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CHAPTER II.

TABES DORSALIS—LOCOMOTOR ATAXIA (POSTERIOR SPINAL SCLEROSIS—LEUCOMYELITIS POSTERIOR CHRONICA).

THE second of the diseases belonging to this group certainly deserves to be considered as one of the most important of those with which we are acquainted, not only because it is to be reckoned among the diseases of the nervous system which occur relatively frequently, and with which the general practitioner is not rarely brought face to face, but also because its clinical picture presents so many essential differences that it requires a large experience to feel at home with it on all occasions. Nobody questions the importance of the recognition of the disease in its early stages if only on account of its bearing upon the treatment, but many do not appreciate the difficulties which this early diagnosis entails. The more cases of tabes we see, the more we are surprised at the protean character of the symptoms, and the more are we convinced that almost every case offers some point of particular interest, and that occasionally even an expert can be sure of the diagnosis only after repeated examinations and long observation.

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Symptoms.—The symptomatology of tabes is so comprehensive that in order to get a clearer idea of it we shall in our description separate the cerebral from the spinal symptoms.

The cerebral symptoms which appear in the course of the

disease are referable either to the cranial nerves or to the brain substance. When the latter is affected, it is sometimes the cortex, at other times the white substance, or again the basal ganglia, which are most deeply implicated.

In considering the cranial nerves, we shall find that there is hardly a single pair which can not be affected and in which lesions have not been repeatedly described in cases of tabes. However, as we shall see later, not all of them are implicated with the same frequency. Among them the nerves supplying the muscles of the eye are most commonly, the facial most rarely, attacked. Between these extremes we may put in descending order the vagus, the optic, the fifth, the olfactory, the glosso-pharyngeal, the accessorius, the hypoglossal, and lastly the auditory. I have observed three cases in which several pairs of nerves were involved at the same time and in which the onset of the disorder was somewhat acute.

The lesions of the olfactory nerve possess no great practical significance, and it is not quite certain that they are not more frequent than is generally supposed. They consist of a weakening or even total loss of the sense of smell, or in the perception of peculiar, often disgusting odors, as we have shown on page 26. We do not know whether these changes are due to anatomical lesions or only to some functional disturbances, and but little is known about the course of such disorders of the sense of smell. Occasionally, when examining into the condition of the sense of taste, one may accidentally discover an affection of the sense of smell without being able to ascertain how long it has already existed, as it can easily have escaped the notice of the patient. Only those who use tobacco or snuff perceive the defect very early and appeal to a physician for advice and help. Unfortunately, we can do but little. The treatment of these affections has been dealt with on page 27.

The most frequent lesion of the optic nerve in tabes is atrophy or gray degeneration. Usually both eyes become affected, if not simultaneously, at least within a short time of each other, and it is quite rare for one eye to be diseased while the other remains healthy for any great length of time. The patients complain that everything seems as if covered by a gray veil. The loss of vision is particularly rapid at first; it then becomes much slower, and the complete amaurosis occurs much later than one would have expected from the brusque

onset of the trouble. Along with this, a narrowing (not always concentric) of the visual field appears, as the peripheral portions of the retina are the first to become impaired in their functions. The perception for color may also be affected, as we pointed out on page 34. The order in which these changes occur is not always the same. As a rule, however, the loss of color perception and the narrowing of the visual field precede the lessening in acuteness of the central vision, and it is exceptional to find diminished acuteness of vision and marked disturbance of color sense combined with a normal visual field.

With regard to the frequency of the affection of the optic nerve in tabes, the usual statements of authors hardly give a correct idea; the more careful our examinations are the more often do we find them. According to my experience, it may certainly be said that they occur in sixty per cent of all cases (cf. the excellent piece of work of Martin, *De l'atrophie du nerf optique et sa valeur pronostique dans la sclérose des cordons postérieurs de la moelle*, Paris, Asselin et Houzeau, 1890).

The ophthalmoscopic examination shows a pale grayish white or bluish white, but not pure white, discoloration of the disk, which is thought to be produced by the obliteration of numerous fine vessels in the optic nerve. When the amblyopia is marked, but no perceptible changes in the disk are found, we must think of a retrobulbar degeneration of the optic nerve. From a pathological standpoint we are dealing with a degenerative atrophy, first of the medullary sheaths, and then of the axis cylinders. The theory that these changes are due to an action of the sympathetic nerves or to changes in the vaso-motor nerves brought about by the spinal disease is quite untenable, for the process is a neuritis in which we have a wasting of the nerve fibres and changes in the interstitial tissue, such as have been described on page 331.

For the optic atrophy the outlook is altogether unfavorable; although a slight improvement or a temporary arrest of its progress may give the patient a delusive hope of recovery, the termination is always in total blindness. It is true that the process may take several years, during which the patient is still able to find his way about by himself with the aid of a stick.

With such a prognosis we shall not be surprised if the treatment is without avail. The subcutaneous injections of strychnine, one milligramme (grs. $\frac{1}{60}$) twice daily in the neigh-

borhood of the eye, as proposed by some, are of value only because they give the patient the comforting satisfaction that something is being done for him, but they really have no curative properties, and it is improbable that they even postpone the unfavorable issue.

In a few isolated cases transient lachrymation has been observed (Patrolacci, Thèse de Montpellier, 1886; Féré, L'Encéphale, 1887, vii, 4).

The nerves which supply the eye muscles—the third, the patheticus, and the abducens—the affections of which have already been considered in Part II, Chapter III, frequently become attacked in the course of tabes. Besides the insufficiency of convergence, the central form of which may be termed motor asthenopia (Hübscher, Deutsche med. Wochenschr., 1892, 17), one often encounters a diplopia resulting from a paralysis of the ocular muscles. This may appear suddenly, and after a longer or shorter duration disappear as quickly; or, again, it may recur repeatedly and be a source of great annoyance to the patient in his daily occupations. An abducens paralysis may also occur by itself, and, finding this, one should always look for a commencing tabes, for it is frequently the first sign of this disease in an apparently quite healthy person. If the affection remains stationary, it is to be regarded as being due to a nuclear lesion; the same remarks apply to a ptosis which, occurring by itself, is also a suspicious sign, and should lead us to look for tabes. In cases of oculomotor paralysis the lesion is also relatively frequently nuclear (page 46). Watteville (Neurol. Centralbl., 1887, 10) has called attention to a paralysis of the movements of convergence, especially in the initial stages of tabes. Borel, in a paper published under the direction of Landolt in Paris (Arch. f. Ophthalm., Novembre, 1887), has dealt with the same symptoms. Several of the extrinsic eye muscles may be affected at the same time, and an ophthalmoplegia externa is not infrequently observed in the course of tabes.

The behavior of the intrinsic eye muscles is not less interesting, and the condition of the pupils deserves the most thorough examination; they are rarely normal and of the same size in both eyes. Frequently some abnormality of reaction is demonstrable; the marked contraction (myosis), the difference in the size of the two pupils (anisocoria), and the loss of the light reflex have already been mentioned. These changes force

us to assume a lesion in the floor of the fourth ventricle (Guillery).

The ophthalmoplegia interna of Hutchinson, in which besides the loss of the light reflex there is also paralysis of the muscles of accommodation, is much rarer. The pupils of those afflicted with tabes may frequently be found to dilate promptly and normally under strong and painful irritation of the skin, as, for example, that produced with the faradic brush.

The rôle which the affections of the fifth nerve play in this disease is quite subordinate; paralytic conditions of either its motor or sensory branches as the result of tabes have, it seems, never been observed except in Westphal's case, in which there was degeneration of the ascending root of the fifth, and among the signs of irritation only the headache, traceable to the nerve endings in the dura, is occasionally met with. A certain relationship is said to exist between tabes and genuine migraine, but in considering these cases one must make sure that the migraine has not been inherited, and furthermore note whether the attacks become more or less severe after the development of the tabetic symptoms. According to some observations, in such cases the headache of the migraine becomes less and less severe, and eventually disappears, while the nausea and vomiting still persist, so that it is then impossible to say whether we are dealing with a gastric crisis of tabes or with an abortive attack of migraine. In certain cases of hemisphericity, if there has been, for instance, a syphilitic infection at some previous time, it is always well to examine carefully for any traces of tabes, more especially for the absence of the patellar reflex. Occasionally one meets with paræsthesias in the face, the patient complaining of a sensation as if one half of the face and the lips were swollen; this is probably also due to an affection of the ascending root of the fifth nerve.

Lesions of the facial nerve are so rare in tabes that, when they occur, one can not help raising the question whether they are not to be regarded merely as accidental complications. Among three hundred and forty-five cases of locomotor ataxia, I have observed only two in which any of the muscles supplied by the facial were affected.

About the same may be said of the auditory nerve. There is no question but that lesions of this nerve may be caused by tabes or develop in the course of the disease, but they are very rare indeed; they manifest themselves by a diminution or a

total loss of the power of hearing. The patients complain of deafness, which may have developed gradually or have come on acutely. In both cases the symptoms are due to organic disease of the nerve; in the former we have to deal with a gradual gray degeneration of the nerve trunk, in the latter with a nuclear affection. Too few cases, however, of involvement of the auditory nerves in tabes have as yet been observed to enable us to speak with much certainty of their pathology (Hermet, *L'Union méd.*, 1884, 86; Morpurgo, *Arch. f. Obrenhik.*, 1891, xxx, 26). Under what conditions the so-called Ménière's symptoms appear in the course of tabes requires to be studied more closely. I have seen them in two of my cases, but they disappeared again in a few weeks, and in these cases, unfortunately, no anatomical examination of the internal ear could be made.

Functional disturbances of the nerve of taste have now and then been described in the course of tabes. In a few instances as in the case of Erben, which we considered on page 108, the nucleus of the glosso-pharyngeal nerve was degenerated, and during life such derangements of the sense of taste existed that the patient was at times unable to distinguish sweet things from those which were acid or salty. To these lesions no great practical significance can be attached.

On the other hand, there is a great variety of manifestations associated with tabes which are due to lesions of the vagus. In this connection we have disturbances more generally of the digestive, but also of the respiratory and circulatory organs. They occur with irregularity, and may disappear again quite suddenly. Following the suggestion of Charcot, we designate them "crises." Of those affecting the digestive system the so-called "pharyngeal crises" are relatively the rarest. These consist of paroxysmal movements of deglutition, which occur from twenty to twenty-four times a minute, and succeed one another in this way for ten or twenty minutes; the attacks may be associated with a noisy inspiration, and may suggest hysterical singultus; in some cases they can be produced at will by pressure on the side of the larynx (Oppenheim).

The gastric crises (Charcot) are far more frequent; they consist of paroxysmal attacks of retching and vomiting, during which the patient, without any particular exertion, may vomit large quantities of strongly acid, slimy, or watery material, some-

times of a blackish appearance, after which he feels greatly relieved. These attacks are repeated for several days, sometimes for a week or two, once, twice, or even oftener, every day, and then disappear entirely for a longer or shorter period. In some cases the vomiting is associated with cardialgia, but usually it is uncomplicated. It is not at all connected with the taking of food; indeed, it not infrequently occurs early in the morning when the stomach is empty, and if the patient be a drinking man it may arouse a suspicion of the morning vomiting of drunkards. The differential diagnosis is, however, not at all difficult; if the vomiting be associated with vertigo, a sensation of anxiety, and a quickening of the pulse, it can not be considered simply as a "gastric crisis." This paroxysmal vomiting is of the greatest importance for the diagnosis of tabes. It is frequently regarded as dependent upon some stomach trouble and treated as such for a long period without any sign of improvement, until finally, perhaps by accident, our attention is drawn to some other symptom which places the diagnosis beyond doubt. If a person have paroxysmal vomiting and complain occasionally of violent rheumatoid pains in the legs, we should examine most carefully for tabes, and we shall frequently be surprised at the ease with which we can make a diagnosis, and wonder that we had ever been under the impression that the patient had simply "chronic gastritis" and "rheumatism." The statement of Eckert (*Die intestinalen Erscheinungen der Tabes*, Inaug.-Diss., Berlin, 1887) that gastric crises must be divided into those of central and those of reflex origin deserves to be investigated more closely. In the central form he assumes, besides a general condition of irritation in the brain, some affection of the nucleus of the vagus, in the reflex form a peripheral irritation of the vagus which, under certain circumstances, may be produced by the ingesta. He holds that in the latter cases the vomiting is not associated with any distressing nausea, so that the patient suffers relatively little.

Sometimes intestinal disturbances manifest themselves by intense "lightning" pains about the rectum and anus, the "anal crises"; in other cases by tenesmus, which forces the patient to go frequently to stool, though he is able to pass little or nothing; and lastly by the so-called tabetic diarrhoea, about the causation of which we are absolutely ignorant. This diarrhoea may be more or less persistent, and be followed by an equally

protracted and obstinate constipation. Incontinence of fæces is rarely present, though on rectal examination we shall occasionally discover sensory disturbances, particularly anæsthesia of the mucous membrane. Paræsthesias may also occur, and the patient may experience a sensation as if he had a foreign body in the rectum.

By "laryngeal crises" we mean those paroxysms of dyspnoea which may occur when the patient is lying down, or, in other cases, only when he attempts to move or walk about. Sometimes they appear in the form of peculiar suffocative attacks, accompanied by violent coughing, and are often preceded by a sighing or whistling inspiration. These attacks may last several minutes, during which the suffering may be so intense that the patient gives up all hopes of recovery. Attacks of even moderate intensity, in which a long, sonorous inspiration follows several short expirations, are most disagreeable for the patient, and appear very serious; under some circumstances they may be mistaken for whooping-cough. These crises are caused by changes of temperature, speaking for a long time, or by strong odors, smoking, etc. The result of the laryngoscopic examination is frequently negative; in other cases one finds paralysis of some of the laryngeal muscles; here also, in all probability, we should distinguish a central and a reflex form.

Abductor paralysis—i. e., paralysis of the muscles that open the glottis—sometimes occurs among the early signs of tabes, and may lead to serious danger of suffocation; but we are unable to say whether this should be attributed exclusively to paralysis of the abductors, or to spasm of the adductors alone, or to both conditions. We may consider the condition described by Gray (*Brain*, January, 1888), in which the voice often breaks and takes on a high falsetto, as a kind of "laryngeal ataxia."

Attacks of angina pectoris, with all its characteristic symptoms, are rarely met with in tabes, though Vulpian, among others, has seen them (*Revue de méd.*, 1885, v, 1).

Lesions of the accessorius are considered as rarities in the course of tabes. They are occasionally found associated with a posticus paralysis when the outer branch of the spinal accessory is also affected; the sterno-cleido-mastoid and trapezius muscles then show atrophic changes. In a case observed by Martius there was an atrophic paralysis of the upper portions

of both trapezii, while the sterno-cleido-mastoids, which also receive fibres from the cervical plexus, were not affected. Whether, and if so under what circumstances, one or both of the two nuclei of the accessory nerve are affected (the nucleus accessorius vagi and the nucleus spinalis) we are absolutely ignorant. It is also uncertain whether symptoms of irritation in the domain of the accessorius—e. g., torticollis—ever occur in the course of tabes.

Among the lesions of the hypoglossus there is one which deserves a special mention in this place—that is, the hemiatrophy of the tongue described above, which Ballet (*lit.* page 144) stated was relatively often observed. He even went so far as to say that, when one found this hemiatrophy, tabes

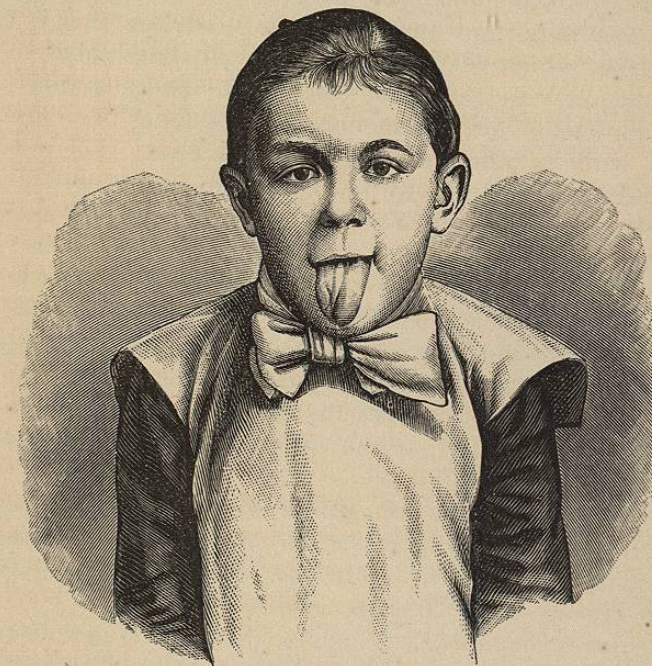


Fig. 170.—HEMIATROPHY OF THE TONGUE IN AN OTHERWISE PERFECTLY HEALTHY CHILD (personal observation).

should always be suspected. We can only agree with him to a limited extent. We have certainly found hemiatrophy in cases of tabes, but one should remember that it is in itself a rare affection, and that it exists more often independently than associated with tabes. In addition to the two cases mentioned on