns. Here the caudate vesicles had their normal size and structure; the pigment, nucleus, and nucleolus being well marked, and the tubular structure unaltered. The cavity in the cord was bounded by a layer of condensed gray substance, which could be separated as a distinct membrane. On its interior surface, forming the lining of the cavity, were a number of delicate, elongated, nuclear bodies, apparently epithelium. One or two granule cells were found scattered amongst the white columns, but no further traces of any active tissue change. The roots of the nerves appeared normal, and contained healthy tubules. The character of the fluid filling the cavity could not be ascertained, as it escaped in the removal of the cord from the spinal canal.



PLATE

Illustrating Dr. Gull's case of Enlargement of the Ventricle of the Cord in the Cervical Region.

Transverse section of the spinal cord, at the origin of the seventh cervical nerve.

The centre of the cord was hollowed out into an irregular square cavity or ventricle, with cornua corresponding to the general disposition of the gray matter.

This cavity was limited by condensed gray matter, and lined by an imperfect epithelium.

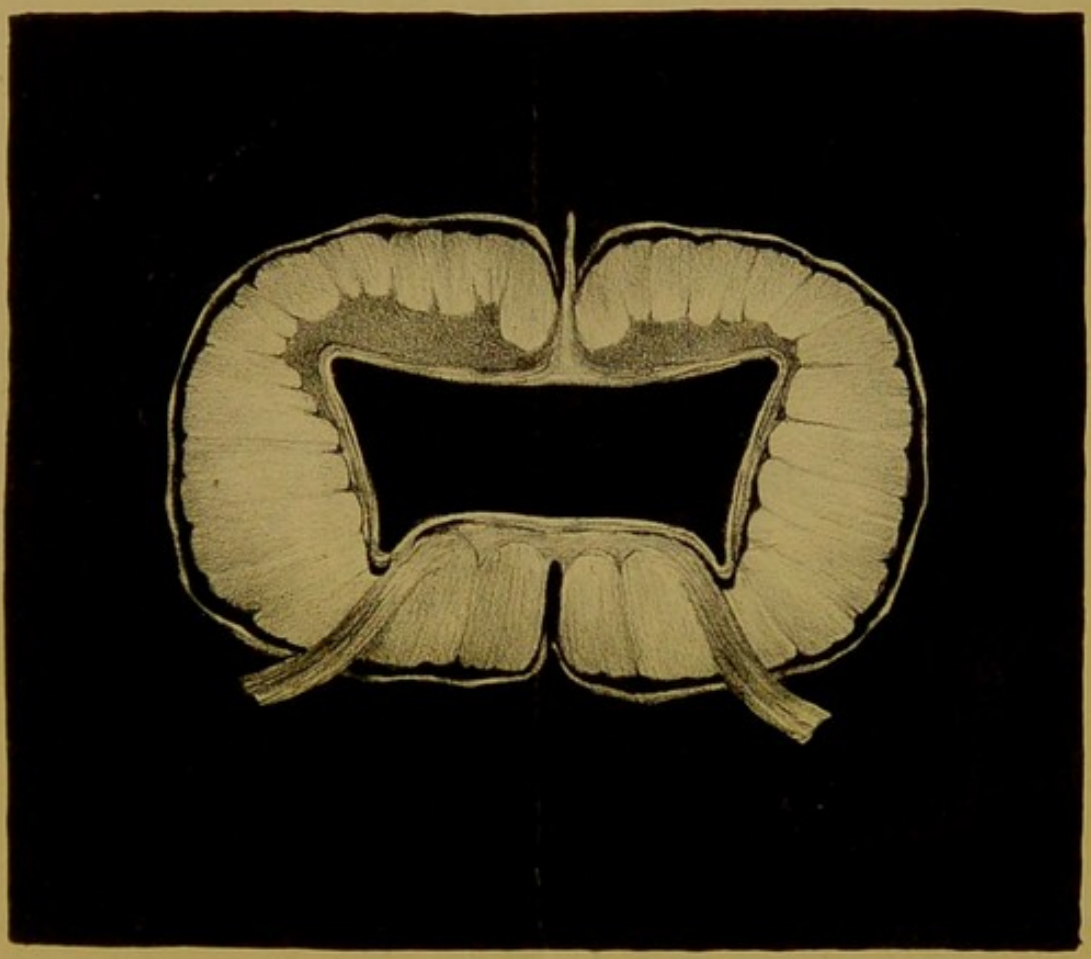
Towards the posterior columns, and at the origin of the posterior roots, it will be seen that little, if any, gray matter was left.

Anteriorly, the gray matter is spread out, and in it, by the microscope, the caudate vesicles could be seen, having their normal structure.

The white columns are shown in the drawing a little disintegrated, and irregularly spread out, owing to the thinness of the section.

They had quite a normal structure, except that, after repeated examination, one or two granule cells were detected in them.

The membranes surrounding the cord, shown in outline in the plate, were healthy.



W. Hurst, delt.

Day & Son, Lith.rs to the Queen.