

## Clinical Cases.

### CASE OF CERVICAL MENINGO-MYELITIS: EPILEPTIFORM SEIZURES: ATROPHY OF NERVE-ROOTS AND MUSCLES: UNUSUAL FORM OF ASCENDING DEGENERATION IN THE SPINAL CORD.

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CHARLES H—, \* aged 32, was admitted on October 22nd, 1881, into the National Hospital, under the care of Dr. Ramskill, who has kindly permitted me to publish the case. He had led a very gay life, and had several chancres as well as epididymitis, but a history of the usual symptoms of syphilis could not be obtained. In May 1881, he had "neuralgic" pains in the left eye, and then "rheumatic" pains in the shoulders. In June he noticed that he saw double, the diplopia gradually improving and disappearing before admission. Soon the fourth and fifth fingers of the left hand became weak, so that he could not extend them. In August he gradually lost power in the left leg, and before long could not walk at all, owing to nearly complete paralysis of the lower extremities. In a few days sensibility had departed from the lower limbs, and from the trunk below the waist, at which level there was a sense of constriction. A little later the right arm became powerless. There was involuntary evacuation of both urine and fæces during three weeks, but this symptom had improved when he was first seen by Dr. Ramskill shortly before admission.

When admitted, he was a good deal emaciated. The lower limbs were much wasted and the muscles soft and flabby. He could just manage to draw up the legs a little, the right better than the left; could flex the knees and ankles, but not extend them; could not move his toes. The legs were not

\* The early notes of this case were taken by Dr. C. E. Beevor, late Resident Medical Officer.

rigid, but were kept in a flexed position, to which they returned when straightened. The reflexes were as follows: plantar, both obtained, but with some difficulty; ankle-clonus obtained, right and left, but not very readily; knee-jerk, both excessive; cremasteric, abdominal and epigastric, absent.

The upper limbs were like the lower, much emaciated. The left arm was much more wasted than the right; the posterior aspect of the left fore-arm being positively concave: the difference between the right and left hands was equally distinct. While there was free movement at all the joints of the right arm, the power over the left was very much diminished; extension at the elbow and all movement of the fingers being quite absent, while the other actions were very feebly performed. As tested with the dynamometer, the right hand possessed a grasping power of 25 lbs., the left of 0 lb.

As regards sensibility, he could localise tactile impressions correctly on the left foot and leg and on the right leg, but less correctly when touched on the right foot. The sensibility of the rest of the body was good. There was no analgesia. There was no tenderness over, or irregularity in, the course of the spine. He had to obey the call to pass his urine immediately he felt the desire, and his bowels were constipated. The urine was healthy.

He was unable to turn the left eye outwards beyond the middle line, but there was no other abnormality of head or face, and no optic neuritis. There was slight nocturnal elevation of temperature, probably due to an extensive bed sore which exposed a large portion of the sacrum.

*Progress of the Case.*—The patient suffered greatly from his bed sore, but after a time began to improve. His legs remained drawn up at the knees, at which point they were pressed together with such force as to cause him great pain.

On March 6th, 1882, he could for the first time lift both feet clear of the bed, and his right hand was much stronger. The paralysis of the left external rectus had disappeared some time, and the large bed sore had nearly healed. His left arm was kept habitually in a position of extreme abduction at the shoulder, flexion at the elbow, and dorso-flexion at the wrist.

On May 4th he became suddenly much worse, was delirious, and vomited three times. His temperature rose to 105° and his pulse to 150. There was no optic neuritis at this or at any time. Tremors and spasm of the legs were incessant and caused him to shriek out. The explanation of his altered condition became evident in a few days, when a large bed sore made itself visible through the thick skin of his left heel, and shortly afterwards a large portion of the os calcis

became bare. From May 6th until the day of his death, any movement of the right, that is, the non-paralysed arm, caused him excessive pain. On the 12th there was subsultus, and occasional delirium.

On the 13th he was attacked with three fits, which are of interest, on account of the fact that the only lesion discoverable at the autopsy was spinal. The first fit occurred at 7.30 A.M., and lasted a few minutes. It affected at first only the left arm and leg, and the left side of the face, and later the right side of the face and right arm and leg. The head and eyes at first were turned to the left, later to the right. The second fit lasted also a few minutes, and was exactly the counterpart of the previous one, with the difference that it began on the right side and terminated on the left. The above facts I wrote down, as they were given to me by an attendant on whose correctness of observation I can rely. Consciousness was retained during the second fit, but not during the first. When I first saw him, at 8.30 A.M., the third fit had been half an hour in progress. I took the following note: "Patient is lying on his back; head turned to the left; eyes open. There are small clonic spasms of the eyes from right to left; also of the mouth, chiefly on the left side; also of both fore-arms, both of which are nearly completely flexed. The fingers of both hands are semiflexed and rather rigid, but not affected by clonic spasm. The right leg is not at all affected. There are clonic spasms of the left foot from the ordinary position to that of dorso-flexion, and the leg is so thin that the *tibialis anticus* and *extensor longus digitorum* can be seen to contract with each spasm. The respirations are 120 per minute, and very noisy: the breathing is entirely abdominal. The pulse is 120 and very weak. The pupils are much dilated and equal. The face is sweating and very flushed. He is not unconscious."

Two hours later I continued my note: "Is still in the same epileptiform convulsion. Is conscious. The muscles affected are the right sterno-mastoid, the face being turned upwards and to the left, the levators of the eyelids, the left external rectus and the right internal, the flexors of the left thigh, the hamstrings on the same side, and the dorso-flexors of the left foot. All these muscles are subject to clonic spasm, occurring at the slow rate of two a second, as often seen at the end of an ordinary epileptic seizure. The left *transversalis abdominis* is affected, but not the diaphragm. The respirations are now at the rate of forty a minute." This fit lasted until 12.30 P.M., that is, four and a half hours.

The patient continued in the typhoid condition, and in two or three days developed signs of pneumonia. He died on May 19th. The treatment employed by Dr. Ramskill con-

sisted in giving large doses of iodide of potassium, and nerve tonics.

The following are the chief results of the post-mortem examination, which was made 12 hours after death. Rigor mortis, not well developed; body much emaciated. There is extreme atrophy of the muscles of the left hand and arm, and small portions of muscle removed from the left thenar eminence and the extensor surface of the left fore-arm are very pale compared with corresponding specimens from the right arm. The spinal cord is removed without any difficulty, although there are slight adhesions on the left side between the dura and the vertebrae in the cervical region. The dura mater is a good deal thicker than normal in the region of the cervical enlargement. Here over a vertical extent of about three inches the arachnoid and subarachnoid cavities are obliterated by delicate fibrous tissue, chiefly placed laterally around the roots of the nerves, the obliteration being much more complete on the left side than on the right. It is difficult also, both anteriorly and posteriorly, to separate the dura mater from the cord. Both the dura mater and the adhesions are perfectly pale. More exactly the adhesions between the dura mater and the pia extend to the left side from the level of the fifth cervical to that of the second dorsal nerve-roots. The posterior roots on the left side of the nerves between the fifth cervical and second dorsal inclusive are markedly atrophied. The anterior roots of the fifth and sixth cervical nerves are very distinctly atrophied, being hardly more than mere threads. The atrophied nerve-roots are of a dirty-grey colour. On cutting consecutive transverse sections of the cord, the whole of the cervical region, except the highest part, is found to be softer than normal, while over the central and lower two or three inches its substance is nearly fluid, of about the consistence of cream, and of a dirty-white colour. The softening is much more marked in the region of the grey matter than in that of the white, and more on the left than on the right side. The dorsal region of the cord is somewhat atrophied, and the lumbar greatly so, it being hardly larger than the dorsal, and nearly all trace of the lumbar enlargement having vanished. The brain presents no abnormal appearances beyond marked venous congestion, and careful examination fails to detect anything wrong with the cranial nerves. The arteries at the base of the brain are of healthy appearance. The right lung is somewhat oedematous, and its lower lobe congested. The left lung contains pneumonic patches scattered through it. In its lower lobe the consolidation is more general, and a small portion of it cut off and thrown into water sinks. The heart is healthy; the right side is full of black clot; the left side empty, except

for a little pale clot. The liver, kidneys, and spleen are normal. In none of the organs are there any gummata or other signs of syphilis.

Even to the naked eye very marked degeneration in the spinal cord was visible after it had been hardened in Müller's fluid. In the upper cervical region, situated between the columns of Goll and the posterior cornua, and quite distinct from them, were on each side nearly symmetrical tracts of sclerosis. That on the left side was of extreme distinctness, and reached from a little behind the posterior commissure to the posterior margin of the cord. The columns of Goll were sclerosed, but far less densely than the above-named tract on the left side. In the dorsal and lumbar region the antero-lateral columns were sclerosed in the crossed pyramidal tracts, the left a good deal more so than the right. Under the microscope the degenerated tracts exhibited the appearances of dense sclerosis. It is to be regretted that owing to a mistake the medulla was not saved. The microscopical examination of the portion of muscle removed from the left arm showed that many of the fibrillæ were much atrophied. They presented a wavy outline. In some of them the striation was preserved, in others it was very indistinct. There was neither fatty degeneration nor increase in the amount of connective tissue. The normal structure was preserved in the specimens of muscle from the right arm.

Among the numerous interesting features of this case are the epileptiform seizures which occurred a week before the patient's death. In connection with them it may be noted that there was no lesion discoverable in the brain to account for them, and that the patient had never before had any fit. Also that they were of a distinctly unilateral type, and consciousness was at least in two out of three of them not lost. A case reported by Dr. Gowers in vol. xxvii. of the 'Transactions of the Pathological Society,' presented in many respects similar features to the present one. It is that of a young man, aged 30, who, for three months before death, was subject to severe convulsive seizures, commencing unilaterally in the left hand. The patient died in one of these attacks. At the post-mortem the brain was found to be healthy, but there was postero-median sclerosis in the whole extent of the cord, most dense in the lumbar region. The patient had exhibited no spinal symptoms. The region between the columns of Goll and the posterior cornua was healthy. Charcot states, at page 116, vol. ii., of his 'Lessons on the Diseases of the Nervous System' (New Sydenham Society's Translation) that he has collected five cases of compression of the cervical region of the cord, and five cases of compression lower down, in which

there were epileptic seizures. It is to be presumed that in some of them at least there was ascending degeneration of the usual type. In one of these cases the pressure was exerted rather on the bulb than the cord. Was irritation of the medulla the explanation of the epileptic attacks here alluded to, and also of those in the present case?

With regard to the peculiar position of the patient's arm and hand, which is the same as that described by Professor Charcot as occurring in pachymeningitis cervicalis hypertrophica, when the lesion is situated in the lower part of the cervical enlargement, it may be remarked that the softening was diffuse in the present case, and certainly included the lower part of the cervical enlargement. In addition, the adhesions on the left side extended between the level of the fifth cervical and second dorsal nerve-roots. Dr. Ferrier states, in the number of 'BRAIN' for July 1881, that he found that stimulation in the monkey of the sixth cervical anterior nerve-root caused adduction and retraction of the upper arm, and extension of the fore-arm, with pronation and flexion of the wrist, and that stimulation of the fifth anterior root caused extension of the fingers and wrist, supination of the wrist, drawing of the hand to the mouth, and flexion of the fore-arm. It may be reasonably expected that in the atrophy of the fifth or sixth nerve-root the arm would take a position the opposite to that which it assumes when either single root is stimulated. But in simultaneous atrophy of the two roots, a position in some respects agreeing with that deduced as likely to occur in atrophy of the fifth root, and in other respects with that to be expected in atrophy of the sixth root, would be looked for. The patient's left arm was in such a position. It was abducted at the shoulder, and the hand was semiprone, as might be expected in simultaneous atrophy of the fifth and sixth roots. It was flexed at the elbow, and the wrist was extended, which would be expected in atrophy of the sixth root, but not when the fifth was affected. All the muscles of the left arm were wasted to an extraordinary extent. This agrees with the greater degree of softening on the left side, and the atrophy of the left nerve-roots.

On seeing the spinal cord in the hardened condition, Dr. Bastian at once recognised the sclerosed tract outside the columns of Goll, in the upper cervical region, as exactly similar to a tract of secondary degeneration which he found in a case several years ago, and kindly told me where I should find the case recorded, namely, in the 'Medico-Chirurgical Society's Transactions' for 1867. It is that of a man who died after receiving an injury to the spinal cord, at the level of the fifth and sixth cervical nerves, caused by a fall through a space

of twenty-five feet. There was found secondary degeneration of a descending character in the direct and crossed pyramidal tracts, while above the lesion, both in the columns of Goll and symmetrically on the two sides, in a narrow region between those columns and the posterior cornua, there was a separate tract of sclerosis.

It might naturally have been expected that, with the atrophic condition of the posterior roots which was found in the cervical region, the patient would have suffered from partial anæsthesia, at least in the left arm, but such was not the case. The tactile sensibility was repeatedly tested, and never found to be impaired over either arm, or in the region of the chest or back. Nor is this very surprising, if the common clinical experience be remembered of the frequency with which the sensibility in paraplegia both is less affected than the motor power, and the greater rapidity with which, when impaired, it returns than does the latter. That there was neither pain nor tenderness in the region of the spine, and that there were no pains in the arms, such as one might have looked for in cervical meningitis, is an instance of the fact that the classical symptoms of meningitis are not always present. As has been above stated, however, there was severe pain whenever the right arm was moved, for a fortnight before death.