PART II.

DISEASES OF THE GENERAL NERVOUS SYSTEM WITH KNOWN ANATOMICAL BASIS.

THE anatomical changes, which are found in the diseases belonging to this category, concern the central nervous system as well as the peripheral nerves. The former always suffers, the latter are only in certain cases affected. Whether the changes in the peripheral nerves are to be regarded as secondary, or whether the entire nervous system becomes affected in all its parts at the same time, so that the peripheral and the central lesions progress *pari passu*, can not be definitely decided. The nature of the anatomical changes will be discussed under the head of each individual affection. Combinations of the functional neuroses and organic diseases of the nervous system are, on the whole, rare. Such instances have been carefully studied by Oppenheim (Neurol. Centralblatt, 1890, 16).

CHAPTER I.

MULTIPLE SCLEROSIS—DISSEMINATED SCLEROSIS—INSULAR SCLEROSIS—SCLÉROSE EN PLAQUES—SCLEROSIS CEREBRO-SPINALIS DISSEMINATA S. MULTIPLEX.

ALTHOUGH multiple sclerosis is not one of the common affections of the nervous system, it is desirable and important for the general practitioner to possess a clear understanding of it, because the clinical appearances by which the different cases manifest themselves vary within such wide limits and remind us now of this, now of that spinal or cerebral affection, without ever completely simulating any one definite disease. The typical course given in the books is not very often met with in practice. Much more commonly one or the other of the classical symptoms is not found at all, or, if present, is only very slightly developed. On the other hand, symptoms are occa-

sionally encountered which are not included in the usual descriptions of the disease. In a word, multiple sclerosis is quite inconstant in its manifestations, a circumstance which often makes the diagnosis very difficult. In the investigation into the pathology as well as the clinical aspect, Charcot has done admirable and lasting service.

Symptoms and Course.—The course of a classical case is usually as follows: The patient first complains of general symptoms—headache, vertigo, digestive disorders—soon, also, of sensory disturbances in the upper and lower extremities, slight weakness, and a readiness to become fatigued. These symptoms may persist for months, yet relatively early one or several apoplectiform attacks may occur which sufficiently indicate the seriousness of the condition. It strikes the patient, as well as those who surround him, as a peculiar thing, that whenever

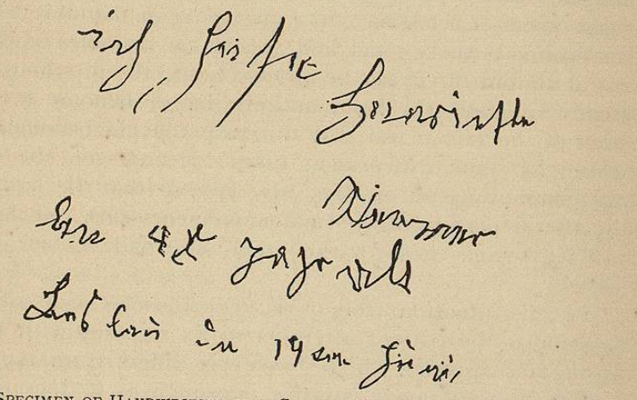

Fig. 164.—SPECIMEN OF HANDWRITING IN A CASE OF MULTIPLE SCLEROSIS. (Ich heise (heisse) Henriette Sterner, bin 48 Iahre alt. Breslau, den 19 Juni.)

he attempts to pick up something with his hands, or to make any other movement, a tremor appears, in exceptional cases implicating the facial muscles also (Cohn, Deutsche med. Wochenschr., 1890, 13), but usually confined to the upper extremities, which frustrates the intended movement more or less completely. If he attempts to raise a full glass to his mouth, he spills some of the contents. If he attempts to eat, the food is jerked off his fork, etc. Co-ordinated movements, such as are required for writing or playing the piano, become difficult, the handwriting becomes almost illegible (Fig. 164), and the condi-

tion is materially aggravated if the tremor is not confined to the upper, but if also the lower extremities, the trunk, neck, and head are attacked, so that on voluntary movements—on attempts to walk, for instance—the whole body first begins to tremble, and finally shakes so violently that the patient is forced to sit or lie down at once. This symptom, which is almost pathognomonic for multiple sclerosis, or at any rate most significant, is called "intention tremor," a term which does not, however, imply that the tremor is "intentional," but only that it appears on voluntary ("intended") movements. During rest no trace of it is observed. When the patient lies quietly and undisturbed in bed no tremor is present, whereas, if he is spoken to, examined, made to answer questions, and the like, a tremor over the whole body develops, which, of course, presents various degrees of intensity. It is most marked and characteristic if the patient is asked to bring his hand slowly to an object—for instance, to a pin laid upon the table. At first the motion is fairly good and steady, he trembles but little or not at all, but the closer he approaches to the pin the more unsteady becomes the hand and the larger become the excursions of the tremor, so that to grasp the pin becomes impossible. In some exceptional cases I have seen the shaking movements appear on one side only, so that the patient was capable of performing normal movements with one hand and one leg, when those of the other side had become entirely useless.

In this intention tremor the eye muscles also take part; as soon as the patient attempts to fix a point with his eyes nystagmus appears, which, however, differs from the tremor of the other voluntary muscles, inasmuch as it does not completely disappear during rest. As a subjective symptom the very annoying sensation of giddiness must be mentioned in this connection, which leaves the patient only when he lies quietly in bed, whereas it otherwise impedes him a good deal in his movements, especially in walking. Owing to the faulty innervation of the tongue and larynx, we meet with a peculiar speech disturbance; the patient talks slowly, in a monotonous tone, and awkwardly, and his speech is scanning, as he makes a pause after each word, almost after each syllable, so that it takes him a much longer time to express his thoughts than a healthy man: "Yes—doctor—I—am—very—much—fa—tigued—and—worn—out." As this is spoken in the manner indicated,

without any change of intonation, it is very characteristic indeed, and it is, together with the intention tremor and the nystagmus, pathognomonic for multiple sclerosis. It impresses itself so much upon the mind that once heard it can never be forgotten or misinterpreted.

To give a physiological explanation of the intention tremor is out of our power, and it is more especially not clear why it is so extremely common in multiple sclerosis, where we have such an irregular distribution of the anatomical lesions, whereas in most of the other cerebral affections it is absent. Whether Charcot's idea is correct, according to which the long persistence of the axis cylinders in the sclerotic foci has some connection with the tremor, or whether we should hold with Strümpell that the loss of the myeline sheaths, in consequence of which an abnormal diffusion of the nerve current from fibre to fibre occurs, is responsible for this, we can not decide, nor have we any proof of the correctness of Stephan's view (cf. lit.) that the existence of sclerotic foci in the thalamus gives rise to the phenomena, nor of Cramer's (cf. lit.) that the intention tremor has to be explained as analogous to the tremor which comes on after hard muscular exertion.

Though we may be justified in looking upon these three symptoms as constituting in a manner the typical picture of multiple sclerosis, we must, as we have said above, at once familiarize ourselves with the fact that even these may not all be pronounced, or, again, that there may be others to be found in conjunction with them, developing in the course of the disease. Among these latter we may mention certain spastic symptoms—rigidity of the muscles, increased tendon and skin reflexes, the above-described spastic walk—which, together, are liable to simulate, at least for a time, the picture of spastic spinal paralysis. This is the more likely as there are no sensory disturbances at all to be noted in multiple sclerosis; only in rare exceptions paræsthesias are observed, owing to which tabes and myelitis may be diagnosticated, especially if, as sometimes happens, bladder disorders are superadded. A careful study of the sensory changes has been made by Freund (Arch. f. Psych., 1890–'92, p. 319). That bladder diseases are by no means so rare in multiple sclerosis as was formerly supposed, has been pointed out by Erb, and after him by Oppenheim (Deutsche Med.-Ztg., 1889, 32). Glycosuria will be found associated with the disease if there are foci situated in the floor

of the fourth ventricle (Richardière, Revue de méd., Juillet, 1887).

Participation of the optic and other cranial nerves is not very rare, yet it is here much less important for the diagnosis, and much less significant for the course of the disease than, for example, in tabes. Diplopia is rarely met with; and equally uncommon is the neuritis and atrophy of the optic which leads to amaurosis. Uhthoff (cf. lit.) has pointed out, in an admirable study, that if optic atrophy occurs it is not like the primary atrophy in tabes, but that here it is a secondary process, which follows an active increase of the fine connective-tissue elements. It is self-evident that various disorders of sight are associated with this, yet they often present temporary improvement, and have usually a less serious issue than those of tabes. In general, it is characteristic of multiple sclerosis that its course is not uninterruptedly progressive, but that it shows remissions, during which the hopes of the patient as well as of his friends for his complete recovery are aroused. I have seen instances in which such remissions lasted for years and the symptoms disappeared to a great extent, and in which, just owing to this peculiarity in the course of the disease, the diagnosis could be made with some certainty.

Cerebral manifestations are not uncommon, and frequently a slight degree of dementia develops, which to the patient himself makes his condition more bearable. It must also be regarded as a sign of beginning mental weakness, I think, that in some cases the patient frequently laughs boisterously without a cause. One of my patients had spells of loud laughter, which lasted from one to three minutes, and which appeared usually without sufficient motive. I have never had occasion to observe pronounced states of depression or exaltation in the course of this disease. The vertigo, which of course must also be regarded as a cerebral symptom, has been spoken of above. Apoplectiform attacks in the beginning of the disease are not rare; epileptiform seizures may be found, if the cerebral cortex is more especially implicated.

It has been shown by Charcot that in certain cases the development of the symptoms appears to be abortive and the affection, one might almost say, remains latent and can only be recognized by the peculiar shaking tremor. He proposed for these instances Trousseau's designation, "*formes frustes*," and it seems that in multiple sclerosis such forms are observed relatively frequently. Soucques studied these carefully under the direction of Charcot (Progrès méd., 1891, 11). As an example of the general course which the disease may run I insert here the following history of a patient in my wards, who is still living:

Paul W., thirty-one years old, began to be sick ten years ago during his military service. At first, at times he could not feel his rifle in his left arm, and then in the same year he was often conscious of a slight feeling of fatigue, which was associated with vertigo. He had a good deal of difficulty with his arms and his legs; they always felt as if they were asleep, and any muscular action necessitated the greatest exertion. He could not go through his salutes in the proper manner, and he was repeatedly punished on this account. At the same time he had now and then vomiting and weakness of the bladder for quite a long time, so that, on coughing, small quantities of urine were passed involuntarily. On examination, we are told, Romberg's sign was absent and the patellar reflexes were increased. A few months later, marked weakness in the right arm and the right leg became manifest, and the acuteness of hearing became diminished on that side. The patient complained of an annoying double vision. In 1879 he had some difficulty in swallowing; the bolus would stick in his throat, so that he had to force it down. In 1880 pronounced deliberation in speaking is said to have been noticeable, and the patient at that time also complained that he could not lay his tongue upon certain words which he wished to use. The speech disturbance soon passed off, but the patient suffered from various troubles till January, 1884, in which month I saw him for the first time. He then presented the symptoms of an incipient tabes, but it was noted as a remarkable feature that the patellar reflexes were retained. The lancinating pains, however, the paresis of the legs, the diplopia, the paresis of the bladder, the unsteadiness which appeared especially in the dark, seemed sufficient to warrant the diagnosis of tabes, and in the out-patient department this diagnosis was made, although with some reservation. The patient declined to enter the hospital. He was therefore ordered galvanism, but was lost sight of in the summer of 1884. Two years later he was treated at a hospital in this city for six months. Although I was unable to obtain a record of the case, I heard that the tabetic symptoms were very indistinct, and that the condition suggested rather a spastic paralysis. The patient was again lost sight of. Finally, on January 8, 1888, he was admitted to the medical ward of the city almshouse, where he still remains. From a note made on January 10, 1888, the following is extracted: The patient is a well-nourished man; as he lies quietly in bed, the general aspect suggests nothing

abnormal; if, however, he is asked to perform any movement, the whole body—trunk, head, and extremities—is seized with a violent shaking tremor, which makes it difficult for him to get up, and impossible for him to walk without assistance from another person,

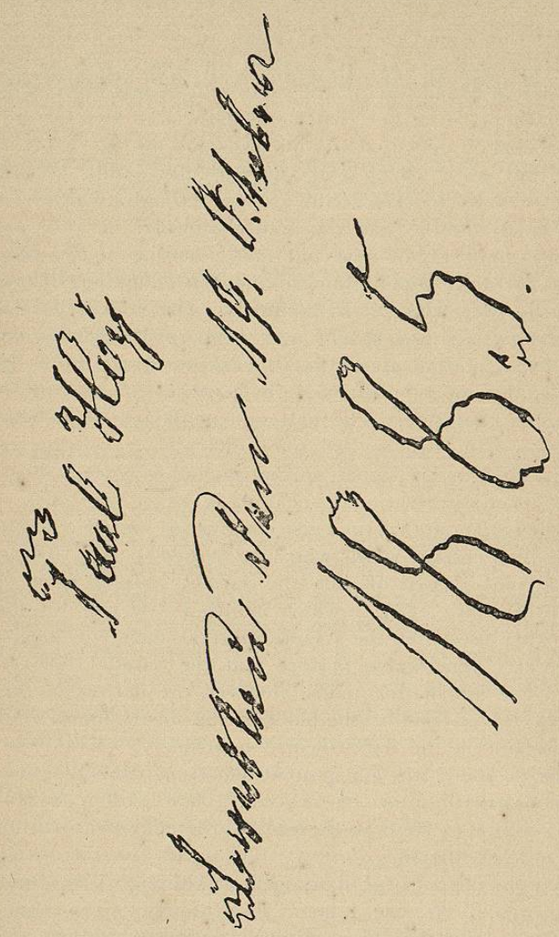

Fig. 165.—Specimen of Handwriting illustrating Alcoholic Tremor.

even when supported by two canes. If he is allowed to discontinue all attempts at moving, the tremor gradually abates, and five or ten minutes later he is perfectly quiet again. The patient is unable to feed himself, and can not occupy himself with anything. The muscular strength is retained everywhere. In the domain of the cranial

nerves nothing but nystagmus can be noticed, which is especially well marked on the right side. The facial, hypoglossal, etc., are normal. The tongue is protruded steadily and straight. Speech is slow, although not distinctly scanning. There are no motor or sensory speech disturbances. The tendon reflexes in the upper, but

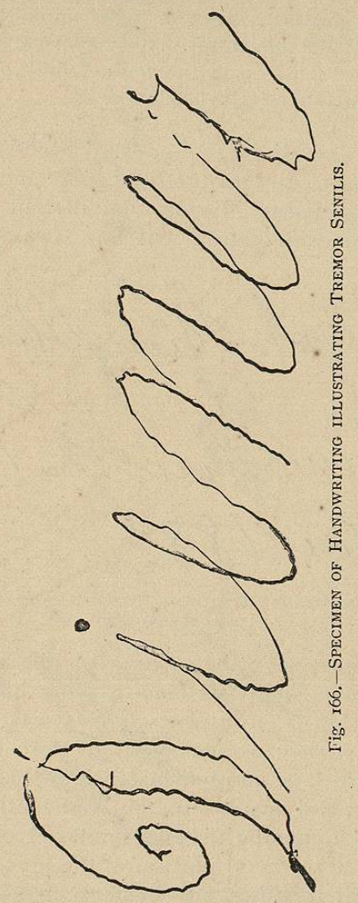

Fig. 166.—Specimen of Handwriting illustrating Tremor Senilis.

more especially in the lower extremities, are increased, and the skin reflexes are without exception well marked. Sensory changes can nowhere be demonstrated, and the bladder disturbances, which were present on previous occasions, have disappeared. The spinal column is nowhere tender on pressure. Among the subjective complaints