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A CASE OF UNILATERAL PROGRESSIVE ASCENDING PARALYSIS, PROBABLY REPRESENTING A NEW FORM OF DEGENERATIVE DISEASE.[1]

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The patient, a man, 52 years old, was referred to me for diagnosis by Dr. Hugh T. Patrick, by whom and several other neurologists, and also by several ophthalmologists of Chicago, he had been carefully examined. About two years previous to coming under observation the patient's wife and some of his friends noticed that he occasionally stubbed his right toes and scraped his right heel, and gave other evidences of slight weakness or awkwardness of the right lower extremity. The patient was quite sure that the weakness in his leg came on slowly, the paresis up to the present time increasing with almost imperceptible gradations. It was not until the involvement of the right lower extremity had been apparent eighteen months that he noticed any weakness of the right arm. This weakness of the arm then appeared and soon became more and more evident, it being accompanied by a tendency to carry the arm rested against the body and flexed at the elbow. The paresis in the upper extremity had slowly become worse, although it had not nearly reached the degree of impairment observable in the leg. The patient can still use the arm for most purposes, but it soon aches from use. He can write slowly but with some difficulty and much aching. Up to the time of my examination his attention had not been called to any affection of the muscles of the face, but investigation showed some paresis in both the upper and lower distribution of the facial nerve; the forehead corrugated distinctly less on the right side than on the left in looking upward; the right face drooped slightly, its folds and contour being somewhat smoothed out. The time when the face was first involved could not be fixed.

About six months after the weakness of the right leg was first noticed the patient experienced much pain in the right gluteal region and upper and outer aspect of the right thigh. Later he had considerable hyperesthesia over the right lumbar and lumbosacral region, and two weeks later herpes appeared in this region with lessening of the hyperesthesia, although the entire outer and anterior aspect of the thigh now

[1] Read before the Philadelphia Neurological Society, December 18, 1899. For discussion on this paper see p. 222.

became hyperesthetic. Shortly after the paresis in the arm was first noted he also had some hyperesthesia of the upper and outer aspect of the arm, but no herpes. The hyperesthesia of the lumbar region of the right lower and upper extremities disappeared in a few weeks.

The examination showed distinct wasting of the right lower extremity, apparently uniform throughout the limb, that is, not localized in any muscular groups. The measurements were as follows: Right thigh, seven inches above the middle of the patella, seventeen and three-eighths inches; left thigh, eighteen and five-eighths inches; right leg, six inches below the middle of the patella, twelve and three-fourths inches; left leg, thirteen and three-eighths inches. The measurements, therefore, showed a difference of one and one-fourth inches for the thigh and of five-eighths of an inch for the leg. The various movements of the right leg were distinctly weaker than those of the left, but were nowhere absolutely abolished. Similarly all the movements of the right arm were distinctly impaired, but were nowhere absolutely lost. The dynamometer showed 180 for the right and 160 for the left. Faradic contractility was retained. The affected limbs were not spastic nor contractured. Careful examination showed retention of all forms of sensation. The tendon and muscle phenomena on the right side were all somewhat exaggerated. Knee-jerk was plus on the left side, but was considerably more exaggerated on the right. Patellar clonus was present on the right but not on the left, and the right side showed a slight ankle-clonus, which was absent on the left. The plantar reflex was normal on the left; but on the right, while the Babinski reflex was not present, the normal response was distinctly less marked than on the left. It might be described as between normal plantar flexion of the toes and the dorsal flexion of the Babinski reflex. Ocular movements and pupillary reflexes were normal and ophthalmoscopic examination showed no changes in the fundus. The patient had lost his hearing in the right ear as the result of a shell concussion during the Civil War, but otherwise the special senses were normal.

My examination corresponded with the results obtained by Dr. Patrick and others who had investigated the case in Chicago, and who regarded it as purely motor.

This case, simple in its characteristics and easily described, represents a rare and possibly a unique form of disease. Between seventeen and eighteen years ago I saw a case somewhat similar in its features, similar at least during the months

that the patient remained under my observation. This patient was a woman, 43 years old, who three years before coming under my observation, while carrying her last child, began to notice weakness in her left leg, and soon had a slightly shuffling and limping gait. She had had some sciatica during her pregnancy. The left arm became noticeably paretic a few months after the left leg, and both leg and arm were in much the condition of the patient here described at the time when the woman was under my care. She had been examined by the late Dr. E. C. Seguin, who, in a note to me, said, "The presence of so much increase in the reflexes on the affected side would lead me to cling to the idea that there was a central (cerebral) lesion causing changes in the crossed pyramidal fasciculus analogous to lateral sclerosis." Neither arm nor leg was contractured during the time that the patient was under my own observation, although the paresis of the affected limbs slowly increased. Sensibility was preserved. The patient complained, however, that she had at times had pains which seemed like neuralgia in the limbs, more severe in the leg, the pain being relieved by a very hot foot bath. She also complained of what she called nervous twitchings in both the leg and arm, but more marked in the leg. The special senses were not impaired. She had slight impairment of her control over the bladder; as she expressed it, her urine was hard to hold. This patient passed from my observation, but was alive three or four years since, and was then affected in the extremities of both sides, but just to what degree and in what manner I have not had the opportunity to learn. She had, however, become entirely unable to walk.

Several diagnoses are suggested by a study of this case, as (1) an unusual form of unilateral disseminated sclerosis; (2) unilateral amyotrophic sclerosis; (3) a progressive hemiplegia due to slowly increasing focal cerebral lesion involving the motor subcortex or the internal capsule; (4) a degenerative motor neuritis; and (5) a functional hemiparesis; but the case does not seem to fit in exactly with any one of these diagnoses. A hemiplegic form of disseminated sclerosis has been described. Nodules of sclerosis appearing at successive periods in lower and later in higher levels of the pyramidal system

might be suggested as an explanation. A case has recently been reported which bears upon this diagnosis; but does not correspond exactly with the case reported this evening.

This case was studied in the Salpêtrière service of Prof. J. Dejerine, and has recently been recorded by Drs. A. Thomas and E. Long.[2]

The patient was a man 47 years old, with a history of having acquired syphilis at the age of thirty-six. At the age of forty, that is, seven years before his death, he began to experience progressive enfeeblement of the right leg, which proceeded gradually to complete paralysis with diminution of sensibility on the same side. Later he had incontinence of urine and feces. After a short stay at the hospital he improved, the amelioration in his condition being maintained until the end of 1894, when the paralytic symptoms reappeared in the right leg with also additional features of numbness and paralysis in the right arm. When he again entered the hospital in 1895 he had almost complete paralysis in the right lower extremity, with contracture on extension, the hemiplegic gait, exaggeration of the knee-jerk, and ankle-clonus, on the paralyzed side, and a slight degree of muscular atrophy on the same side. The knee-jerk was also exaggerated on the left side. The right upper extremity was a little paretic; the reflexes at the wrist and elbow were exaggerated on both sides, but more on the right. Sensibility was greatly diminished in the leg and trunk of the right side, while the right upper extremity presented a slight hypoesthesia at the level of the hand and forearm; the arm proper, the shoulder, the neck and the head were free from anesthesia. Incontinence of urine and feces persisted. The special senses were intact. His condition remained stationary until 1896, during which year he died, following an attack of acute pleurisy.

This case presents several points of difference from the patient reported this evening. Diminution of sensibility was present in the right leg from the first, and this went on until it was extremely marked in both the lower extremity and the trunk; hypoesthesia was also present in the wrist and forearm

[2] Comptes rendus hebdomedaire des seances de la Société de Biologie, Oct. 13, 1899, 2 ser., t. 1, No. 28, p. 768.

after the arm became paretic. In my case no anesthesia has as yet been detected. Incontinence of urine and feces present, in the case of Thomas and Long, there have not been symptoms. My patient also denies any history of syphilis.

In the case of Thomas and Long a careful autopsy and microscopical examination were made. Several plaques of sclerosis were found. One of these was at the level of the fifth cervical root and occupied the entire right side, except a slight band of the antero-lateral cord. The plaque or nodule had disappeared entirely at the third cervical root above, and below at the level of the sixth cervical root. Another small plaque was found at the level of the seventh cervical root. This began at the dorsal part of the right column of Burdach and invaded the entire right dorsal horn as far as the base of the ventral horn. In the superior thoracic region some diffuse sclerosis was revealed, invading on the right the crossed pyramidal, direct cerebellar and ventral tracts; on the left the dorsal horn and tract of Gowers; and on both sides the columns of Goll and of Burdach, especially on the left. The same conditions were present in the mid-thoracic region.

The histological appearances are given in detail. The plaques of cervical sclerosis were notable because of the few vascular alterations. It is questionable whether the primary alteration was of the true nervous elements or of neurogliar tissue. The vascular origin of the diffuse sclerosis in the thoracic region appeared more probable, and recalled some of the lesions of spinal syphilis.

The ordinary train of symptoms found in disseminated sclerosis were not here present; but the lesions as described fairly explain the summary of symptoms given by the recorders of the case. The main feature of the disseminated sclerosis was its monoplegic form and the fact that the diagnosis was not made clear by intention tremor.

Several cases of unilateral amyotrophic lateral sclerosis have been put on record. In the paper read by Dr. Spiller at the last meeting of this society he referred to the fact that seven cases of the kind had been collected. Some of the diagnostic features of amyotrophic sclerosis are, however, wanting in our patient, and especially the absence of localized atrophy,

spasticity and contractures. It is not impossible, however, that these may develop later, and I cannot but feel that the case may represent a somewhat unusual form of amyotrophic lateral sclerosis. Indeed, the most probable diagnosis would seem to be that of a slowly increasing degeneration of the pyramidal fasciculi or of the cerebral motor neuron system.

A slowly increasing focal cerebral lesion does not seem probable, as such a lesion would not be likely to select for its slowly destructive effects first the motor fasciculi for the leg and then those for the arm and face.

Special and general symptoms of focal lesions, such as tumor, hemorrhage, softening and abscess are not present.

A degenerative motor neuritis is, of course, possible, but it is unlikely that such a peripheral affection would first select the leg, then the arm and then the face of the same side. The exaggeration of the deep reflexes is also an argument against a peripheral nerve affection.

The persistence and progressive increase of the symptoms and the absence of hysterical stigmata make a functional hemiparesis improbable.

Although some one-sided atrophy was present in this case, its other features do not seem to be in accord with the cases described by Brissaud[3] in discussing the cephalic trophoneuroses, which he shows are sometimes of cerebral origin, although the true trophic centers are situated at lower levels.

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24. WEITERE MITTHEILUNGEN UEBER VORSTELLUNGSREFLEX DEP PUPILLEN (Report on Pupillary Reflexes). J. Piltz (Neurol. Centralbl., 18, 1899, p. 496).

As a result of many experiments, Piltz arrives at the following results. There are pupillary movements associated with psychical conditions (cortex reflexes, attention reflexes) that consist in contraction of the pupil when the attention is fixed on a bright object lying to the side of the line of vision, and in dilatation of the pupil when the object is dark. Furthermore, the mere calling up of images results similarly; that is when the image embraces objects, the pupil contracts and vice versa with an image of dark objects. JELLIFFE.

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[3] Brissaud, E., "Leçons sur les maladies nerveuses," 2 s., Par., 1899.