PRIMARY LATERAL SPINAL SCLEROSIS.*

BY J. B. MARVIN, M. D.

Professor Theory and Practice and Clinical Medicine, Kentucky School of Medicine.

The disease designated by different authors as primary lateral spinal sclerosis, spastic spinal paralysis, spasmodic tabes dorsalis, is rare, and only recently has been recognized as a distinct affection, principally through the writings of Erb, 1875, and Charcot, 1876. Seguin, of New York, in 1873 recorded five cases of what he termed tetanoid paraplegia, and to him belongs the credit of priority in describing the symptom group. The disease comes on very gradually, is protopathic in origin, is associated with systematic, symmetrical sclerosis of the lateral columns of the cord in the crossed pyramidal tract; the sclerosis is not a secondary degeneration due to a lesion higher up in the cord or the brain. The disease is an affection of adult life, generally appearing between the ages of twenty and fifty years. According to most writers, males are more liable to the disease than females. In forty-nine cases cited by Althaus, there were twenty-four males and twenty-five females. The real cause of the disease is unknown; the neurotic temperament is marked in many cases; external injury, rarely syphilis, scarlet fever, typhoid fever, and other acute diseases may be followed by it. Brunelli, of Rome (International

* Read before the Kentucky State Medical Society, June 26, 1883.

Medical Congress, 1881), reported eleven cases occurring near Rome, caused by eating *Lathyrus cicera*. The disease is attended by loss of power in the extremities, with muscular rigidity, spasmodic twitchings and tremor, and an increase in the tendon reflexes. The muscles are well nourished, and there are no disorders of sensation. The positive symptoms, *i. e.*, muscular contraction and increased reflexes, and the negative symptoms, *i. e.*, no sensory, vesical, or rectal troubles, show unmistakably that the lesion is located in the crossed pyramidal tract. The symptoms are striking, yet the disease seems so far to have eluded the grasp of the pathologist. Ross, Bastian, and others, attribute to Drs. Dreschfield and Morgan, of Manchester, England, the honor of being the first to have proved by dissection the connection of the symptoms of the disease with sclerosis of the lateral columns in a primary and uncomplicated case of the disease.

The account given by Dr. Dreschfield (Transactions of the International Medical Congress, London, 1881) states that the giant cells of the anterior cornuæ were diseased. Althaus, in his recent work on Sclerosis of the Spinal Cord, claims that this case was of the amyotrophic variety. The only case which he accepts as of undoubted and uncomplicated primary lateral sclerosis is the one recently recorded by Minkowsky,* occurring at the University Hospital of Königsberg, under the care of Professor

*Deutsches Archiv fur Klin. Med., vol. xxxiv, page 433. 1884.

Naunyn. The patient was a woman, aged nineteen years, suffering with secondary syphilis. Fourteen months after admission the patient died. Microscopical examination of sections of the cord showed sclerosis of the postero-lateral columns, corresponding to the crossed pyramidal and direct cerebellar column. The gray matter of the anterior cornua was perfectly normal. Primary lateral sclerosis is a very rare disease, and there is danger of confounding it with the comparatively common condition of secondary descending degeneration, which may produce the same motor symptoms. A diagnosis of primary lateral sclerosis is only justified when the history shows a very slow and chronic onset, and there is no lesion present which could give rise to secondary degeneration. Special attention should be given to the condition of sensibility and to the state of the vertebral column. The disease is especially liable to be confounded with transverse myelitis. Cases of transverse myelitis of the dorsal region, in which the sensory disorders which were present at first have disappeared, while the motor phenomena have remained, are extremely liable to be confounded with primary lateral sclerosis. In the latter disease there are no sensory disorders. Motor rigidity and weakness are developed together; rigidity is more marked than paralysis, and there is no muscular atrophy at the upper level of the paralysis. Hughes Bennet has recently drawn attention to the fact that young women may have all the symptoms of primary lateral sclerosis, and he la-

ments that there is not a single point by means of which a diagnosis can be made.

Though it is generally easy to distinguish the functional from the organic lesion, yet the fact remains that in some cases competent observers have been able to differentiate sclerosis and pseudo-sclerosis only on the post-mortem table. Charcot claims that the lesion in primary sclerosis is wedge-shaped, and extends exteriorly to the pia mater and interiorly as far as the posterior cornua. This peculiarity in the localization of the disease, he claims, distinguishes it from secondary lateral sclerosis consequent on cerebral injuries; the lesion in this case being rounded, and not extending exteriorly as far as the pia mater, and also from sclerosis following myelitis, etc., when a small layer of healthy white matter remains between the posterior cornua and the sclerosed tissue.

The disease is probably the most chronic of any spinal trouble; it may extend over a period of many years; uncomplicated cases do not tend to shorten life. The disease is liable to extend into the anterior cornua, or the postero-external columns, wasting, bedsores, cystitis, etc., supervening; death being due to exhaustion or some intercurrent complication. Cases of recovery are recorded; I am skeptic enough to doubt the correctness of the diagnosis. In the line of treatment but little is to be expected; arsenic, bromides, nitrate of silver, anti-syphilitics, galvanism, and water-cure, have each their advocates.

The following brief notes will give you

a good clinical picture of the disease. I gave the case the severest scrutiny and feel justified in my diagnosis:

In June, 1883, Mr. B., of Tennessee, consulted me on account of a peculiar stiffness of his legs and great difficulty in walking. Mr. B. was twenty-seven years of age, unmarried, and of rather slight stature, though compactly and well built. He gave a good family history on both sides. As a child he was always healthy and active, he was reared in the country; when old enough he had plowed and performed the usual farm work. He had never been sick prior to the onset of his present trouble. In the spring of 1874 he first noticed a tired, heavy sensation in his legs. He attached no importance to these symptoms, as he thought they were due to hard work. Shortly his feet began to drag, his legs feeling very stiff and heavy, his toes turn in and he easily stumbles, his knees knock together, and his legs tremble violently when he attempts to walk. He kept his bed for some time, hoping in vain for improvement. He could walk only by the aid of a cane. His general health was good. Mr. B. now left his farm and went to Nashville, where he was closely confined for several years as a book-keeper.

Present condition: on examination I fail to get any history of syphilis, or injury to head or back. The muscles of the legs and back are well developed; there is no atrophy; no change in the electrical reaction of the muscles; he has a marked spastic gait, the posterior leg muscles are

very rigid, his heels are drawn up, and in walking his toes drag and appear to stick to the floor, his knees lock and strike together, his body sways; back strongly arched and chest thrown forward. He can cross his legs only with the greatest difficulty. When he is tired or excited, or standing in an awkward posture, his legs tremble violently. When sitting, if he presses his toes on the floor, violent reflex trembling is excited in the legs; ankle and knee clonus are well marked, superficial reflexes not as well marked as the deep. He has never had any symptoms referable to the bladder or rectum. He has never had any head symptoms. He does not complain of pain any where, but on close questioning he acknowledges that when he has walked much or overexerted himself, he has a dull, aching sensation in the lower dorsal region and in the calves of his legs. There are no tender spots, and no evidence of any thing abnormal about the spinal column.

In the discussion following the report of this case to the Louisville Medico-chirurgical Society, Prof. J. W. Holland stated that several years ago he had seen a patient with Prof. W. O. Roberts, whom he thought presented all the features of primary lateral sclerosis, and he had so diagnosed it. Prof. Roberts stated that the patient, a woman, aged nineteen years, had been confined to her bed for several months; recently bedsores and cystitis had developed. The patient dying within a few weeks, Prof. Roberts kindly notified Prof. Holland and myself that we might make an

autopsy. On account of difficulties interposed by friends of the deceased, the brain and cervical portion of the cord were not examined; the dorsal and lumbar portions of the cord only were carefully removed.

The cord externally presented nothing abnormal. It was suspended in Muller's fluid for several days, and then, with a freezing microtome, I made a large number of very thin sections. These sections to the naked eye revealed a dull, grayish, waxy patch in the lateral columns. Sections were stained in carmine, picro-carmine, aniline blue, and osmic acid. Carmine and picro-carmine gave the best results. The sections I pass around show beautifully the deeply-stained diseased patches in the lateral columns, extending from the posterior cornua to the periphery. Sections from the dorsal regions, examined under the microscope, reveal no disease of the gray matter; the ganglionic cells are numerous, well-formed, and present no evidence of granular or other degeneration. The disease is limited to the lateral columns proper. The nerve-tubes are almost entirely destroyed, the neuroglia greatly overgrown, numerous spider cells, and in some sections oil globules and amyloid bodies are seen. At the lumbar enlargement the sclerosed patch in the lateral columns is not so extensive. A slight patch of sclerosis is seen in the postero-external columns. The ganglionic cells are apparently healthy. The question arises, is this a case of primary sclerosis? The existence of bedsores and cystitis point to an extension of the disease to the gray matter.

Other than a slight patch of sclerosed tissue in the postero-external columns in the lumbar region, I am unable to detect any disease in the gray matter.*

There is one flaw in the case, viz., not examining the brain and cervical cord; but from the history of the case, and the microscopic appearances of the sections from the dorsal and lumbar cord, the lesion being symmetrical, and presenting all the features as laid down by Charcot and Bouchard, as distinguishing primary from secondary degeneration, I am inclined to adhere to Professor Holland's original diagnosis, and class this among the few cases of primary lateral sclerosis in which a post-mortem examination has demonstrated the connection between the lesion and the symptoms.

*A number of sections were shown with the microscope, and micro-photographs projected on the screen with magic lanterns.

LOUISVILLE.