REMARKS

ON

THE PATHOLOGICAL CHANGES IN A CASE OF
CHRONIC SYPHILITIC SPINAL
PARALYSIS.

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SYPHILITIC disease of the spinal cord may give rise to various symptoms, according to the nature of the pathological changes. The symptoms may be those of acute myelitis, of tumour (gumma), of meningitis, of meningo-myelitis, of pseudo-tabes, and other forms of spinal syphilis have been described. Professor Erb¹ has specially drawn attention to a group of cases of chronic syphilitic disease of the spinal cord which present a certain combination of symptoms. To these cases the name of "Erb's syphilitic spinal paralysis" is frequently given. The patient presents superficially the familiar symptoms of spastic paresis or paralysis, as regards gait, attitude, and movements. The patella tendon reflexes are increased, and ankle clonus is present, but there is relatively only slight muscular rigidity. The bladder is constantly affected. As a rule there is only slight, though constant, affection of sensation. The onset of the disease is generally gradual, seldom rapid. Marked spastic paresis gradually develops, but only exceptionally is there complete paraplegia. The upper half of the body is unaffected. There is often a tendency to improvement under antisiphilitic treatment.

A large number of cases have been recorded which have presented the combination of symptoms to which Erb has drawn attention; but there has been much discussion as to whether these symptoms ought to be regarded as characteristic of spinal syphilis, and only a few pathological examinations have been made.

Oppenheim² admits the frequency of Erb's group of symptoms, but believes that they are simply due to a dorsal myelitis of syphilitic origin, not extending over the whole transverse section of the cord. He believes that there has been a meningo-myelitis in the dorsal region, but that the meningeal affection has subsided, and only the results of the myelitis and secondary degeneration have been left behind.

Whatever opinions may be held as to the advisability of regarding Erb's group of symptoms as those of a special affection, there can be no doubt that cases of syphilitic disease of the spinal cord are not infrequently met with, which present this combination of symptoms. Hitherto only a very few pathological examinations have been made in this form of spinal syphilis, and hence the records of the pathological changes in the following case, which clinically corresponded in most points with Erb's type, may be of interest. During life the patient was under the care of Dr. Joseph, Honorary Surgeon to the Warrington Infirmary, and the abstract of the clinical history is taken from notes kindly furnished by Dr. Langdale, House-Surgeon to the hospital. Dr. Langdale also made the *post-mortem* examination, and to both of these gentlemen I am indebted for kindly forwarding the spinal cord to me for examination.

CLINICAL HISTORY.

The patient was a man, aged 27, labourer, who first came under treatment in November, 1894. He had been a soldier, and had contracted syphilis in 1888, at the age of 21. In 1893 he began to be troubled with difficulty in micturition. This increased, complete retention occurred, and for almost twelve months before he first came under observation he had been obliged to use a catheter to draw off the urine.

When first examined, in 1894, he complained of retention of urine, of weakness of the legs, and of a tired feeling at the back of the knees on going upstairs. There was at that time no definite paralysis or wasting of the muscles of the legs. The plantar reflexes and the knee-jerks were increased; ankle clonus was present on the right side, doubtful on the left; the gluteal, cremasteric, and abdominal reflexes were present. There was no anesthesia, no pain in the back, no girdle sensation and no inco-ordination. The pupils reacted normally, and the optic discs were normal.

Ten months later (in August, 1895) he began to suffer from pain in the back, especially at the lower part. In November, 1895, the legs were so weak that he was unable to walk without assistance. The movements of the left leg were greatly impaired, those of the right leg to a less extent. There was still complete retention of urine. The knee-jerks were increased, and ankle clonus was present on both sides. On the right leg,

and on the abdomen on both sides as high as the umbilicus, there was slight impairment of cutaneous sensibility; on the left leg sensation was normal. The urine contained pus. After rest in bed the condition of the bladder improved, and he was able to pass urine without the aid of a catheter. The legs also improved rapidly, and the patient was able to walk again, but the gait was unsteady. Sensation became quite normal.

In December, 1896, the patient again came under observation. There was then some difficulty in micturition. He was able to walk, but there was inco-ordination of the lower limbs. At a later date there was obstinate vomiting and hiccough, and the urine contained pus. Death occurred in February, 1898.

The *post-mortem* examination was made by Dr. Langdale, who kindly sent me the spinal cord and portions of the brain for examination. The necropsy revealed extensive double pyelonephritis, but no other macroscopic changes of interest were detected. The cerebrum, cerebellum, pons and medulla appeared normal macroscopically.

PATHOLOGICAL EXAMINATION.

The spinal cord was hardened in Müller fluid; numerous pieces were embedded in celloidin. Sections were prepared and stained according to Weigert's method, and with logwood and eosin, and with aniline blue black. Diagram 1 represents the distribution of the changes in the spinal cord at various levels. The portions of the figures in this diagram which are shaded by dots represent the regions of degeneration and secondary ascending and descending sclerosis—that is, the parts in which, in the sections stained according to Weigert's method, nerve fibres are absent or scanty.

MICROSCOPICAL EXAMINATION.

In the pia mater and arachnoid evidences of slight meningitis were found in all regions of the cord, but the changes were most marked in the dorsal region, least marked in the cervical region. The meninges were slightly thickened by fibrous tissue, and in various parts there was slight infiltration with round cells chiefly around the blood vessels.

Diagram 1.

Diagram 2.

Diagram 3.

Diagram 4.

Diagram 1.—Showing distribution of lesions in the spinal cord at various levels. The sclerotic and degenerated parts are shaded. Diagram 2.—Section of meningeal artery showing endarteritis. Diagram 3.—Section of small vessel in the white matter of the cord showing hyaline thickening of its walls. Diagram 4.—Section of spinal cord in upper dorsal region. Weigert's stain. Pale parts=regions of sclerosis. Black dots=normal nerve fibres.

The spinal blood vessels presented well-marked changes in numerous parts. The larger arteries in the meninges, especially at the posterior part of the cord, near the posterior nerve roots, often presented marked endarteritis. The internal coat was often much thickened by abundant new cell formation on the inner side of the elastic lamina, the lumen of the vessel being thereby much diminished (see Diagram 2). The

middle and external coats were generally not much altered; but frequently there was an infiltration of leucocytes around the vessels. The veins were either unaffected or the changes (proliferation of the intima cells) were less than in the arteries. In many places the veins were surrounded by cell infiltration. The very small meningeal arteries presented thickening of the walls and slight proliferation of the cells of the intima, and they were often surrounded by round-celled infiltration.

The meningeal vessels were generally well filled with red corpuscles, but in a few places the lumen of a vessel was partially occluded by a thrombus.

Within the cord, especially in the diseased parts indicated on p. 1922, the walls of the small vessels were often very much thickened, and had a homogeneous hyaline appearance, also the endothelial cells of the intima were slightly proliferated (see Diagram 3). In some places these hyaline vessels were surrounded by slight infiltration of leucocytes.

The two prominent features of the vascular changes were the endarteritis of the larger meningeal arteries and the hyaline thickening of the walls of the small vessels within the cord.

The changes in the white matter will be best indicated by describing the sections at various levels.

In the cervical region there was marked degeneration in the posterior median columns, and the degenerated region presented the usual appearances of secondary sclerosis, namely, absence of nerve fibres and increase of neuroglia connective tissue. The pia mater was slightly thickened by fibrous tissue, and in the lateral region of the cord, just beneath the pia mater, in the region of the direct cerebellar tract, and a short distance in front of it, there was an increase in the neuroglia connective tissue and a diminution of the number of nerve fibres. But this peripheral sclerotic change was very slight, and did not extend to the crossed pyramidal tract. The last mentioned tract and all other parts of the white matter appeared normal. The grey matter was normal.

In the upper dorsal region marked degeneration was seen in the posterior columns, localised to the posterior median column and a little of the adjacent part of each posterior external column. The sclerosis at the anterior part of this region—that is, close to the grey commissure—was slight. In the lateral region of the cord there was a peripheral strip of degeneration just under the thickened pia mater. The degeneration extended inwards, and affected the outer part of each crossed pyramidal tract (see second figure, Diagram 1, and also Diagram 4). The degenerated parts, both in the posterior and lateral regions, presented the ordinary appearances of secondary sclerosis—namely, disappearance or diminution of the nerve fibres and great excess of neuroglia connective tissue with dilated blood vessels.

The sclerotic changes were more marked in the lateral column of the right side, and at one point on this side there was a gummatous infiltration. This commenced from the pia mater and surface of the cord, and extended inwards as an irregular, somewhat oval mass, just anterior to the position of the crossed pyramidal tract (see Diagram 5). In the centre

Diagram 5.—Representing section stained with logwood and eosin. Gummatous mass surrounded by cell infiltration, extending inwards from the surface of the cord. Upper dorsal region.

of the patch was a small mass of amorphous caseous substance. This was surrounded by nucleated fibrous tissue, and externally was a zone of abundant cell infiltration. The small vessels presented marked hyaline thickening of their walls. In some of the sections this gummatous infiltration was seen to invade the grey matter slightly.

In the mid-dorsal region there was a similar distribution

of the peripheral sclerosis in the lateral columns (as in Diagram 4). But in the posterior columns the intense sclerosis in the region of the posterior median columns had disappeared. There was, however, a slight diffuse increase of the neuroglia fibrous tissue all over the posterior columns, both external and median. The neuroglia was well supplied with nuclei and the small blood vessels were dilated. Nerve fibres were seen at all parts of the posterior columns in this region, but their number was much less than in a healthy spinal cord, and they were more widely separated by neuroglia connective tissue. The grey matter was normal.

In the lower dorsal region there was a small degenerated sclerotic patch in the posterior columns adjacent to each posterior horn of grey matter (as shown in the third figure of Diagram 1). The one on the left side implicated the grey matter of the posterior horn. There was also a peripheral zone of sclerosis in each lateral region. At a lower level only one of the sclerotic patches in the posterior column was seen (as shown in the fourth figure of Diagram 1). In the lowest dorsal region, just above the lumbar enlargement, there was a large patch of degeneration in the posterior columns of the left side. It extended from the posterior median fissure up to the posterior horn of grey matter (as shown in the fifth figure of Diagram 1 and also in Diagram 6). In this degener-

Diagram 6.—Section of spinal cord in lowest dorsal region (Weigert's stain); affected parts pale; normal nerve fibres represented by black dots.

ated patch, sections stained according to Weigert's method showed a complete absence of nerve fibres. The periphery of the patch was bounded by a broad zone of dense fibrous tissue. Internal to this zone was a region in which there were a considerable number of blood vessels with hyaline walls, surrounded by a network of well-nucleated fibrous tissue. In the centre of the patch were numerous tortuous blood vessels, presenting marked hyaline degeneration of the walls, and surrounded by round-celled infiltration. This cell infiltration varied in amount at different parts; it consisted of leucocytes, together with a few compound granular cells, but none of the large epithelioid-like cells, so common in acute non-syphilitic myelitis, could be detected. In the lateral part of the cord at this level there was the same peripheral zone of degeneration (Diagram 6) as in the mid-dorsal region.

In the lumbar region of the cord the only change, apart from the affection of the meninges and their vessels (described above), was sclerosis, just limited to the region of the crossed pyramidal tracts (as shown in the last figure of Diagram 1).

Grey Matter of the Cord.—As mentioned above, at one point in the lower dorsal region a sclerotic patch invaded the whole of the posterior horn of grey matter; also at one spot in the upper dorsal region the outer half of the grey matter was invaded by gummatous infiltration on the right side; but in all other parts of the cord the grey matter was normal, with the exception of dilated and diseased blood vessels. The nerve cells and fine nerve fibres of the grey matter seen in Weigert's specimens were everywhere normal, except at the points just mentioned. The columns of Clarke appeared normal.

As regard the distribution of the changes above described, it is evident that the degeneration in the posterior median columns in the cervical region was of the nature of secondary ascending sclerosis; it was less extensive than that in the posterior columns of the upper dorsal region. The degenera-

tive changes in the lateral region of the cord were evidently due to a sclerosis produced by the extension inwards of inflammatory changes beginning in the pia mater. As mentioned above, they affected the peripheral region of the cord, and also implicated the crossed pyramidal tract. In the lumbar region, however, there was distinct secondary descending sclerosis, exactly limited to the crossed pyramidal tract.

A prominent feature of the pathological changes was the endarteritis of the larger vessels, especially in the meninges and the hyaline degeneration of the smaller vessels. All the degenerated portions of the cord presented sclerotic changes except those in the posterior columns in the lowest dorsal region, and at one point in the antero-lateral columns on the right side in the upper dorsal region. Here the changes were those of a gummatous infiltration.

The pathological changes were certainly not those of disseminated sclerosis; their distribution was not sufficiently irregular; there was also marked secondary ascending sclerosis in the cervical region, which is very rare in disseminated sclerosis; and during life the symptoms were not those of the disease just mentioned. The typical endarteritis, the slight meningeal changes, the extension of the lesion from the meninges into the adjacent parts of the cord, the histological (gummatous) nature of the most recent patches, and the syphilitic history of the patient, all indicate clearly that the case was one of spinal syphilis.

The pathological and microscopical examinations of the spinal cord, which have hitherto been recorded in cases of Erb's syphilitic spinal paralysis, are too few to enable one to draw definite conclusions as to the lesions, and possibly they may not always be of the same nature and distribution. But the above record furnishes a good example of the changes which may be met with, in a case which presented during life most of the symptoms of "Erb's syphilitic spinal paralysis." The chief point of difference from Erb's cases was the gait; there was considerable inco-ordination in the movements of the legs; but this has been recorded in other cases also.

Briefly the pathological changes may be summarised as follows: Endarteritis and hyaline degeneration of the arteries of the spinal cord and meninges; slight meningitis; gummatous infiltration of the right antero-lateral columns in the upper dorsal region; sclerosis of the periphery of the cord in the lateral columns in the whole of the dorsal region; sclerosis in the posterior median columns in the upper dorsal region; irregular sclerotic patches, with one patch of cell infiltration (gummatous) in the lowest dorsal region; descending sclerosis in the lumbar crossed pyramidal tracts; ascending sclerosis in the cervical posterior median columns.

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A CASE OF SYRINGOMYELIA.

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It is only within the last few years that the possibility of giving a clinical picture of syringomyelia has been recognised. The presence of abnormal cavities in the spinal cord was first recorded by Morgagni¹ nearly two centuries ago, and these for long were looked upon as mere pathological curiosities. In later years, however, the pathology of this condition has been pretty clearly established, and now syringomyelia is recognised as having a definite symptomatology, and the striking statement is made by Starr² that "it is not less common than such a well-known disease as disseminated sclerosis," and Charcot agreed with Schultze that "it was at least as common as amyotrophic lateral sclerosis." The rapidity with which this advance has been made in our knowledge of the disease is strikingly brought out by comparing the well-established group of symptoms now regarded as diagnostic, and described in detail by Bruhl,⁴ with the statement made by Erb, even in 1878, that "all experience thus far teaches that

"the development of cavities, as such, produces no sort of symptoms by which it could be recognised during life"; and again, that "no special symptom, no peculiarity in the course of the disease, betrays the complication."³ The following case is instructive, especially as it illustrates some of the variations from the classical type that may be met with, and I have Sir William T. Gairdner's kind permission to publish it in detail:

J. B., aged 33, a farm servant, was admitted to Sir William Gairdner's ward in the Glasgow Western Infirmary on February 9th, 1898, suffering from a chronic nervous illness, the most prominent features of which were wasting of the right hand and forearm, marked spastic paraplegia, and some sensory disturbance.

History.—As a child he was healthy, the only illness he remembers having had being measles. He had no convulsions, but at the age of 10 he says he was nervous and easily frightened, though not prevented from attending school. At the age of 12 he left school and began to work on a farm, and two years later, during a severe winter, he suffered much from fissures on his hands, which were long in healing and more troublesome on the right hand. From this time the right hand was noticed to be weak, and he began in his work to use the left hand more than the right. During the following winters he suffered much from chilblains on his hands (right more than left), and in the spring of 1883 (aged 18) he had a whitlow on the ring finger of the right hand, which took a long time to heal. The weakness of the right hand meanwhile was gradually increasing, and the muscles were becoming atrophied. In July, 1883, he suffered much from vertical and occipital headache, and also had indigestion with occasional vomiting, and a gnawing pain in the back between the shoulders. He went on with his work till towards the end of the month, when for a few days the headache was very severe, and he could not bear the light on his eyes, and was sleepless. One day at this time, when walking in the garden, he suddenly got dizzy and fell, but without hurt or loss of consciousness, and he was able to get up and walk into the house. Next morning he fell down in the house, and had to be lifted into bed, a movement of which he was quite unconscious. He remembers being out of bed again by the time the doctor arrived, and also that he was out of bed a day or two later, having his head shaved and a blister applied. However, within a week he was completely paralysed, and was wholly confined to bed for fully a month. At first he could not move arms, legs, or back, but gradually recovery took place, more in arms and back than in legs, so that he was soon able to get out of bed, and walk with the aid of sticks. It should be added that he suffered from retention of urine during the first fortnight he was confined to bed, but after that there were urinary difficulties, though never complete retention. After this headache was occasional, and he often felt a sharp pain in the right occipital region, but he practically suffered no pain apart from this headache. He came to the Western Infirmary in July, 1884, and remained under treatment for about five months. His case at this time was indexed "spastic paraplegia," there being noted in addition some permanent inequality of the pupils, atrophy and weakness of the right arm, and some sensory disturbance. Very little change took place during his residence, and soon after leaving he proceeded to the University College Hospital, London, where he was under the care of Sir William Gowers. Since then his general condition has not changed much. He has not been able to do any regular work. His right arm has gradually been getting more wasted and weaker, and latterly there have been some subjective sensory disturbances in the left hand, which is getting more susceptible to cold, and seems to be getting slightly weak, and also wasted. The spastic condition of the lower limbs and the urinary difficulties have remained much the same. In August, 1897, the patient was in the Glasgow Royal Infirmary.

Family History.—His parents are both dead, the father at age of 67, after an attack of paralysis of the right side; the mother at the age of 66, from influenza with pulmonary complications. He is one of a family of three sons, his two brothers being alive and well. No relation is known to have suffered from any nervous illness.

Present State.—The patient is a healthy-looking man. There is some oedema of the feet and legs. There is slight postero-lateral curvature of the spine in the upper dorsal region, the convexity being to the right. There is a slight want of symmetry between the two sides of the face, but no relative difference in the power of the facial muscles can be appreciated. There is marked paresis of the right arm, especially of the hand, but the grasp of the left hand is probably also deficient. With the dynamometer the grasp of the right hand is registered as 10, and that of the left as 45. There is considerable atrophy of the right hand and forearm as compared with the left. The thenar and hypothenar eminences are practically gone, and the interosseal muscles very deficient. There is characteristic deficiency in the movements of the hand. He cannot oppose the thumb to any of the fingers, and when he tries to fully extend the fingers there is over-extension of the first phalanges and flexion of the terminal ones. There is distinct though less marked wasting of the thenar and hypothenar eminences of the left hand, and this is associated with an inability to oppose the left thumb to the little finger. No relative wasting is made out in the shoulder muscles. Fibrillary tremor is present in the muscles of both hands. There is no rigidity of the upper limbs, though there is some tendency to contraction of the third, fourth, and fifth fingers of the right hand. No triceps, biceps, or wrist jerks are got on either arm. There is paresis of both legs, slightly more marked on the right side. Secondary rigidity is very marked, and the attitude of the limbs characteristic. The feet are extended at the ankles, and the first phalanges of the toes are over-extended, while the terminal phalanges are fully flexed. The great toes are specially affected in this way, that of the left foot being quite dislocated at the metatarsophalangeal joint. The muscles of the right leg seem to be less bulky than those of the left, and at the thickest part of the calf, though the circumferential measurement varies to some extent with the oedema, the right is always one-half to one inch less than the left.

The knee-jerks and ankle clonus are exaggerated on both sides, and the plantar reflexes are very distinct. The epigastric, abdominal, and cremasteric reflexes are not got. There are no tremors of the limbs.

There is no evidence of trophic change in the skin or its appendages