Clinical Cases.

ON A CASE OF DIFFUSE (SYPHILITIC ?) SCLEROSIS OF THE SPINAL CORD PRODUCING SYMPTOMS OF POSTERO-LATERAL SCLEROSIS.

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W. H., æt. 60, porter, was admitted into the Manchester Infirmary on April 7th, 1866.

Family history.—The patient's father died of apoplexy, his mother of dropsy and heart disease. He lost three brothers, one of whom died from consumption; the cause of death of the other two is not known to the patient. One sister is living, and enjoying good health. The patient has three sons and three daughters, all enjoying good health.

Previous history.—He has been employed as a porter in removing furniture; he had to work often twelve to sixteen hours per day for a whole week, and has often exposed himself to cold and wet. He has led a steady life. He has been married twice; the first time forty years ago, the second time eighteen months ago.

Previous health.—He had inflammation of the lungs when a child; 12 years ago he suffered from an attack of gonorrhœa, which, however, soon passed off. He remained well till five weeks after his second marriage, when he noticed a swelling in the left groin, and also some urethral discharge. He did not notice any sore on the penis. He went to his family medical attendant, who told him that he had contracted syphilis. He soon after suffered from an inflammation of the eyes, but had no other symptoms, such as a rash, sore throat, &c. Under active treatment the swelling in the groin disappeared, the discharge ceased, and the inflammation of the eyes subsided. Four months before admission he began to stagger in his walk, and he noticed that he began to lose the control over his legs. Occasionally he felt his legs give way, his legs felt weaker, yet he was able to walk well with the help of stick. He suffered from some pain in the lower part of the spine, but had no pains in the legs, nor did he feel any numbness in the feet or legs. Towards the end of February the patient fell, and soon after noticed that the legs were more paralysed, so that it

was impossible for him to walk. He then also noticed that at times he was unconscious of his passing urine or fæces, while at other times he was conscious of these acts. A few weeks before admission he found that he had lost the control over his bladder completely, and the urine dribbled away. His general health remained fairly good; he never suffered from headache or vertigo.

State on admission.—The patient appears well nourished, but is somewhat anæmic. The examination of the external surface only reveals the presence of a few indurated glands in the left groin and in the back of the neck. The examination of the thoracic and abdominal viscera shows beyond the bladder and rectum affection normal relations. The tongue had a glistening, smooth appearance, and showed the characteristic signs of superficial syphilitic glossitis. The anterior part has quite a lobulated appearance, due to masses of firm fibrous tissue passing in different directions; several small superficial erosions were also visible. The temperature is normal; pulse 88, small, and compressible.

Examination of the nervous system.—The pupils are equal, the left a little irregular in its outline, due to adhesion of the iris. They react but very feebly to light, but better to accommodation. The field of vision is good; the fundus of the eye shows anæmia of the disk, but nothing more. There is no affection of the oculo-motor muscles. There is no affection of any of the other special sense organs or cerebral nerves. Pressure on the parietal bone on either side causes pain. The patient can freely move his head and neck, but is not able to raise himself from the recumbent posture. The cutaneous sensibility of the trunk, both in front and behind, is normal. The spine is not painful on percussion; but the patient complains of spontaneous pain in the lumbar portion of the spine, and also over the epigastrium.

The upper extremities show but few noteworthy changes. They can be freely moved; the muscles show considerable force, and there is no anæsthesia. The movements are co-ordinate as long as the patient keeps the eyes open; but with the eyes shut they become somewhat inco-ordinate, and the finer movements (such as buttoning the shirt) cannot be well executed. The superficial reflexes are normal; the deep reflexes over radius and triceps are present.

The lower extremities are markedly affected. When in the recumbent posture the patient can flex and extend the thigh, can also flex and extend the knee and foot. These movements however are performed slowly, owing to tendency to contractures, and feebly. He cannot raise the whole leg from the bed for more than a few seconds, and abduction and adduction of the whole leg is very feebly performed. The sensibility to tactile impressions to pain and to temperature is normal. There is however a distinct delay of the perception of tactile impressions on both feet and legs, when compared with the thighs and trunk. With the eyes closed the movements are inco-ordinate, and there is a distinct alteration of the muscular sense. The plantar reflex is present on both sides; the cremaster and abdominal reflexes are absent; the

epigastric reflex is well marked. The deep reflexes are very much exaggerated, and there is well-marked ankle clonus and the phenomenon spinal epilepsy. The electrical reactions of both upper and lower extremities are normal.

The patient cannot hold the urine well; he has to empty the bladder as soon as he has the desire to micturate, and occasionally the urine is passed involuntarily. The urine is alkaline; contain pus s.g. 1012. The bowels are confined.

Diagnosis.—From the history and present symptoms the case was looked upon as a case of syphilitic disease of the spinal cord in the lower dorsal and upper lumbar region, affecting chiefly the posterior and lateral columns. The history of the case and the presence of traces of iritis and glossitis pointed distinctly to a syphilitic affection of the cord, and the onset and symptoms pointed more to an affection of the lateral and posterior column than to a transverse myelites.

Treatment.—The patient was treated with large doses of Iodide of Potassium, and with mercurial inunction.

Progress.—For a few weeks the patient's condition remained stationary, then he improved, and on September 11th we found the following report on his condition:—Patient is now able to move his legs in any direction, he can stand erect, but walking without the help of two sticks is impossible. When he walks the gait is at once spastic and ataxic, there being the stiffness and rigidity seen in spastic paralysis, associated with the staggering gait of ataxia. With the eyes closed the patient falls at once. When in the recumbent posture the patient can move his legs very freely, when he shuts his eyes the movements become very inco-ordinate. The condition of the reflexes is the same as before. The bladder is slightly better, and the patient can hold the water a little longer than before the improvement set in. The bowels are still very much confined. The patient still complains of a pain in the epigastrium and lumbar region.

For the last few days he has been troubled with cough and expectoration; the physical examination of the chest shows extensive bronchial catarrh.

On September 17th the patient had several rigors, and a rise of temperature from normal to 102°. The temperature remained high for a few days, and the urine contained more pus. The chest symptoms improved. During the remainder of September and the whole of October the patient's condition underwent little change. Occasionally there would be a rise of temperature for a few days, ushered in by a rigor and followed by nausea and vomiting. The patient began to lose all control over his bladder, and the urine, in spite of treatment, became highly ammoniacal. The new set of symptoms were attributed to the condition of the urinary organs. The anti-syphilitic treatment was stopped, and the patient put on quinine and salicylic acid. The patient however became gradually worse, the temperature rose high, the appetite left him, he became weaker, though there was a very noteworthy change in the condition of the nervous

system, except that with the general weakness the weakness in the legs also increased. Though the patient could move the legs when in the recumbent posture, these movements were less powerful and extensive. The tendon reflexes remained in the same increased state as before, and the tendency to contractures when any movements were attempted became very marked.

On November 12th the patient had another severe rigor with considerable rise of temperature ($104\cdot5$), and died on November 15th.

The post-mortem examination was made by Dr. Harris (Pathological Registrar to the Manchester Infirmary). The most noteworthy changes were those found in connection with the urinary organs and the spinal cord. The kidneys were not enlarged, on section they showed several pale and soft spots. The pelvis of the kidney was found dilated and filled with purulent fluid, the ureters were also found dilated, the bladder was found in a state of catarrh, the mucous membrane discoloured and softened, and the contents consisted of purulent fluid.

The brain and its membranes showed no evidence of disease; portions of the cerebral cortex forming part of the motor area were placed in ammonium bichromate, and examined microscopically after having been sufficiently hardened, but showed nothing abnormal.

The several parts of the pons and medulla were carefully examined, and showed no changes.

The membranes of the cord were healthy, and the cord of normal appearance externally, and of normal consistence. Numerous transverse sections of the cord made at different levels showed already to the naked eye certain changes. The transverse sections showed from the upper cervical to the lower dorsal region a patch of degeneration, firmer than the rest of the cord, greyish in appearance, and appearing slightly depressed when compared with the surrounding structure. This patch occupied the region of Goll's tract, and below seemed to extend a little beyond that tract on each side. In the lowest dorsal region the transverse sections had a different appearance. Patches of sclerotic tissue were found occupying nearly the whole of the posterior columns, and extending over the greater part of the lateral columns, and a small patch was found in the anterior columns, corresponding to the region of Türck's tract, a smaller patch on the left, and a larger one on the right side. Successive sections showed that these lesions diminished somewhat in extent and opposite the last dorsal nerve; the sections showed a patch in the posterior columns, extending not quite to the grey matter latterly, and a patch in the lateral columns corresponding to the pyramidal tract. The affected parts in the anterior columns were here reduced to small strips close to the anterior fissure. Sections in the upper part of lumbar cord showed a still further diminution of the sclerotic patch in the posterior columns, and lower still, towards the cauda equina, the only lesion noticed was a pale greyish patch corresponding to the pyramidal tract, whilst the posterior columns appeared perfectly healthy.

The localisation of the lesions in the different sections was particularly well seen in the cord after due hardening in ammonium bichromate.

The microscopic examination of the hardened cord allowed to study more closely the exact extent and the nature of the affection. For this purpose the sections were stained both in carmine and after Weigert's method.

Throughout the whole of the cervical and upper dorsal region the affection was limited to Goll's tract, reaching posteriorly to the pia mater, which in itself was not altered, and reaching anteriorly to close but not quite up to the posterior commissure. In the lower dorsal region the lesion extended latterly to the adjacent parts of Burdach's tract, but did not reach the posterior horn, while anteriorly it extended quite up to the posterior commissure in the centre, but not at the sides.

The anterior and posterior horns appeared perfectly normal.

For the lower dorsal region the affection was of the nature of a diffuse sclerosis, and included Goll's tract and Burdach's tract, with the exception of a small strip on the inner boundary of the posterior horns; the lateral columns were affected almost in their entirety posteriorly, anteriorly the affection stopped short a little of the anterior horns, externally it reached as far as the pia mater, which here again appeared unaffected.

In the anterior columns the affection seemed limited on the right side to Türck's columns, while on the left side only a small strip, adjacent to the anterior fissure, showed a sclerotic condition. The anterior and post. grey matter showed some cell infiltration, the arteries here showed well-marked endarteritis, and by means of Weigert's method changes in the ganglia cells and nerve fibres in Clark's columns were made evident.

In the lowest dorsal and upper lumbar region the affection corresponded closely as regards extent to a postero-lateral sclerosis. In the posterior columns Goll's and Burdach's tracts were affected; but it was noted that the affection did not extend beyond a line drawn vertically from the innermost part of the substantia gelatinosa to the posterior periphery, whilst the changes in the lateral columns seemed entirely limited to the region of the pyramidal tract. The affection in the anterior column was only apparent on the right side, in the form of a small stripe close to the anterior fissures, the left anterior column appeared normal. The grey matter was affected similarly as in the lower dorsal region, as stated above.

For the lower lumbar region the affection was entirely limited to a small triangular patch on the outer and posterior part of the lateral columns, whilst the posterior columns, the anterior column and the grey matter appeared fairly healthy, with the exception of slight cellular infiltration and enteritis.

The pia mater, the posterior and interior roots appeared healthy throughout the length of the cord.

As regards the histological changes, we have to notice numerous blood-vessels with thickened walls, the thickness depending in

part on an endarteritis, which in some places caused marked diminution of the lumen of the vessels, and in part on an infiltration of the perivascular sheath. Round the vessels were noticed masses of round and spindle cells, arranged in some spots concentrically round the blood-vessel. The rest of the altered tissue showed numerous nuclei, tracts of fibrous tissue, thick and coarse, fine granular matter, a sparing number of Deiters' cells. The nerve fibres showed the usual changes, hypertrophied axis-cylinder nerve fibres, in which the white substance had taken the carmine-staining and atrophied nerve fibres. In the grey matter the blood-vessels showed likewise endarteritic changes and perivascular infiltration. Besides these a large number of nuclei and fine granules were seen in the neighbourhood of the ganglia cells. The cells themselves (with the exception of those in Clarke's columns) showed but little alteration; some of the cells in the anterior horns however had a hyaline appearance, in others the nuclei had not taken the staining, and others had their cell processes broken or shrunk.

The case presents several points of interest.

The first question which arises refers to the nature of the affection of the cord. Have we here a syphilitic inflammation, or is the sclerosis (for such the naked eye and microscopic appearance declare it to be), though etiologically connected with syphilis, yet not itself of a syphilitic nature? Does it stand in the same relation to the syphilis as locomotor ataxy coming on in a syphilitic subject? Though the question cannot be answered positively, yet there are several factors which point to the lesion as of syphilitic nature. Thus the symptoms came on soon after the syphilitic affection, whilst in most cases of locomotor ataxy syphilis has preceded the ataxic symptoms by a much longer period of time; the symptoms also yielded at first to an anti-syphilitic treatment, whilst such a treatment has no effect in the largest majority of cases of locomotor ataxy.

The anatomical structure likewise presents some peculiarities speaking in favour of syphilis. Thus in a non-syphilitic affection we should expect at so early a period after the onset (for the disease lasted scarcely twelve months in our patient) to find an appearance corresponding more to myelites than to sclerosis. The histological examination showed extensive endarteritis and perarteritis, and though this can no longer be considered as characteristic of syphilis, yet it has, as regards the nervous system, been found most markedly in syphilitic sclerosis. Thus Rumpf¹ gives a case of diffuse syphilitic infiltration of the cord where the histological

¹ 'Die Syphilitischen Erkrankungen des Nervensystems,' p. 341, and 'Arch. f. Psychiatric,' vol. xvi. p. 410

changes were very similar to those found in our case; Greiff¹ describes a case of syphilis with paresis of the lower extremities where in the vessels well-marked obliterating endarteritis was found. Ballet and Minor² describe a case of diffuse sclerosis in a syphilitic subject which, both as regards the histological changes and in many respects also as regards the localisation, resembles our case, and draw special attention to the vascular changes which cause them to distinguish between a perivascular sclerosis and a peritubular sclerosis (seen in the ordinary primary inflammation of the cord, and for which the term parenchymatous inflammation would perhaps be better). Savard³ likewise draws attention to the vascular changes, and to the cell infiltration and poliferation in syphilitic diseases of the cord. I could besides cite Charcot and Gombault,⁴ Homolle,⁵ Leyden,⁶ Westphal,⁷ and Schulze.⁸ We have however to bear in mind, that similar changes are found in the non-syphilitic form of sclerosis, especially in acute and chronic myelitis, and I have seen repeatedly very marked endarteritis in otherwise healthy cords of persons who have died from granular kidney; as was long ago pointed out by Gull and Sutton. In many of the recorded cases of syphilitic sclerosis the inflammatory process affected the pia mater also; this, however, was not so in our cases, where the pia mater, as already stated, was perfectly intact.

Another feature of the lesion was its diffuse character; it affected not only the white matter, but also, though to a much smaller extent, the grey matter of the cord. This has also been found in some of the cases of syphilitic myelitis, especially in the case of Rumpf cited above; yet such a diffuse infiltration occurs also in the ordinary non-specific form of myelitis.

From these considerations it is evident that, though the syphilitic nature of this infiltration is highly probable, yet we cannot speak on this point very positively. The detection of the syphilis bacillus might in these cases materially assist us in our differential diagnosis, but as yet our methods of staining these bacilli cannot be said to be satisfactory, and as they are but sparingly found even in primary syphilitic sores, it is not very likely that our efforts to detect them in syphilitic

¹ 'Arch. f. Psych.,' vol. xii. p. 564.

² 'Arch. de Neurologie,' vol. vii. p. 44.

³ 'Etude sur les myélites syphilitiques,' 'Thèse de Paris,' 1882.

⁴ 'Arch. de Phys.,' 1873, p. 143.

⁵ 'Progrès Méd.,' 1876.

⁶ 'Charité Annalen,' 1876, p. 260.

⁷ 'Arch. f. Psychiatrie,' vol. vi. p. 245.

⁸ 'Ibid.,' vol. viii. p. 222.

infiltrations of the cord would be very successful. Unfortunately the cord from our case had been kept in ammonium bichromate for some time, which would render an examination for the bacillus still more difficult, if not altogether impossible, for it is doubtful whether the method by means of which we can detect the tubercle bacillus in tissues hardened in ammonium bichromate is also applicable for the bacillus found in syphilitic lesions.

Our case presents another point of interest. The affection in the posterior columns, especially in the upper lumbar region, did not extend beyond a line drawn vertically from the substantia gelatinosa to the posterior border of the cord; the case thus supports Westphal's observations on the persistence of the knee tendon reflex in posterior sclerosis in such cases, and we can thus understand its presence in our case, and its exaggeration owing to the implication of the lateral columns.

If we sum up our case briefly, we have a patient who soon after having been affected with syphilis shows symptoms of ataxic paraplegia with affection of the bladder, and as the cause of this we find a diffuse sclerosis of the lower dorsal region of the spinal cord affecting for a short extent the anterior lateral and posterior columns, and limiting itself in the lowest dorsal and upper lumbar region to the Goll's and Burdach's tracts in the posterior columns, the indirect pyramidal and direct cerebellar tract (as far as they extend) in the lateral columns, and the cells and fibres in Clarke's columns.

The changes in the upper dorsal and cervical region we are inclined to look upon as secondary ascending, those in the pyramidal tract in the lower lumbar regions as secondary descending degeneration. The histological examination of these parts showed a somewhat different appearance when compared with that in the lower dorsal region: the vascular and perivascular changes were much less marked, the neuroglia showed a larger quantity of granules, but fewer nuclei, and many of the axis-cylinders in the affected zone had still a normal appearance.

Cases like the one given are by no means rare, but they have been mostly described as postero-lateral sclerosis, and have been looked upon as combination of several systemic affections, *i.e.* of Goll's tract with the adjacent portions of Burdach's tract, the pyramidal tract, and cerebellar tract.

That such combined affections do exist can no longer be doubted. The cases related by Strumpell,¹ Westphal,² Kahler,

¹ 'Arch. f. Psych.', vol. xi. p. 27, and vol. xvii. p. 217.

² Ibid., vol. xv. p. 224.

and Pick,¹ and more recently by Babinsky,² and several others, show this; such cases are, however, by no means common. Clinically they are chiefly characterised either by spastic paraplegia, without implication of the bladder and rectum, and without many sensory disturbances, like one of the cases recorded by Strumpell, or we have symptoms characteristic of *tabes dorsalis* (*i.e.* inco-ordination, absence of tendon reflexes, and sensory disturbances, etc.), together with marked paralysis and contractures.

A number of other cases, however, have been classed by some authors, such as Grasset,³ who has collected no less than thirty-three fatal cases, under the heading of Combined Tabes, which included many cases like ours, of diffuse sclerosis.

We are inclined to agree with Ballet and Minor,⁴ who speak of these cases as false combined systemic sclerosis. A glance at the post-mortem appearances, as described by some of the authors, shows at once that the lesions found are by no means always the same. The posterior columns are affected in a very varying degree, both as regards lateral and vertical extension; in some cases, like those of Zacher⁵ and Déjérine,⁶ and both Goll's and Burdach's tracts were affected, at least in some regions of the spinal cord, whilst in others (Strumpell, Westphal) the lesion in the post. columns was almost entirely limited to Goll's tracts. The pyramidal tract also showed by no means the uniform well-defined lesion as seen *f. i.* in amyotrophic lateral sclerosis or secondary descending sclerosis; the direct pyramidal tract in the anterior columns has but rarely been found affected, and the cerebellar tracts, while distinctly affected in some cases (Strumpell, Kahler and Pick, Westphal), were found intact in others. In some cases moreover the pia mater showed signs of inflammation.

With such a variability of the lesions, it is not astonishing that the symptoms of the so-called postero-lateral sclerosis should be so diverse.

In some cases⁷ we have the symptoms of locomotor ataxy, to which are added more or less pronounced paralysis and contractures; in others (like those of Strumpell cited above) we have almost pure spastic spinal paralysis, with scarcely any sensory troubles, and with increased tendon reflexes; in some, bladder and rectum are affected early in the disease, in others these

¹ 'Arch. f. Psych.,' vol. viii. p. 251, etc.

² 'Revue de Médecine,' 1887.

³ 'Arch. de Neurolog.,' vols. xi. and xii.

⁴ *Ibid.*, vol. vii. p. 44.

⁵ 'Arch. f. Psych.,' vol. xv. p. 405.

⁶ 'Arch. de Phys.,' 1884, iv.

⁷ Kahler and Pick, 'Arch. f. Psych.,' vol. viii. p. 252; Westphal, 'Arch. f. Psych.,' vol. viii. p. 470.

organs may remain unaffected, or become only slightly affected towards the end.

We can thus no longer speak of one form of combined postero-lateral sclerosis, but must admit, both from the clinical and pathological standpoint, several forms; those resembling *tabes dorsalis* and those resembling *Friedreich's disease*,¹ and those resembling *primary lateral sclerosis*.²

There remain however still a large number of other cases, which, like the one given above, are due to a diffuse sclerosis. That the symptoms in these cases should vary in the individual cases is easily understood, and be dependent on the situation and extent of the sclerosis in the posterior and lateral columns respectively. These cases then, clinically considered, are difficult to distinguish from cases of pure combined systemic sclerosis except by their history and course, such as the gradually developing paresis with contractures and the presence of bladder and rectum troubles. In many cases the upper extremities remain free, and show neither ataxic nor paretic symptoms.

Pathologically the chief distinction lies in the vascular and perivascular changes, and the marked increase of the fibrous element in the neuroglia, whilst the nerve-fibres show still a fair number of normal or simply atrophied axis-cylinders. The process in some cases can be traced from the pia mater, which is found thickened and altered by inflammatory deposits.

¹ Kahler and Pick; Schulze.

² Strümpell.