

watch only within half an inch; ordinarily loud voice at ten feet; left ear hears the watch only in contact, or ordinary voice at four feet; speech broken, Dutch-English, probably normal; vision, right eye counts fingers at two feet, left eye counts fingers at twelve feet; urine normal. Ophthalmoscopic examination shows intense double optic neuritis, with hæmorrhages, and infiltration of retina and discs. There is probably tumor of the cerebellum involving the tubercula quadrigemina.

June 25th.—The left pupil a little small, reacts imperfectly to light. Right pupil half the size of left, reacts very imperfectly; percussion on the temples hurts a little, on the forehead less; head twenty-four inches in circumference.

July 15th.—Last evening and this morning refused to take his medicine, saying there was something in it to poison him.

July 24th.—Discharged at his own request.

Case 3.—Sarah B., æt. 36, single, admitted into St. John Hospital February 9, 1881. First noticed two months ago that the left eye could not see daylight. Three days ago the right eye failed so much as to be unable to read ordinary type. Three years since was confined to bed for six weeks with constant, severe pain in the top of the head. Was at times very chilly and again very hot and thirsty. The head was very hot. No vomiting. This pain was finally much relieved by the application of iced-water to the head, but has been very severe most of the time since, up to the present. Memory is failing, and she feels stupid. Pulse eighty, weak; sleeps very badly; appetite poor; no history of syphilis; mother and one brother died of phthisis. With right eye can count fingers within six inches. The left has no perception of light. The fundus of the right eye normal in appearance. Advanced optic atrophy of the left. The atrophy probably neuritic, and the result of meningitis.

March 26th.—Has been treated with potass. iod., potass. brom., and hydrarg perchlor. and cold water to the shaved head. Right disc normal in appearance. Discharged relieved of pain, but vision about the same as upon entering. Returned to the hospital on August 17, 1881, with headache and nausea. The sight of the right eye began, three weeks ago, to get worse, and she cannot now distinguish light with either eye. Pupils large and immovable. There is now atrophy of the right optic disc, as well as of the left. Pulse 104, small and weak. R. Hydrarg. submur., gr.  $\frac{1}{2}$  every hour.

Aug. 20th.—Severe pain in the head continues. R. Potass. iod., grs. x, t. i. d.

Aug. 22d.—No pain in the head. A prominence noticed over the left frontal eminence (osteoma).

Oct. 27th.—Discharged with perception of light with right eye only, the head-ache absent for several weeks, and the frontal prominence much reduced.

Remarks.—The prompt relief of the head-ache and the frontal prominence by potass. iod. points to a specific origin of the atrophy of optic discs in spite of the absence of a syphilitic history.

Case 4.—Concussion of the brain, and probably of the retinae, followed by atrophy of the discs. Rev.

D. B. consulted me Oct. 5th, 1883. Last April was thrown backwards from a buggy, striking the ground with the back of his head. After recovering in a few minutes from insensibility, felt nausea. The injury did not confine him to the house. Since then has had a feeling of pressure on the front of the head, and sometimes behind the eyes. Every few nights is sleepless and has tinnitus aurium, "rain-like sounds." The sight began to fail a few days after the accident, and in a fortnight he was unable to read. Has photopsies. Vision, right or left eye,  $\frac{1}{16}$ . Blueish white atrophy of both discs. R. Strych. sulph., gr.  $\frac{1}{16}$  t. i. d. R. Potass. brom., grs. xv and repeat p. r. n. in evening. The patient was not again heard from.

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### A CASE OF SPINAL ATAXIA WITHOUT LOSS OF SENSATION AND WITH INCREASED PATELLAR-TENDON REFLEX. A CONTRIBUTION TO THE STUDY OF SPINAL ATAXY.<sup>1</sup>

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The case of ataxia which I am about to describe is one of exceptional interest and especially so when studied in connection with Dr. Smith's very interesting series of cases of hereditary ataxia, with which it has many points of similarity. It resembles hereditary ataxia in the age of the patient (15 years) at which the symptoms developed, the absence of anæsthesia and in fact of all sensory disturbances, the presence of anterior curvature of the spine and nystagmus, and the rapidity with which the symptoms developed. It differs on the other hand from this form of ataxia in the facts, of the freedom, thus far, of the rest of the family from the disease, and in the presence and indeed exaggeration of the patellar-tendon reflex. It differs again from ordinary tabes in this last particular, in the presence of nystagmus, in the absence of all sensory disturbances of every kind, of all cerebral, ocular (pareses) and bladder symptoms. It may be a question as to where it should be placed, and indeed the presence of the tendon-reflex might be thought to exclude it entirely as tabes, but still I am strongly inclined to believe, for reasons which I shall presently state, that it belongs to the systemic spinal ataxias. Another and not the least interesting point connected with the case is the bearing which it has on the disputed question of the pathology of ataxia. There is still a difference of opinion existing among neurologists as to the cause of ataxia. Some holding that it depends purely upon the loss of sensation, (including sense of touch, muscular sense, etc.); others that it is a purely motor disturbance due to an impairment of the motor coördinating impulses. The sensory theory would be disproved if, as Erb has pointed out on the one hand, cases of complete anæsthesia should be discovered in which no ataxia existed, and on the other if well-marked ataxia should

<sup>1</sup>Exhibited before the Section for Clinical Medical Pathology and Hygiene of the Suffolk District Medical Society, June 9, 1885.

be found with no loss of sensation. A few cases of this latter class have already been reported, and one in particular by Erb,<sup>2</sup> which will be mentioned more in detail later. In this case there was also complete absence of anæsthesia in every form, while the tendon reflexes were present. These cases show that though loss of sensory impulses may possibly by themselves induce the loss of the power of coördinating movements, ataxia must also have a motor origin and probably be due to a disturbance of the motor coördinating tracts. The complete overthrow of the sensory theory would be accomplished by the occurrence of a case of total anæsthesia without ataxia.

Mary D., 17 years of age on September 23, 1885. Her present illness is dated by her mother from January 3, 1884, when the girl came home from school complaining of weakness of the legs and malaise. She has never been to school since. The mother admits, however, that during the preceding two months, the daughter had not felt well, and complained of weakness. I first saw the patient in the early summer of 1884, and have examined her occasionally from time to time since. As her symptoms have remained, with the exception of one or two particulars, essentially the same during all this time her present condition will be here given, stating at the same time such changes as have occurred in the progress of the disease.

*Gait, muscular power, etc.* The patient can hardly walk or stand without the aid of support. She habitually uses crutches, and only can be made to walk unassisted after persuasion, when left to herself she easily topples over though she can manage to get across the room. It is evident that she has not the typical ataxic gait, but rather scuffs along with her feet, taking short, slow, careful steps. She gives at first the impression of having advanced paresis of the legs. But further examination showed this view to be erroneous; for when supported laterally so that she cannot fall, she can lower or raise herself on either leg. When told to flex and extend her legs against resistance, it is manifest that their muscular strength is good, though I think they are not as strong as normal. When, again, lateral support is given to her by placing the hands firmly against both her sides, and her courage is restored, she can raise her feet and walk fairly naturally. When walking, or when standing with her heels and her toes together but eyes open, she topples over after a few seconds and must be well balanced to stand at all. With her eyes closed she tumbles somewhat more readily. She stands and walks with her feet wide apart. She does not reel, or pitch in one direction more than another, but rather tumbles like any inanimate thing which has lost its equilibrium. In other words there exists *static ataxia*,

There also exists incoördination of the motion of the legs, but in a minor degree. When told to describe a circle on the floor with the toe, a most irregular figure results. She cannot accurately touch with the toe any small object, but reaches it by moderately zig-zag movements. It is a question whether more of an ataxic gait would not be shown if she could maintain the equilibrium, and could walk with

less caution and more quickly. The left leg is more ataxic than the right. The hands are also ataxic; the left decidedly, the right slightly. This is shown by difficulty in picking up small objects, buttoning her clothes, touching the top of her nose with her eyes shut.

*Sensation.* There is absolutely no loss of the senses of touch, pain, or temperature. When I first saw her, about fourteen months ago, I thought there was slight diminution of the muscular sense, not very marked, but still sufficient to be detected. I am unable, however, to discover any at all. She recognizes the slightest passive move of her legs, can accurately locate their position, and can feel the slightest faradic contraction of the muscles. Sensation as tested by faradism is also normal. There have been no lightning pains or girdle sensations. When first seen in 1884, she said she had a slight feeling of numbness in legs, but this has disappeared. At that time there was no objective anæsthesia.

*Reflexes.*—The patellar-tendon reflexes are not only present, but exaggerated. No ankle clonus; the plantar reflexes can only with difficulty be obtained. The lumbar reflexes are lively.

*Muscles.*—There is no atrophy or rigidity. There is no disturbance of the functions of bladder or bowel, no tremor or local paralysis, excepting the possible weakness of legs above mentioned.

*Nystagmus* exists when the eyes are turned to right and left, but none noticeable when the eyes are at rest. Last June I did not notice the nystagmus, and do not think it was present at that time. I saw it for the first time at the next examination in September.

There exists marked *anterior curvature of the spine* in upper dorsal region. There have been no psychological symptoms of any kind. No vomiting, headache, vertigo (excepting slight dizziness when she attempts difficult feats, and loses her balance).

The eyes were kindly examined for me by Dr. Charles F. Williams in June last, and within a few days by Dr. O. F. Wadsworth. Both report nothing abnormal in the fundus of either eye. As Dr. Williams did not observe the nystagmus, there is additional ground for not believing it to be present in June.

There is no syphilitic history, and no neurosis in the parents or collateral branches can be discovered. Up to date, there have been eight children in the family. The youngest is at time of writing only five days old, and one died in infancy. Of the other six children, I have personally examined four and found all but the subject of this paper healthy. The mother describes the remaining two free from all disease.

As Mary D., first complained of weakness in November or December of 1883, she was little more than 15 years old when the first symptoms developed.

That the diagnosis in this case is not placed beyond doubt, must be frankly admitted. But, it seems to me that it is highly improbable that the case is one of cerebellar disease, in view of the character of the drug, and of the absence of all headache, vertigo, vomiting, convulsive attacks, and the normal condition of the eyes.

Of the spinal diseases, the only ones to be thought of are multiple sclerosis and Friedreich's disease.

<sup>2</sup>Neurologisches Centralbl. No. 2, 1885.

On first seeing the case, I was inclined to regard it as one of the multiple sclerosis because of the lively patellar-reflex; but now, after watching the case for nearly a year and a half, I believe the lesion will prove to be that of Friedreich's disease. This view is based on the absence of the peculiar tremor of multiple sclerosis, the absence of ankle clonus, local paralysis (especially of the ocular muscles), and of spastic rigidity of the muscles; the absence of all cerebral symptoms, as vertigo, headache, psychical disturbances, etc. Furthermore, it is as difficult to explain the increased patellar reflex by a multiple sclerosis as by a posterior sclerosis; for any lesion of the lumbar enlargement which would cause the ataxia, would abolish the reflex, and if the lumbar enlargement is not involved, the increased reflex is compatible with a posterior sclerosis. This will be referred to again in speaking of the pathology of the disease.

It may be well to recall here the features which distinguish Friedreich's disease from ordinary tabes.<sup>1</sup> In the first place, there is the tendency to attack many members of the same generation, in the same family, while ordinary tabes does not run in families. Then Friedreich's disease attacks early in life, while ordinary tabes is a disease of middle age or of later life. Out of thirty-six cases, in fifteen the first symptoms came on at 10 years of age or under, and thirty-two were 20 or under.

In ordinary tabes, pain and anæsthesia are prominent and almost constant symptoms; in Friedreich's form, disturbances of sensibility are usually insignificant, come on late in the disease, and often are entirely absent. Other peculiarities of the latter form, are the rapid extension of the ataxy from the feet to the hands, impaired articulation, nystagmus, and curvature of the spine. On the other hand, many symptoms which are common in typical tabes are absent in the hereditary variety,—such as lightning pains, anæsthesia, disturbances of the bladder, transient paralysis, loss of pupillary reflex, optic atrophy, visceral lines, etc. (Omerod). In both, the patellar-reflex is absent. It will be noticed that the symptoms in Mary D's case present a typical picture of the hereditary form, excepting that none of her brothers or sisters have thus far been attacked (all are younger than the patient), and the patellar-reflex is present and rather exaggerated.

There is the youth of the patient, the rapid progress of the ataxia spreading to the arms within six months, the nystagmus, the spinal curvature, and the absence of the other symptoms commonly met with in ordinary tabes. Difficult articulation we should hardly expect to meet at this early stage of the disease. The fact that no other case has thus far occurred in the same family, will hardly militate against this case being regarded as Friedreich's form of tabes, considering the ages of the other children, and also the fact that a case has already been recorded which stood alone in the family. The absence of sensory disturbances is peculiar to this form of tabes.

As to the retention of the patellar-tendon reflex, it is a significant fact that it is not a very rare thing in

ordinary tabes for the patellar-reflex to be preserved, and two cases have been observed, one by Ross and one by Erb, in which the patellar-tendon reflex was increased. But more than this, the clinical evidence in favor of regarding the case of Mary D., as one of Friedreich's ataxia, (posterior sclerosis), is still further strengthened by three observed cases of ataxia in one, of which *normal sensation* was combined with *normal patellar-reflex*, and in the other two with *exaggerated patellar-reflex*. These cases are so valuable that it will not be out of place to briefly mention them here. The first case is reported by Erb.<sup>2</sup> It was that of a man 52 years of age, who, when he came under Erb's care, exhibited marked and typical ataxia. When he walked side by side with a patient, with typical tabes, no difference in the gait could be distinguished. There had been no lightning pains; there was no paræsthesia or anæsthesia of any kind and no girdle pains; the pupils and ocular muscles were normal, there were no bladder symptoms. There were also no cerebral symptoms of any kind. The patellar-tendon reflexes were present. For the same reasons I have given above in the case of Mary D., Erb regarded this case as one of spinal ataxia, though unwilling to commit himself as to the exact lesion of the cord.

This seems to be the place, before giving the other two cases, to speak of Kast's<sup>3</sup> two cases, in which with slight anæsthesia and no loss of muscular sense at all, there was exquisite ataxia, while the patellar-tendon reflex was present.

The first of these cases was one of myelitis from compression following fracture of the vertebræ caused by a fall. There was complete paraplegia, slight loss of sensation, but gradual motor recovery in eight weeks. When full motor power in the legs had returned, ataxia was developed. Both knee-jerks were readily obtained, but there was no ankle clonus. The plantar reflexes were active. In the right leg ataxia and loss of sensibility to touch and pain were more marked than in the left, but there was no loss of muscular sense; the sphincters were not affected.

The second case was one of acute transverse myelitis, with complete motor and almost complete paralysis of both legs to touch, temperature and pain. In this case also, on return of motor power, decided ataxia developed, and persisted even after complete return of motor power. The knee-jerk was well marked and there was ankle clonus on left side but not on the right.

In these two cases, a stage intervened between the complete motor paralysis and return of muscular power, in which, with almost complete motor power, there were considerable defects of coördination and ataxia. I have not been able to obtain Kast's original article, and therefore cannot speak positively of the exact amount of sensory disturbances present, but Erb, referring to them, says: "There was the most exquisite ataxy, without very marked disturbance of sensibility, and especially without any loss of the muscular sense."<sup>4</sup>

<sup>1</sup>Ross, Diseases of the Nervous System.

<sup>2</sup>Neurolog. Centralbl. No. 2, 1885.

<sup>3</sup>Schmidt's Jahrbücher, 1883. B. 200, Brain, Vol. vii, p. 553.

<sup>4</sup>Neurolog. Centralbl. No. 2, 1885.

<sup>1</sup>A very excellent digest with a table of all the cases up to date is given by Omerod in Brain, Vol. vii, page 105.

The two cases above referred to, wherein normal sensation was combined with increased patellar-reflex, were reported by Seeligmüller<sup>1</sup> as cases of Freidreich's disease. They were two brothers whose parents were first cousins, and whose mother and maternal aunt were highly neurotic. Both brothers had locomotor and static ataxy, in one the static being predominant. In both there was absolutely no loss of sensation, and there existed *increased* patellar-reflex, and nystagmus. The plantar reflex was absent in both; the abdominal reflexes were present. There was no ankle clonus, and no disturbance of speech.

Freidreich, to whom Seeligmüller sent his cases for an opinion, expressed a doubt as to whether they were true cases of his disease, presumably in view of the presence of retained patellar-reflex and the presence of psychical symptoms which existed in both, namely: forgetfulness and dreamy states in one, and forgetfulness, migraine, dreaminess, and inverted sexual passions in the other. But, as Seeligmüller points out, the father, who was otherwise perfectly well, had the same forgetfulness, and, as Omerod remarks, the connection between tabes and paralysis of the insane is well known. Even if there should have been some cerebral disease, it would scarcely account for the spinal symptoms. It seems to me, all things considered, that Seeligmüller was more likely right.

Not the least interesting question connected with the case of Mary D. is the seat of the lesions in the cord. It is a remarkable fact, considering the grouping of the symptoms, that out of seven autopsies<sup>2</sup> of Freidrich's disease thus far recorded<sup>3</sup> (including the case reported by Kahler and Pick), in six there was found sclerosis, not only of the posterior columns, but of the lateral as well. In several there was sclerosis of anterior columns. Of these seven cases, the condition of the tendon-reflexes was, unfortunately, noted in only three; in all three the patellar-reflex was absent, though there was lateral sclerosis as well.

How are we to explain the absence of all symptoms of lateral sclerosis in these cases on one hand, and, on the other hand, of the presence of the knee-jerk in exceptional cases of ordinary tabes, and of the exaggerated jerk in Seeligmüller's cases and mine? The key to this question I believe is given by the autopsies of other cases of systemic spinal disease, not Freidreich's, in which a combination of systemic lateral and posterior sclerosis existed. An excellent digest<sup>4</sup> of these cases has also been made by Omerod, who has collected twenty cases in all. Of these, the patellar-reflex was lost in eight and exaggerated in six cases, reported by Strümpell (two cases), Westphal (one case), Raymond (one case), Rabasin (one case), and Déjérène one case. Of these, the most instructive was Déjérène's.<sup>5</sup> The symptoms were those of tabes, namely: slight locomotor but marked static ataxy; *exaggeration of patellar-reflex; ankle clonus*; paresis of legs; anesthesia and analgesia of the legs, distributed in patches. Death from broncho-

pneumonia. At the autopsy there was found chronic posterior spinal meningitis, atrophy of posterior nerve roots and cutaneous nerves, sclerosis of lateral columns in lumbar and dorsal regions. This sclerosis was wedge-shaped, the bone reaching the periphery and occupying the posterior part of the lateral columns, but stopped before reaching the posterior bone (wrist cerebellar and crossed pyramidal tract?). The posterior columns were sclerosed the whole length, *excepting in the lumbar enlargement*, which was nearly unaffected.

In the other four cases, all of which, however, presented more decided spastic symptoms, the disease of the posterior columns did not reach the lumbar enlargement. These cases, therefore, support Westphal's opinion, based on his one case where the spastic symptoms were present, and several other cases where they were not—that if the posterior sclerosis involved the lumbar enlargement, the lateral sclerosis was manifested by paresis only, without rigidity, and the patellar-reflex is absent, but if the posterior sclerosis did not extend into the lumbar enlargement, the symptoms of lateral sclerosis predominate.<sup>1</sup>

If the case of Mary D. (the subject of this paper), is properly classified as one of Freidreich's disease, we may assume that the disease has not to any great extent involved the lumbar enlargement; and thus explain the fact that the patellar-reflex is not abolished. Finally, the exaggeration of the latter may be owing to the same conditions which cause an increased patellar-reflex in transverse myelitis when the lesion is above the lumbar enlargement; or, if we suppose the same anatomical changes to be present which were found in the six autopsies of Freidreich's disease mentioned above, it is possible that the lateral columns are slightly affected. This would account for whatever amount of paresis is present.

## SUN-BLINDNESS.\*

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The danger of blindness from the direct rays of the sun, which appears to have been generally recognized not only by the medical profession but also by the laity of the past generation, is apparently unrecognized to-day. During the recent eclipse, many might have been seen looking at it with the naked eye. It is not uncommon for patients to tell the physician, as a proof of strong vision, that they can look steadily at bright lights, and even at the sun without blinking. Children not infrequently undertake competitive trials to see which can gaze longest and most steadily at the sun.

In many of the recent smaller works on ophthalmology the subject of sun-blindness is not mentioned. Nettleship, Lawson, Williams, Schweiger and De Wecker refer to the possibility of blindness being caused by flashes of lightning, by the bright glare

<sup>1</sup>Archiv. für Psychiat., etc. Bd. 10, S. 222.

<sup>2</sup>This does not include Dr. Smith's case.

<sup>3</sup>Abstracts of this article will be found in Omerod's Digest.

<sup>4</sup>Brain, July, 1885.

<sup>5</sup>Archiv. de Physiolog., Nov. 16, 1884.

<sup>1</sup>Omerod, loc. cit.

<sup>2</sup>Read before the Chicago Society of Ophthalmology and Otology, August 11, 1885.