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ORIGINAL ARTICLES.

THE POSSIBILITY OF THE EARLY DIAGNOSIS OF LOCOMOTOR ATAXIA BY THE EYE-SYMPTOMS.

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A review of the voluminous literature on the subject of the eye symptoms in tabes will impress one with the overwhelming importance of a careful and thorough inspection of the extrinsic muscles, the pupils and accommodation, and the optic nerve in the diagnosis of that disease. And this is particularly true in its earliest stages. A well marked case in its second or third stage presents no difficulties. The symptoms are of such a pronounced and individual character and so frequently found in association that "he who runs may read." But in that variable and usually extended period before the spinal sclerosis has developed its distinctive features of altered locomotion, incoördination, perverted and increased sensibility and paralysis, when a diagnosis is of greater value to the patient, an investigation of the ocular apparatus will not infrequently furnish reliable data from which positive deductions concerning the distant future can be drawn.

I will take up, in this paper, eye symptoms of the first or pre-ataxic stage only. They include disturbances of,

First.—The Extrinsic Muscles.—Graefe,¹ in speaking of sclerosis of the posterior column says, "paralysis of the eye-muscles may appear in a very early period of the disease, for we are bound to accept the conclusion that either independently of the disease process in the cord, or in connection with it, patches of degeneration analagous to those in the cord, form in the nerve nucleus or in the encephalic course of the fibres of the nerves supplying the muscles, and the paralysis can no more be attributed to the diseased patches in the cord than can the changes in the optic nerves which are found under similar conditions."

Schmeichler says² "the disturbance of the eye muscles in tabes consists in paresis of one or several. It begins, if it develops at all, almost always with the very first symptoms of the tabes, but at times it precedes the latter by many years."

E. Berger³ says, "in my cases (109) the far greater number of the ocular paralyses was in the stradium preactacticum."

Gowers⁴ says "paralysis of the external ocular muscles is also common in tabes and occurs in several forms. 1. Transient weakness lasting a few days

or weeks, and then passing away. 2. Permanent paralysis, complete or incomplete, of a single nerve or part of a nerve. Either form may occur at any stage, but the first is more common in the early and the second in the later stages of the disease."

Swansy says⁵ "in the premonitory stages of tabes dorsalis ephemeral partial paralyses affecting now one, and again another of the orbital muscles, may sometimes be observed."

Noyes⁶ says, "in special cases the paralysis is likely to be incomplete and not to be permanent. No other sign of spinal cord disease may occur for a long time and this symptom, while unsupported by others, will remain of doubtful significance."

R.P. Howard⁷ details the history of a case from which I extract the following paragraph: "Patient, an educated man fifty-four years of age, was of studious and very temperate habits, and free from evidences of syphilis, the possibility of which disease he positively denied. He married at thirty-three and was the father of sixteen children, the youngest eight months old. About the beginning of August, 1883, he suffered from severe headache, which he regarded as migraine, having been afflicted with that neurosis in his youth; at the same time he became irascible. Toward the end of the month strabismus, diplopia, and slight ptosis on the left side supervened, but these gradually disappeared in about six weeks. Some five months later deviation of the right eye, with diplopia, set in, and in about two months was followed by ptosis, which like the other symptoms, developed gradually, but unlike them, has proved permanent." Among the reasons for making the diagnosis of tabes, he gives "the early disturbance of the external and internal ocular muscles."

Roberts⁸ says, "certain so-called premonitory symptoms are usually observed, which may last for months or years, but these are really the early symptoms of the disease." Among them he mentions slight strabismus or ptosis.

In an article by Dr. J.A. Jeffries⁹ are found the following statements: "In tabes, as has long been known, there has been frequently a history of transitory diplopia during the prodromal period." "The origin of these fugacious paralysis is not known, but their significance, when combined with previous syphilitic infection is gravely suggestive of tabes to follow." "An eye paralysis, however simple it may seem, is always a just cause for suspicion of trouble to come, and demands a prompt and thorough examination of the patient."

Alexander,¹⁰ in speaking of paralysis of single branches of the 3d nerve, says, "since the ptosis frequently disappears we must attribute to a transi-

¹ Graefe and Saemisch, Bd. vi., p. 69.

² Arch. of Ophthal., Vol. xii, Nos. 3 and 4, p. 359.

³ Arch. of Ophthal., Vol. xix, No. 4, p. 485.

⁴ Diseases of the Nervous System, Am. Ed., p. 295.

⁵ Diseases of the Eye, 3d. Ed, p. 432.

⁶ Diseases of the Eye, p. 151.

⁷ Am. Jour. Med. Sci., Nov., 1889.

⁸ Practice of Medicine, 5th. Ed., p. 939.

⁹ Boston Med. and Surg. Jour., Oct. 27, 1892.

¹⁰ Syphilis and Auge, p. 137.

tory disturbance of circulation, which sometimes precedes by years the development of tabes."

Cornwell¹¹ states that "in tabes dorsalis the paralysis (of the eye muscles) often precedes the other symptoms some months, and at times, years." Hughlings Jackson found in six out of nineteen cases, diplopia to be the first symptom. . . . After it has fully developed and remained stationary for a time, it is apt to disappear.

Moore¹² says, "we must not omit the paralysis of the external muscles of the eye, that are so commonly seen in tabes, usually either the abducens or the motor oculi, and rarely the fourth nerve, which gives rise to various symptoms of dizziness, diplopia and strabismus. Graefe has pointed out that tabetic patients show little disposition to fuse the images in binocular vision, and this is taken as a sign of the central origin of the affection. These muscular paralyses are frequent in the early stages¹³ of the diseases; they are transient often in character, and this fact has not been very readily explained. Ptosis is also present when no other branch of the third nerve is involved."

Dr. Max Karger, of Berlin, in an analysis of 117 cases of locomotor ataxia, "finds at the commencement of the disease . . . slight and transitory paralysis of eye muscles."¹⁴

A sufficient number of authorities has been quoted to warrant the statement that ocular paralyses are among the earliest symptoms in the preatactic stage of tabes. The muscles affected are, in the order of their frequency, the external rectus, the levator palpebrarum, and finally other branches of the 3d nerve. Two features characterize early spinal paralysis and help to differentiate it from that and from other affections of the nervous system. 1. Its incompleteness; 2. Its transitoriness.

My first case was sent by Dr. Frank Woodbury, who kindly furnished the following history:

Case 1.—Mr. Blank, 61 years of age, of German parentage. Father died at about 60 years of age with catarrhal jaundice; mother died when he was a child. A sister older than himself died of phthisis; one brother died with pneumonia, the other with hepatic abscess after being afflicted with locomotor ataxia for several years. The patient is of highly nervous temperament and is excessive in his attention to himself and his health. As the result of a naturally good constitution and absence of all excesses, he has enjoyed remarkable immunity from sickness. He had croup as a child and had the other ordinary diseases of childhood; but his adult life has been exceptionally healthy. He has decided fondness for the pleasures of the table and as a result, has symptoms of chronic indigestion, constantly coated tongue, etc., which, however, causes him little annoyance. He has probably had some gastric catarrh for years and also post nasal catarrh, the latter having been cured by treatment. At one time he had middle ear inflammation, which permanently impaired auditory power in the right ear. Even previous to this time he had had tinnitus in this ear, which still persists, although the ears present no evidences of lesion. He has no vertigo; but on account of double vision is referred to Dr. Hansell for examination of the eyes.

Four months ago he noticed double vision. Examination revealed paresis ext. rect. of L., candle light 20' distance, exactly in front of face appeared double; images fused by prism varying, at different times, from 15° to 30°, base in; pupils contracted, L. more than R. and slightly irregular in outline. No synechia. Both contract on accommodation but not to light. Slight cortical capacity L. lens. Fundus of each normal excepting one vein in the left retina slightly tortuous. Nerves healthy in color and outlines distinct. Knee jerks exaggerated. No dizziness or ver-

tigo. Rhomberts symptom absent. No incoördination. No disturbance in sensibility. No pains.

Second—The Pupil.—A consideration of the pupillary symptoms in the earliest stages of tabes involves a study of their size, their inequality, and their reaction to light and to convergence. Normal pupillary movements depend, in the absence of ocular disease, upon the sensitiveness of those fibres in the retina and optic nerve which convey impressions of degree of light; upon a healthy condition of the nuclei of the pupils in the floor of the fourth ventricle and intercommunicating fibres; upon the soundness of the pupillary branch of the 3d nerve, from its nucleus to the iris, and upon the cilio-spinal centre in the region of 1st and 2d dorsal vertebrae and its communications with the cervical cavernous plexus, lenticular ganglion and the long ciliary nerves. Disease, therefore, of any one of these parts will cause abnormal contraction or dilatation, and will destroy reflex, consensual or associated action.

That such interference is found in the preataxic stage of tabes is shown by many authorities, and would probably be mentioned more frequently but that myosis may exist for years without attracting a patient's attention or bringing him to the notice of the neurologist, and no standard of size, under the same condition of light, seems to be accepted. It is, however, safe to assume that a pupil smaller than 2.5 mm. in diameter, in moderate intensity of light, is pathological.

Schmeichler (*loc. cit.*) relates the case of an "intelligent man who related to me that four year ago his attention had been directed to his narrow pupils; that he had followed their progressive narrowing since then before the mirror, and feared very much that he would become blind by their closure."

Berger found that six cases out of a total of 26 with myosis in stad. preatax. In the case of Mr. Blank, myosis is a conspicuous symptom. According to his own statement he has frequently had the size of his pupils brought to his attention during several years. A peculiarity noticed by Schmeichler of the tabetic pupil is the unusual length of time, after the instillation of a drop of atropine solution, before the pupil regains its previous size—from three to four times as long as in average normal eyes. I used one drop of an 8 grain solution of homatropine to dilate Mr. Blank's pupil to facilitate the examination of the fundus, and it was fully seven days before the pupil had returned to its previous size, five days more than is usually required. Again, the dilatation produced by a mydriatic is less in degree, rarely exceeding 4-5 mm. in width—a condition noticed under the homatropine mydriasis in Mr. Blank. A few writers speak of the inequality in the size of the two pupils in the early stages of tabes. Thus Berger says, "There is no marked difference in the relative frequency of inequality of the pupils in the different stages of tabes; it is but slightly more frequent in the initial stages."

Charcot¹⁵ has also called attention to this symptom.

Jessup¹⁶ says "unequal pupils are often found in cases of apoplexy, acute meningitis, chronic and acute alcoholism, general paralysis of the insane, locomotor ataxia."

Case 2.—Mrs. V., Dec., 1889. R. pupil contracted. L. normal,

¹¹ Am. Jour. Med. Sci., April, 1884.

¹² Journal of Nervous and Mental Diseases, 1888, p. 237.

¹³ Italics mine.—H. F. H.

¹⁴ Jour. Mental and Nerv. Dis., Vol. xv, 1888.

¹⁵ Rec. of Ophthal., Nov. 1887.

¹⁶ Oph. Review, July, 1888.

both respond to light and convergence. Dec., 1892. Had an attack of dizziness one month ago. Says she has rheumatic pain in feet. R. pupil contracted and does not respond to light, but contracts with convergence. L. pupil normal in size and reaction. Knee jerk on L. side almost absent. A positive diagnosis cannot be made, but the anisocoria, unilateral Argylle-Robertson pupil, limited color field with no diminution in the field for white, partial loss of knee jerk and fleeting pains in the feet would seem to point to a commencing sclerosis in the posterior columns of the cord. The Argylle-Robertson pupil is a well known symptom of tabes in all three stages. It is most common in the second, but exceeds in frequency even in the first stage, the normal. Good reaction of the pupil was retained until the paralytic stages in 109 cases studied by Berger, in only four. When that stage was reached all pupils were altered in their reactions.

The Argylle-Robertson pupil has been observed in progressive paralysis, in cerebral syphilis, and possibly in disseminated sclerosis but so rarely that its presence alone would almost warrant the assumption of a commencing post-sclerosis.

Third—The optic nerve.—The earliest sign of degenerative changes in the optic nerve is a diminution of the size of the color fields, then a loss of sensibility to green, with a relative decrease in the fields of other colors, a concentric limitation for white, and, finally, total loss of vision. In many cases it is quite impossible to recognize ophthalmoscopic changes until the field for white becomes limited. Hence the perimeter is more important than the ophthalmoscope in the early diagnosis of tabes. Perimetric measurements, to be accurate, must be conducted with great care, and often repeated with the same set of tests and under the same conditions of illumination. It must be remembered, too, that the ability to distinguish and perceive colors varies among different persons, and according to the method of examination. Ophthalmoscopically, the earliest stages of tabes may be suspected by the clear cut nerve with a pronounced excavation and distinctly seen lamina cribosa and irregularly surrounded by pigment, loss of color, particularly on the nasal side, dilatation and tortuosity of one or two veins, perhaps limited to one and usually the lower segment of the retina (Schmeichler) otherwise vessels normal in calibre and unaccompanied by the white lines which indicate a thickening of their walls. Authorities differ as to whether the temporal or nasal half of the disc shows first the discoloration and hence we must conclude that it is sometimes one, sometimes the other, and frequently the color of the entire disc is normal. By far the greater portion of the cases shows the L. optic nerve to be the first implicated. That atrophy of the optic nerve may precede general symptoms of tabes is abundantly proven. In seven of Berger's cases it was the first symptom, spinal symptoms appearing later. He quotes Charcot, Förster, Graves and Michel in relation to the largest interval of time between the commencing "deterioration" of vision and the earliest and most common symptom of spinal irritation—lancinating pains—and states it to be from two to twenty years.

In case 1, left eye, examination on October 5 and 6, showed decided limitation for green. It was not recognized outside of a circle 5° from point of fixation. Two months later, after salivation and iodide of potassium in large doses the field for green became almost normal in extent. Again, a large retinal vein running down and in, was dilated and tortuous. After treatment it became normal in calibre and course.

Case 3.—Mrs. G. In September of 1883, complained that for 18 months she has not been able to see to go about alone at night, excepting in the moonlight, nor could she see to sew on any colored material. V. R., 20-100, L. 20-20. Both discs white. Fields for white limited on nasal side. Color fields not taken. Pupils very small but responsive to light and convergence. In November of the same year commencing tabes diagnosed by Dr. Chas. K. Mills, to whom I took her in consultation. "She now complains of sharp pains in the legs. Rhombert's symptom." (Extract from my note book). Five years later the disease ended fatally. While an investigation of the signs of spinal disease, had it been made during the 18 months prior to the first visit to me, might have revealed commencing degeneration of the cord, the first and only symptom complained of during that period, was deterioration of vision due to atrophy of the optic nerves, well advanced in the R. A diagnosis of tabes might safely have been made at this period (1883) from the eye symptoms. Myotic pupils, scarcely responsive, white discs and limitation of the fields for white and probably for colors (since she hesitated in matching colors—the only test to which she was subjected).

Pershing, in an instructive article "Pre-ataxic Tabes Dorsalis with Optic Nerve Atrophy"¹⁷ relates two cases, which I quote in part.

Case 1.—C. L., printer, 38 years old. His mother died of "neuralgia of the stomach." One of the mother's sisters was insane, and the patient's only sister is very nervous and excitable. . . . syphilitic history. . . . V. began to fail in the L. in April, 1889, and grew steadily worse until January, 1890, when V in R. also became much impaired, L. seeming to improve. Transient ptosis on the left side was noticed several times. The knee jerks were found absent in November, 1889. Present condition November, 1890. Can barely see a hand close to face. Cannot count fingers. Pupils equal, moderately contracted; light reflex scarcely perceptible; accommodation reflex could not be tested. White atrophy of both optic discs. Knee jerks entirely absent even with Jendrassik's reinforcement. Plantar and cremasteric reflexes present; abdominal excessive. Station with eyes closed normal; stands on either foot. Walks backward perfectly well. No ataxia in arms or legs. . . .

Case 2.—E. P., 53, married. Mother died at 76, of softening of the brain. Father, brothers and sisters said to be very nervous. No syphilitic history. . . . Present condition Jan. 15, 1891. V. R. 5-6, L. 5-10. The fields are much contracted, both for white and colors, especially on the nasal side. Both optic papillæ are sharply defined and of a grayish tint. The R. pupil reacts well to light; the L. is contracted, irregular in contour, with light reaction barely perceptible. Both react well on convergence. The knee jerks are present, and are increased by reinforcement. . . . With eyes closed he can stand on either foot and walks well. There is no ataxia of arms or legs. (All other symptoms absent).

In determining the diagnosis, we must remember that the ocular symptoms illustrated by the cases recorded in the literature as well as my own three cases may be the result of other causes than posterior spinal sclerosis. The causes of ocular paralysis are traumatism, orbital disease, pressure upon the nerve in its course by gumma or other tumors, meningitis, dropsy of the third ventricle, nuclear disease, sclerosis of the brain, spinal disease, etc. Myosis may be medicinal (eserine, opium) irritative, in apoplexy, early stage of meningitis, inflammation of the eye, or paralytic, in affections of the lower cervical and upper dorsal spinal nerves, due to traumatism or disease. The Argylle-Robertson pupil is sometimes found in progressive paralysis but in the large proportion of cases it is a symptom of the first and second stages of tabes. Simple primary atrophy of the optic nerves (without signs of neuritis) is more frequently found in tabes than in all other organic nervous affections. To quote from Pershing's article, "Indeed primary bilateral atrophy alone makes the existence of tabes probable. Nettleship found that of 76 cases, 58 certainly and 10 more

¹⁷ Medical News, March 26, 1892.

probably were due to the same causes that give rise to degenerative changes in the spinal cord and brain. Peltesohn found that of 98 cases of spinal or cerebrospinal atrophy occurring in Hirschberg's clinic, 78 were cases of *tabes dorsalis*.¹⁸ Among cerebral causes may be mentioned, hydrocephalus, tubercular masses pressing upon the chiasm, softening and sclerosis of brain tissue. V. Graefe attributes 30 per cent. of all optic nerve atrophy to spinal causes—degeneration of the posterior and lateral columns from sclerosis or chronic myelitis, idiopathic or traumatic.

Various and contradicting opinions are offered by investigators as explanatory of the ocular paralyses associated with *tabes*. We may, however, accept as probably true, that the paralysis of the extrinsic muscles is, when transient, the result of vaso-motor disturbance and when permanent, of peripheral neuritis, and rarely nuclear, excepting in the last stages, that the myosis and Argyll-Robertson pupil are symptoms of disease of the cilio-spinal centre and that the optic nerve atrophy is a true primary degeneration of the nerve in no way different from that of the cord but not dependent upon it.

My conclusions may be thus summarized:¹⁸

1. That a transient external paralysis, for example, a ptosis or esotropia, in the absence of known cause might be an indication of incipient *tabes*.

2. The spinal myosis, the reflex to light wanting but to convergence preserved, monocular or binocular would warrant a strong suspicion of incipient *tabes*.

3. "Idiopathic" optic nerve atrophy, non-inflammatory, of unequal degree, often precedes by months or years, the appearance of the spinal symptoms of *tabes*.

4. That pareses of one or two extrinsic muscles, spinal myosis and Argyll-Robertson pupil, and incipient optic nerve atrophy, coexistent in a person of from 30 to 60 years of age, with inherited neurotic tendencies are forerunners in the majority of cases of posterior spinal sclerosis.

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HIP DISEASE.

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The term hip disease is used for any chronic inflammation in the synovial membrane of the hip joint, in the acetabulum, in the head, neck, or great trochanter of the femur, or in the soft parts immediately surrounding these, which, if allowed to progress without treatment, would ultimately present the symptoms of a tubercular arthritis. From a clinical standpoint—the standpoint from which this article is written—it does not appear possible to the authors to accurately diagnose the situation of the primary lesion in more than a few cases; nor is it possible to differentiate between a small tubercular

focus in the acetabulum and one in the head or neck of the femur, and often it is not possible, for some time, to differentiate between a condition which will ultimately result in the separation of the head of the femur, and one in which will end in the separation of the great trochanter. Further, we are not able to point out the difference between tuberculosis immediately within the capsule and the same state of affairs immediately without that membrane. From a clinical standpoint we can only approximately locate a lesion, which, unrestricted, ulti-

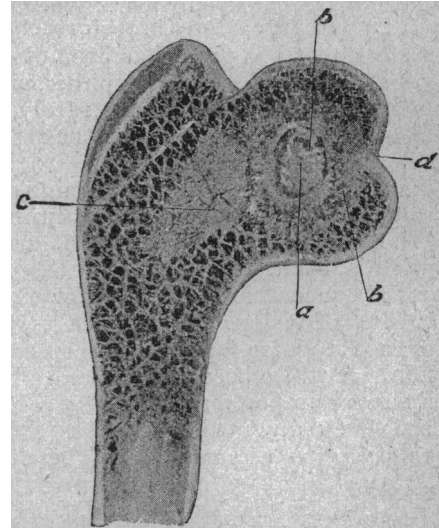


FIG. 1.—Primary tubercular infection in the head of the femur. *a* Cheesey sequestrum; *b*, tubercular infiltration; *d*, pressure groove from resting against the upper rim of the acetabulum, the head being partly displaced from the cotyloid cavity.

mately involves all or nearly all of the neighboring tissues; which under treatment usually recovers, leaving us still in doubt as to its precise habitat, and which wherever be its origin demands one and the same treatment. This then in a routine way we characterize by the general term, hip disease.

Tuberculosis at the hip-joint usually commences as a primary (Fig. 1) or secondary (Fig. 2) osteitis, and more frequently in the femur than in the acetabulum.

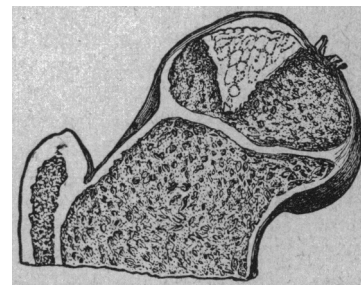


FIG. 2.—Secondary tubercular infection; cone-shaped cheesey sequestrum; cartilage lifted up as a vesicle.

bulum. It may undoubtedly commence as a primary tubercular synovitis, but many cases of synovitis follow from a traumatic rather than from a tubercular origin. Peri-arthritis inflammation seems to derive its origin more frequently from the infectious fevers than from other causes. The favorite habitat of these lesions in the children of syphilis is in the neighborhood of the epiphyseal line.

However the joint disease may commence, it is at the beginning, or very soon becomes tubercular. In the same way, wherever the lesion may be primarily found, if left untreated it goes progressively on,

¹⁸ If cases 1 and 2 should develop into unmistakable *tabes*, they will be illustrations of its earliest stages: if not, they must serve simply as examples of the eye symptoms common to that disease, and must be regarded as idiopathic or forerunners of some other central nerve affection than *tabes*. The conclusions are drawn from case 3, and those recorded in the literature.