that the excitability also undergoes qualitative changes and we find reaction of degeneration. (3) The loss of the tendon reflexes, which is sufficiently explained by the disappearance of the ganglionic cells, a part of the reflex arc. It is only because the lower extremities are rarely affected that the patellar reflexes are usually retained. (4) Sensibility remains everywhere and for all kinds of impressions intact (touch, pressure, pain, temperature). The coldness and blueness of the hands is to be attributed to the disuse of the muscles. True trophic disturbances of the skin, as well as bladder and rectal symptoms, are usually absent.

Diagnosis.—Remembering, then, the different points just alluded to, the diagnosis should be easy, and it will not be difficult to avoid confounding the disease with myelitis, neuritis, or syringomyelia. The flaccid paralysis, the absence of all symptoms of motor irritation and sensory disturbances, is especially of moment in differentiating this disease from myelitis. More particularly characteristic is the commencement, the onset of the disease in the small muscles of the hands. If this has been well pronounced, an error in diagnosis is unpardonable.

Ætiology.—With reference to the ætiology a little more is known about this disease than about spinal infantile paralysis; for certain cases at least it has been shown that overexertion of the muscles, as happens sometimes to those who work with the sewing machine, has a causative influence, or at any rate the disease has been preceded by some overexertion of the muscles, to which we are then justified in attributing an ætiological importance. The conditions, however, under which paralysis and fatigue of the muscles lead to atrophy—why, for instance, the serratus magnus (Chvostek) is in some cases at first thus affected—we are wholly ignorant of, just as we do not know the conditions under which the genuine hypertrophy develops which we often find in the biceps of blacksmiths. Recently, again, attention has been called to the fact that the disease may be hereditary, by Bernhardt (Virchow's Arch., 1889, 115, 2) and by Werdnig (Arch. f. Psych., xxii, 2).

Little need be said about the therapeutics; there is no effectual treatment, and all measures that have been tried have not been efficient in hindering the progress of the disease.

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II. LESIONS OF THE WHITE MATTER OF THE SPINAL CORD—"LEUCOMYELITIS."

While, as we have said on page 424, the lesions affecting the gray matter (poliomyelitis) are almost entirely confined to one portion of it—namely, the anterior horns—we shall soon see that this is different with the lesions of the white matter, to which the general name leucomyelitis may be given (λευκος, white). Here different parts can be attacked, either alone or in conjunction with others, and it is of great importance to differentiate between the clinical symptoms which occur in

the diseases of the different columns or "systems" (Flechsig), hence called "system diseases."

The affection is either a primary one, when it is often impossible to ascertain any ætiological factor, or it occurs secondarily and as a consequence of certain affections of the brain and the spinal cord itself, such as traumatic inflammations and compression. We shall consider both separately.

A. Primary Lesions of the White Columns.

Regarded from an anatomical standpoint, the primary tract-degenerations of the white substance consist in a destruction of the nerve fibres and a simultaneous increase of the neuroglia. The medullary sheaths are the first to disappear; the axis cylinders, which are more resistant, do not degenerate till later. Compound granular corpuscles, which remove the detritus from the diseased regions (Ziegler), accumulate in the lymph sheaths of the vessels. The increasing neuroglia crowds in and displaces the empty nerve tubes, a process which, in conjunction with the thickening of the walls of the vessels, which develops at the same time, is described under the name of sclerosis, or gray degeneration.

An affection confined to one nerve tract or system has up to this time only been observed in the antero-lateral but not in the posterior columns. In the former, the anatomical arrangement of which has been described above, we meet especially frequently with sclerosis of the so-called crossed pyramidal tracts, but the lesion does not necessarily extend over the whole length of the tract, but may be only partial (Westphal). Most of the cases which have come under observation were, however, not pure instances, but presented other anatomical changes as well, and there is only one case reported, by Dreschfeld, in 1881, which, viewed from an anatomical standpoint, can pass for a pure lateral sclerosis.

The primary sclerosis of the lateral columns—spastic spinal paralysis, *tabes dorsale spasmodique*—was first described by Erb and Charcot in 1875, and characterized by them as a motor paralysis with remarkable increase in the tendon reflexes; and, indeed, if we examine such patients, all we find is that they have lost to a greater or lesser extent the use of their legs; they are unable to walk, the feet are glued, as it were, to the floor, and the patient can only shuffle along, the inner margin of the foot never leaving the ground. At the same time the

muscles feel firm and hard, the legs are in extension, and any attempt at flexion is difficult. If such a patient is made to sit on the edge of a table the legs do not hang down flaccidly, as might be expected, but are thrown into a state of tetanic tremor, produced by contractions of the quadriceps extensor. There is an enormous exaggeration of the patellar reflexes, and the ankle clonus is obtained without the slightest difficulty. With the exception of the inability to walk, the patient has no subjective complaints; neither sensation nor the functions of the bladder, rectum, or the sexual apparatus show any abnormity. An implication of but one of these would at once exclude the diagnosis of lateral sclerosis, as would also (and this should be especially remembered) the existence of any muscular atrophy. The very characteristic spastic or spastic-paretic walk of the patient, the traces which his feet leave on a gravel path, for instance, and which can be followed up as distinct continuous streaks, the shuffling noise which accompanies every step when he attempts to walk about the room, these are of great diagnostic value; the examination of the soles of the patient's shoes, which appear thinner and more worn down on the inner side, will be of interest and value.

The disease may be congenital (Lorenz, Bernhardt, cf. lit.), and may occur in more than one member of the same family, as we have stated above (page 274), but it often begins later in youth or in middle life, attacking first the one then the other leg, without, as a rule, extending to the arms or trunk, yet the upper extremities are said to be occasionally affected (Strümpell). The disease may last years or tens of years without presenting any decided change for the worse. Death is brought about by intercurrent diseases. It is not known whether, as in progressive muscular atrophy, overexertion is of ætiological importance; instances, however, in which acrobats (Donkin) and hod-carriers (Munter) were attacked seem to suggest this. Morgan pointed out that exposure to cold, such as long standing in water, may be the immediate cause of the disease (Morton, Lancet, January 19, 1881).

The form of spastic paralysis, analogous to a tabes developing on a syphilitic basis, which has been regarded by Charcot as a transverse syphilitic myelitis, and which has been studied carefully first by Erb, later by Muchin, P. Marie, and Kowalewsky (Neurol. Centralbl., 1893, 12), must be regarded as a distinct disease. It occurs much less frequently than tabes, and

differs from the spinal paralysis just described, inasmuch as here we find sensory and trophic changes as well as eye-muscle palsies. The differential diagnosis may, however, be impossible.

Much more frequent than a lesion confined to the crossed pyramidal tracts is one which implicates not only these, but with them the posterior columns and the direct cerebellar tracts, in which, although not always, Clarke's columns take part. The anatomical character of this "combined system disease" which results from these lesions has been repeatedly described (Westphal, Gowers, Strümpell). The symptoms vary according to the distribution of the lesion; thus, if the disease of the lateral columns extends low down, while the posterior columns in the dorsal and lumbar region present no changes, rigidity of the muscles and increase of the reflexes will be found. If, on the other hand, the disease in the posterior columns extends farther downward, these symptoms will be absent, the lesion in the lateral being neutralized, as it were, by that in the posterior columns (Westphal).

Not rarely the affection seems to depend on faulty development, a condition which we may meet with in more than one member of the same family, and which may be hereditary. In these cases the disease appears in early childhood, and, as we said, sometimes in several children of the same family. It has been called, after the author who first described it, Friedreich's "hereditary ataxia." Senator (cf. lit.) has called attention to the possibility of a congenital atrophy of the cerebellum, the medulla oblongata, and the spinal cord. The motor disturbances in the children begin in the feet, the walk becomes awkward, they stumble, and in passing over small obstacles have to look at their feet to keep from falling, etc. (Fig. 142). The patellar reflexes disappear; the arms are not affected until later, and, indeed, they are by no means always implicated. The second motor disturbance establishes itself in the muscles of the tongue and the lips which are necessary for speaking, and this produces a very characteristic defect of speech of motor origin. Finally the muscles of the eyes become implicated, and there results a distinct nystagmus. The combination of these three symptoms is pathognomonic for this rare disease. It has no connection with tabes and sensory changes, and bladder symptoms, manifestations which are probably never wanting in cases of tabes, are never met with in the disease under

consideration. Nor can it be mistaken for multiple sclerosis, as vertigo and "scanning speech" are never associated with it. The course is tedious, the prognosis always unfavorable, the muscles undergo atrophy in consequence of inactivity, and contractures occur in the joints.

Similar symptoms are observed in adults in cases of combined lateral and posterior sclerosis; yet there are certain peculiarities to which Gowers especially has drawn attention.

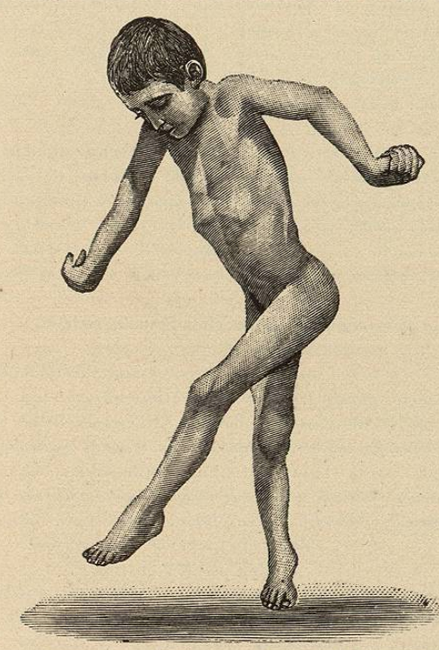

Fig. 142.—FRIEDREICH'S DISEASE. The patient is held under the arms. (After CHAUFFARD.) (Semaine méd., 1893, No. 32.)

The disease was named by him "ataxic paraplegia." The lower extremities are ataxic and paretic, which gives rise to an uncertain, swaying walk; but this is associated with paræsthesias, weakness of the sphincters, and decrease of the sexual power. The patellar reflexes are at first increased, and only at times become lost later in the disease. Such an increase is never known in hereditary ataxia. During the period of increase, rigidity of the muscles, spasm, and ankle clonus are present. It is evident that cases of this kind may be mistaken

for tabes, especially if the patellar reflexes are absent, as exceptionally occurs. Then the history may be of use to us, as syphilis seems to possess no ætiological importance whatever in the combined sclerosis, while exposure to cold and overexertion seem to be of considerable moment.

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B. Secondary Lesions of the White Columns.

Lesions of the motor centres of the brain cortex, or lesions of the path between these centres and the motor ganglia of the spinal cord, the so-called cortico-muscular tract or pyramidal tract, give rise to a descending degeneration of the motor fibres on the same side as the brain lesion. This secondary degeneration is in the cord continued in the crossed pyramidal tract of the opposite side, while the direct pyramidal tract presents only traces of it. About the causes of this sclerosis we possess just as little definite knowledge as about the clinical symptoms