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LATENCY OF ATAXIC SYMPTOMS IN CASES OF OPTIC ATROPHY.¹

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It has been long noted that optic atrophy, when present in locomotor ataxia, appears generally among the earlier symptoms. Indeed, given a case of optic atrophy without recognizable cause, the chances are altogether in favor of characteristic pains, bladder irregularities, or other symptoms, appearing later, which, even if not marked, will enable us to place the case under this category.

Charcot² emphasizes the fact that most women coming to the hospital with amaurosis develop sooner or later, in the majority of cases, ataxic symptoms, perhaps ten, perhaps fifteen years later. In one case cited, the blindness was followed in ten years by shooting pains and girdle pains, these symptoms remaining stationary for ten years more, when appeared symptoms of muscular incoördination.

Gowers³ mentions cases of his own in which optic atrophy preceded ataxia by sixteen and twenty years, and quotes a case reported by Buzzard,⁴ in which atrophy existed fifteen years, associated only with lightning pains and loss of knee-jerk.

In his recent text-book, Gowers⁵ has stated that atrophy of the optic nerve so universally appears early

¹ Read before the Boston Society for Medical Improvement, April 8, 1889.

² *Leçons sur les Maladies du Système Nerveux*, 1880, t. ii, p. 41.

³ *Gowers' Medical Ophthalmoscopy*, p. 164.

⁴ *Brain*, 1878, No. 2, p. 168.

⁵ *Diseases of the Nervous System*, 1888, p. 293.

in the disease, that, after the ataxic gait is fairly established, it is of rare occurrence. Conversely, he states that when optic atrophy has become developed, it is common for the other symptoms of locomotor ataxia to remain in abeyance. To use his own words, "In a large number of cases, the ataxia never comes on, the spinal malady becoming stationary when the nerve suffers." If this is true, its bearing on the question of prognosis is of considerable import. It is certainly something to be able to assure a patient, at any stage of this disease, of his probable, or even possible, immunity from any of its distressing symptoms. In the matter of diagnosis, also, the subject is worthy of consideration.

I have looked through the records of sixty-six consecutive cases of locomotor ataxia, — fifty-two seen in the Neurological department of the Massachusetts General Hospital, and fourteen in private practice, — in the hopes of throwing additional light on this branch of the subject. The result is as follows: Out of the sixty-six cases, the diagnosis optic atrophy was found in fourteen; and six more had decided loss of eyesight. In these six there was no description of the optic nerve given in the notes, which prevents their being classed with certainty under the cases of atrophy, although undoubtedly some, and perhaps all, of them belong there. Out of the fourteen cases of optic atrophy, the degree of incoördination was mentioned in eleven. In three of these eleven, there was no ataxia; in five, it was slight; in three only was there marked ataxia. In the eight cases in which the ataxia was slight or wanting, the duration of the disease was respectively, two, three, four, seven, eight, ten, twelve and twelve years. In nine of these fourteen cases, the knee-jerk was noticed. It was present

in one or both legs, in four out of the nine; and in one other case, where the loss of eyesight was so considerable as to leave little doubt that optic atrophy was present, the knee-jerks were normal. In all of the cases the disease was well advanced. These figures, while far too incomplete to be regarded as statistical, are still of some interest, sufficiently so to encourage further investigation.

The noticeable features are the large proportion of cases in which the knee-jerk was retained in one or both sides, and the large proportion in which ataxia was either absent or very slight. With regard to the ataxia, this has been already considered by neurologists as a symptom not necessary to the diagnosis, hence the tendency to restore the name "tabes dorsalis" in preference to locomotor ataxia. The point to which I would call attention is that to which Gowers has practically alluded, namely, that it is the cases in which optic atrophy is present in which we may expect to find the absence or indefinite postponement of ataxia. Again, with regard to the preservation of the knee-jerk, the loss of knee-jerk in locomotor ataxia is, as a rule, one of the earliest and most constant symptoms, a fact which lends considerable significance to its preservation in four out of nine (probably five out of ten) cases where optic atrophy was present. The practical bearing of these facts is to assist us in both diagnosis and prognosis. As regards diagnosis, they would lead us to place with confidence under the head of locomotor ataxia certain cases of optic atrophy which we may have hesitated to place there on account of the presence of knee-jerk and absence of ataxia. As regards prognosis, they would lead us to predict a comparative latency of the motor symptoms of the disease where optic atrophy has become pronounced.

The constancy of one symptom was noticeable in

the cases with optic atrophy as in those without it, namely, the Argyle-Robertson pupil. This symptom, together with characteristic pains, may, it seems, be looked for early in this class of cases, however latent the motor symptoms. If the Argyle-Robertson pupil were dependent on optic atrophy, it would be expected particularly in these cases; but its constancy in the other cases shows it to be independent, at least in general, of the lack of conducting power in the optic nerve, a fact already well recognized.

It is worthy of note, that, out of sixty-six cases taken indiscriminately, there were fourteen with the diagnosis of optic atrophy, and several more in which it was probably present. Statistics on this point vary greatly. Erb found eight cases in about seventy, while Topinard noted disturbance of vision in forty-nine out of one hundred and two, and Cyon found sixty cases with amblyopia and amaurosis out of two hundred and three. The statistics of more recent observers also vary markedly. The figures of oculists would naturally run higher than those of neurologists. Gowers states that the proportion is much less than is generally believed, certainly not exceeding one case in ten.

I have selected three cases to report this evening, premising that they are presented, not as being in any way unique, but as fairly representing the class of cases under consideration.

CASE I. J. C., aged forty, married, an engraver. Referred to me at the Massachusetts General Hospital, by Dr. Standish, under whose care he is at present. Four years ago he began to suffer from recurring pains in both knee-caps, without swelling or redness, and not increased by movement. He describes the pains

as shooting and stinging, lasting only a few minutes. Similar pains appeared later in the thigh and in the toes of the left foot. Six months ago he noticed failure of eyesight, which rapidly progressed to almost complete blindness within three months. There have been bladder irregularities, but no absolute incontinence. There have been no laryngeal nor gastric crises. He tires easily in going up stairs, and has done so for four or five years. He has never noticed any staggering or numbness. He never saw double.

Physical examination shows absence of tendon reflex on the left, a trace on the right. Both pupils react to accommodation, neither to light. Both pupils are small, the left smaller than the right (said by the patient always to have been so). There is no trace of impairment of any variety of sensation, not excepting muscle sense. There is no special loss of nutrition. The gait is perfect, and the patient stands on either foot without swaying, also with both feet together and the eyes shut. Ophthalmoscopic examination by Dr. Standish, as well as by Dr. Wadsworth, who saw him at the hospital, shows characteristic optic atrophy. The vision is now so reduced that he can only count fingers with the right eye at four feet; vision in the left eye being 0.4, not improved by convex or concave glasses.

CASE II. Mr. —, aged forty-seven, married. Referred to me for examination by Dr. Chandler. He has also been examined by Dr. Wadsworth and Dr. Prince. Has had weakness and pains in the back since 1864, when he was injured by the blowing up of a vessel. He first noticed trouble in sight in 1879, which increased steadily. He can now with the left eye, barely count fingers at a foot: with the right eye, there is perception of light only. Since 1882 he has had pains, sometimes very severe, perhaps three

or four a year; lighter attacks of pain often, lasting ten or twelve days. These pains are generally in the back of the legs and in the hips. During the past eight or nine years a pain runs up the back to the head. This pain comes quickly and recurs perhaps a dozen times a day. Sharp, throbbing pains appear in the legs, lasting from an hour or two to several days. The locality of these pains varies; for example, one day in one foot, one day in the other, again about the knee, no swelling or redness accompanying the pains. In 1885, he saw double for six weeks. There has been no bladder trouble, no laryngeal or gastric crisis, no numbness, nor weakness in the legs; occasionally a slight tendency to stagger. Twelve years ago he had what was considered a chancre, not followed by eruption or falling out of hair.

Physical examination shows the knee-jerk absent in the right, present in the left. There is no swaying with the feet together and eyes shut. He stands well on either foot with the eyes open, and fairly well on each foot with them shut (which is a difficult test). The lightest touch is felt everywhere. The plantar reflex is present. The right calf measures $15\frac{1}{4}$ inches, the left $15\frac{3}{8}$ inches. The right pupil is larger than the left; neither reacts to light, both to accommodation. There is external strabismus in the right eye. The left field is cut off nearly to the horizontal meridian, and moderately contracted in other directions. Ophthalmoscopic examinations by Drs. Wadsworth and Chandler shows gray atrophy of the optic nerve, more pronounced in the right. The pulse is 106. There is no scar on the penis, but the frænum is wanting (said to have been burnt off).

CASE III. A. D., aged thirty-one. Referred to me for an opinion by Dr. Chandler, and afterwards

by his attending physician, Dr. Cutter, of Leominster. He has also been examined by Dr. Knapp. He first noticed trouble in sight seven years ago, and has steadily lost vision up to the present. The only other symptoms complained of, are occasional pains in legs and feet, occasional dizziness, occasional attacks of gastric pain and vomiting (crises?), and a tendency to tire rather easily, with occasional slight unsteadiness. He also states that his water occasionally escapes, and that he sometimes holds it all day without desiring to pass it. There is no definite specific history, but he states that there was considerable falling out of hair after an attack of gonorrhœa twelve years ago. No sore was noticed, and no other secondary symptom.

Physical examination: There is a slight trace of knee-jerk on the left, none on the right. The pupils are unequal, and react to accommodation, not to light. There is no impairment of sensation; no wasting; slight ataxia. Dr. Chandler finds marked optic atrophy. With the right eye he cannot count fingers. The vision in the left eye is .9.

In these cases will be noted not only absence or partial absence, of ataxia, but also that of sensory impairment, at a stage when these symptoms would be expected. It will also be noted that the knee-jerk was not entirely lost in either of the three cases. It is also noticeable that nutrition is unaffected, the patient in one case (Case III) especially, being a stout, well-nourished man, with calves measuring over fifteen inches. This is an exceptional condition in well-advanced locomotor ataxia, although this disease is not characterized by distinct wasting of muscles and groups of muscles, as are diseases affecting the peripheral nerves or the anterior cornua of the cord. The importance of these points in diagnosis is illustrated

by the fact, that in two of the cases, considerable doubt has arisen in the minds of consultants as to the nature of the underlying cause of the optic atrophy.

Acting upon the suggestion offered by Dr. Webber in the discussion of this paper, I append a brief abstract of the symptoms in the cases of optic atrophy.

CASE IV. M. R. Numbness and darting pains for two years; delayed micturition; knee-jerk absent; pupils unlike. Argyle-Robertson pupils; pulse 102; vision blurred, field normal; discs opaque, with sharply defined vessels, especially right.

CASE V. T. M. Atrophy of optic nerve, twelve years; staggering during past eight weeks; Argyle-Robertson pupils; knee-jerk normal. During seven or eight years, beginning about fifteen years ago, patient was subject to attacks of vomiting about twice a week.

CASE VI. Pains, twelve years; pupils unlike, Argyle-Robertson pupils; knee-jerk absent; slight staggering with closed eyes; optic atrophy, (*b*) $V = \frac{1}{3} \frac{1}{6} (b)$.

CASE VII. A. F. Darting pains in legs, seven years; bladder troubles, one year; diplopia and loss of sight, seven years; complete blindness, two and one-half years; vomiting, six years; pupils unlike, Argyle-Robertson; knee-jerk absent; slight ataxia; no anaesthesia.

CASE VIII. C. G. (female). Pains, eight years; failing sight, eight months; bladder irregularities; Argyle-Robertson pupils; loss muscle sense and slight loss tactile sense in middle toes (*b*); slight unsteadiness;

stands fairly well with feet together and eyes shut; ophthalmoscopic examination by Dr. H. W. Bradford shows optic atrophy of both eyes.

CASE IX. W. F. B. (female). Shooting pains, seven years; weakness in legs, several years; staggering, one year; loss of muscle sense; loss of vision, one year; cannot count fingers with left eye; with right eye, vision one-third.

CASE X. N. B. Pains and ataxia, two years; loss of sight lately; knee-jerk absent; tabetic knee joint; slight optic atrophy.

CASE XI. P. C. Pains, a number of years; slight failing, two years; slight bladder irregularities; vision almost lost in left eye, greatly diminished in the right; color sense almost absent (cannot distinguish light blue from red even in the centre of the field).

CASE XII. C. F. Pains in legs; Argyle-Robertson pupils; pupils unlike; scarcely any ataxia with eyes closed; well-marked atrophy of optic nerves.

CASE XIII. F. K. Failing sight, two or three years; now, perception of light only; pains one year; Argyle-Robertson pupils.

CASE XIV. Gradual failing sight, one year; Argyle-Robertson pupils; pupils small; very slight ataxia; shooting pains in legs; well nourished; knee-jerks absent; marked optic atrophy both; (V. O. D. = $\frac{1}{80}$ V. O. S. $\frac{2}{10}$) nerves white; edges of disc well defined; retinal arteries and veins decreased (Dr. Cheney).

The description of the optic nerves in a few of these cases is such as to leave the diagnosis not absolutely certain. In the majority, however, the loss of sight is too great to admit of question. Whatever may be deducted from the number on account of uncertainty may be offset by the cases in which complete records are wanting, and among which exist, probably, certain cases of atrophy. In any event, the character of the investigation precludes reaching absolute statistics.

