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FAMILY SPASTIC PARALYSIS OF SPINAL TYPE ON A HEREDOSYPHILITIC BASIS

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It is only recently that clinical histories have been published concerning patients suffering from heredosyphilis who have developed a syndrome identical to spinal spastic paralysis. Such cases are comparatively rare. Nonne,¹ in the last edition of his book, was able to report only about ten cases, including his own private ones. I have therefore thought it opportune to report the clinical histories of three such cases, occurring in two brothers and a sister, the offspring of a syphilitic father.

REPORT OF CASES

CASE 1.—History.—G. V., aged 54, a tradesman, at the age of 22 suffered from what was diagnosed as soft chancres of the penis. Married soon after, his first-born, a son, remained in good health. At intervals of two years a second son, a daughter and a third son were born. The fifth child, 14 years old, seemed slightly deficient, but was free from the syndrome of the three cases reported. The mother died four months ago from cardiopathic disease.

At the age of 46, the father began to complain of sexual impotence and involuntary urination. Lancinating pains in the lower extremities, a sense of oppression in the chest, incomplete emptying of the bladder, crises of dry cough with spasmodic dyspnea had only recently been complained of. He also complained of loss of memory.

Examination.—(November, 1920). Examination revealed: no motor or sensory disturbance of any of the cranial nerves; slight hypotonia of all four extremities; inequality of patellar and tendo Achillis' reflexes, both being weaker on the right side; normal upper extremity reflexes; the corneae were glistening, pupils punctiform, not reacting to light and reacting sluggishly to accommodation. There was hypalgesia along the ulnar surfaces of the forearms. There was no tenderness over the nerve trunks or grooves. Girdle hypalgesia was more marked on the right side, extending from the fourth intercostal space to the umbilicus. The Romberg sign was present. There was appreciable disturbance of locomotion. Specific senses were normal. The lymphatic glands on the right side of the neck were enlarged. The heart had a marked dilatation of the right ventricle with an aortic diastolic souffle; arteriosclerosis was present. The blood Wassermann test was positive. Lumbar puncture was refused by the patient.

1. Nonne: Syphilis und Nervensystem, Ed. 3, Berlin, 1917.

CASE 2.—History.—This patient was the second son of G. V. He had retarded intelligence from infancy, but no definite syndrome developed until he was 16 years old, when he gradually began to complain of a slight amblyopia of the right eye which progressed to almost complete amaurosis within three years. While blindness was coming on in his right eye, amblyopia of the left eye gradually supervened, progressing to blindness in about two years. A concomitant slow and progressive weakness of the lower extremities was noted, with sensations of constriction in the limbs. He was, however, able to walk without assistance.

Examination.—There were no disturbances of the cranial nerves, with the exception of the optic nerve. Active and passive movements of the neck and upper extremities were normal; speech was normal. Muscular nutrition of the lower extremities was normal; passive movements met some resistance; active movements were correctly carried out, though sluggishly. In the act of walking the feet were only slightly raised and the knees slightly flexed (paretic-spastic gait). The tendon reflexes of the lower extremities were all active; there was no Babinski sign. The pupils were equal and sluggish to light. Sensory reactions and specific senses were normal. Right Eye: There was a soft complete cataract of this eye; the fundus could not be explored; light perception was normal at 3 meters. Left Eye: There was an incomplete soft cataract of this eye, also; the fundus showed atrophy of the optic nerve (post-neuritic); several areas of chorioretinitis in the periphery.

CASE 3.—History.—A daughter of G. V., aged 22 (third born), experienced no disturbances until 16 or 17 years of age when she began to notice a sense of weakness and constriction of the lower extremities which slowly progressed until she not only dragged her feet and barely flexed her knees, but was so weak that, after taking a few steps she had to sit down, requiring assistance in order to prevent her from falling. There had never been any vesical or rectal disturbances, but she suffered from severe ozena. She had not received treatment.

Examination.—The cerebral nerves were normal; speech was, however, halting in type (bradyarthria). The upper extremities were normal. The lower extremities showed resistance to passive movements; active movements were all slow and incomplete, especially those of the feet. In walking, the feet were only slightly raised and the knees slightly flexed; she stood mainly on the toes; the radial reflexes were absent; only the right epigastric reflex was present; both the abdominal and the left epigastric reflex were absent. Patellar and tendo Achillis' reflexes were active; the plantar reflex on the right side spread out in fan shape. The Babinski sign was absent. The pupils were of medium size with a tendency to myosis; they reacted slightly to light. There was normal sensibility of the specific senses and sphincters. The blood Wassermann reaction was + + +. Mental deficiency was present; ideation was infantile; there was fairly well marked apathy even as regarded her vegetative functions. She did not evince any interest in her case. Memory was poor; perception was correct though rather limited to elementary demands.

CASE 4.—History.—A son of G. V., aged 20 (fourth born), from childhood showed marked signs of deficiency. At 16 he began to develop weakness of the lower extremities with rigidity, progressive in type.

Examination.—The cranial nerves were normal as were also the upper extremities. There was diminished strength in the lower extremities with abnormal resistance to passive movements. Abdominal, patellar and tendo

Achillis' reflexes were greatly increased. There were no rectovesical disturbances and no appreciable changes of general sensibility or of specific senses. Blood and spinal fluid tests were refused by the patient. Mentally the same defects were found that were apparent in his sister, but more pronounced; retarded perception, poor memory, apathy, stubbornness, and poor reasoning power.

Conclusions.—It is evident that the father was a tabetic patient; the history and clinical findings were typical. With the exception of the first-born, all of his children were more or less deficient; all three, at about the same age (at puberty) began to suffer from progressive motor disturbances of the lower extremities terminating in the syndrome of family spastic paralysis (spinal type). One of the brothers also suffered from optic atrophy, choroiditis and double cataract.

CASES IN THE LITERATURE

The following similar cases are reported in the literature, most of them in Nonne's book:¹

Friedmann² reports the cases of two children, 10 and 15 years old, who had spastic spinal paralysis on a probable syphilitic base. In the first case there was complete recovery after two attacks, under anti-syphilitic treatment; the second patient also had two attacks with complete recovery after the first one and only improvement after the second.

In Hoffmann's³ case the mother had a history of syphilis. The boy developed spastic paralysis at 12 years of age with arrested mental and physical development. He had mydriasis, paresis of accommodation and pupillary rigidity.

Mendel⁴ reported a case of spastic spinal paralysis on a syphilitic basis manifested at the age of 5, with progressive weakness and obtunded sensibility of the lower extremities. Slight ischiuria was also present.

Sachs⁵ reported the case of a patient with a tabetic mother with a history of two abortions and the early death of three infants. The patient developed the spastic syndrome at the age of 5 with retrogressive mentality. In this case there was also found a spastic paresis of the upper extremities with pupillary disturbances one year later.

Luzenberger⁶ found the syndrome in two brothers whose parents probably had suffered from syphilis.

2. Friedmann: Ueber rezidivierende sogenannte spastische Spinalparalyse im Kindersalter, *Deutsch. Ztschr. f. Nervenhe.* **2**: 1892.

3. Hoffmann: *Neurol. Centralbl.*, No. 13, 1894.

4. Mendel: Quoted by Nonne.

5. Sachs: *The Nervous Manifestations*, *Am. Med. Bull.*, 1896.

6. Luzenberger: Quoted by Nonne.

Vizioli⁷ reported the cases of three brothers and a sister; the father was syphilitic. All four had chronic degenerative anomalies and Hutchinson's teeth. Only one could talk clearly; the others had disturbances of articulation. All four had normal births at term and appeared normal until they began to walk, when rigidity with the typical adduction and internal rotation of the lower limbs was noticed; it was also difficult for them to sit down. Spasticity of the upper extremities was less marked and there was also a certain degree of rigidity in the muscles of the face and neck and of deglutition. The reflexes increased variably. All manifested some degree of intellectuality.

Königstein⁸ reported a case in which there was syphilis in both parents. The patient at the age of 9 showed slight mental defects, spastic paresis of all the extremities and rigid pupillary reflexes.

In one case reported by Nonne the mother had a chancre of the lip two years before her marriage. The son, when 10 months old, had a roseola and psoriasis plantaris, but otherwise developed normally until the age of 4, when disturbances of locomotion manifested themselves. Spastic gait, ankle clonus and the Babinski sign were present; pupils were normal; there was a slight grade of cranial rachitis.

Nonne also reports a case in which the father was syphilitic. The patient was sickly at birth. He began to walk at the age of 3 and from then on retrogressed mentally. Examination showed subnormal bodily development, stupid expression, left convergent strabismus, spastic paresis of the lower extremities, ankle clonus and a Babinski sign. There were rigid reflexes of the left eye, weak reflexes of the right eye with bilateral partial optic atrophy and multiple foci of choroiditis.

In a third case reported by Nonne the father was syphilitic. The child was born prematurely (7 months). It was weak and began to talk rather late. First dentition was also retarded. On account of the spastic paresis of the lower extremities, it never walked. The child looked like an imbecile. There was slight left convergent strabismus. The cranium was slightly microcephalic. The teeth were carious. Speech was normal. No sensory and sphincteric disturbances were found.

In a fourth case reported by Nonne the mother had given birth to four older children who had all died in infancy. The patient at 9 years of age had weakness and rigidity of the extremities; he was queer, bad tempered and avoided other children. The gait was

7. Vizioli: Quattro casi di diplegia spastica familiare infantile eredo-sifilitica, *Ann. di Neurol.* **12**: 1898.

8. Königstein: Ein Fall vonluetischer Spinalparalyse und reflektorischer Pupillenstarre, *Mitt. d. ges. inn. Med. u. Kinderh.* **9**:246, 1910.

pareticospastic, slight foot clonus and a bilateral Babinski sign were present; the right pupil was rigid, the left reacted only to convergence. The patient suffered from general weakness and was somewhat imbecile. Wassermann reaction: blood +, spinal fluid +, globulin positive. In the mother the Wassermann reaction was pronounced.

Artigalas⁹ reported the cases of three brothers all displaying spastic phenomena and muscular pseudohypertrophy. The syndrome gradually diminished under mercurial treatment.

Finizio¹⁰ reported the cases of two children of syphilitic parents who began to show, at the age of 3, pareticospastic phenomena. One child also exhibited bradylalia, and the other deficiency of the extrinsic eye muscles.

Déjerine¹¹ reported the case of a child who at the age of 8 began to show locomotor disturbances which slowly resulted in contractures. The thighs and legs were extended, while plantar flexion coexisted. Mental development was retarded. The pupils were rigid and unequal. The spinal fluid showed a profuse lymphocytosis.

SUMMARY

The preceding cases allow me to form a clinical grouping of the syndrome observed in each case.

The first signs of the spastic spinal syndrome due to heredosyphilis may manifest themselves just as the child begins to walk; locomotion may begin normally or be retarded; in other cases the first signs appear after the child has learned to walk properly. Thus, Nonne's first case had its onset when the patient was 4 years of age, Mendel's at 5, Déjerine's at 8, Hoffmann's at 12 and mine at puberty.

The motor disturbances—spastic paresis—may be limited to the lower extremities, as in Nonne's second and third cases, in Mendel's case and in my cases; or they may extend to the upper extremities, as in Sachs', Vizioli's, Königstein's and in the first and fourth cases of Nonne. The paresis and spasms were sometimes uniform, rarely severe enough to prevent the patient from standing up (Nonne's second and third cases). The spasms have a gradual onset and sometimes attain a severe degree of rigidity; sometimes even extension contractures of the thighs and legs and plantar flexion of the feet have been present. In one case spasm of the muscles of the neck was found. Reports on the condition of the lower extremity reflexes are

9. Artigalas: Quoted by Sandri in *La sifilide ereditaria del sistema nervoso*, Milan, 1911.

10. Finizio: Quoted by O. Sandri in *La sifilide ereditaria del sistema nervoso*, Milan, 1911.

11. Déjerine: *Paraplégie spasmodique de l'enfance*, Soc. de Neurol. de Paris, 1904.

incomplete; generally speaking, they are exaggerated (my own cases). Foot clonus may be present (Cases 1, 2 and 4 of Nonne). The Babinski sign was seldom present. In many patients the pupils did not react to light (Déjerine, Hoffmann, Sachs, Königstein, and the fourth case of Nonne). Anisocoria is less frequent; in my cases, only the second son had this symptom, but he also had a bilateral cataract and optic atrophy, the latter condition being also a feature (unilaterally) in Nonne's second case. In two cases there were left convergent strabismus and weakness of the external ocular muscles (Finizio). General sensibility is usually well preserved. In one case, rectovesical disturbances were recorded, resembling Erb's syphilitic paralysis. Disturbances of articulation (bradyarthria) were noticed in a few cases.

Psychic disturbances were common, but none seemed to attain a severe degree. My own patients all manifested varying degrees of imbecility—deficient attention, poor ideation, weak reasoning power, apathy, etc. Some patients, notably Vizioli's, had a normal mentality. The Wassermann reaction was positive in my cases and in Nonne's fourth case; it was negative in the spinal fluid of Nonne's second patient. Syphilis was assumed as the result of specific therapy (Artigalas) or after examining the spinal fluid (Déjerine). Syphilis had undoubtedly infected either the mother (Hoffmann, Sachs, first and fourth cases of Nonne) or the father (Vizioli, fourth case of Nonne and my cases) or both parents (Luzenberger, Finizio). This does not mean that in any of these cases one of the parents was immune; it is likely that an important factor in the production of this disease is the lack of antisyphilitic treatment in the parents. In Hoffmann's case, the mother had had two abortions and one child who lived only fourteen days; in Sachs' case there had been two abortions and three children who died in infancy; in Nonne's fourth case the mother had lost four infants. In my cases, the children were born shortly after the father had acquired syphilis, for which he had taken no treatment. Many authors state that there were no degenerative or heredosyphilitic stigmas in their cases; Vizioli alone reports cranial anomalies and Hutchinson's teeth. Often the spinal spastic paralysis assumes the family type (Vizioli, Luzenberger, Finizio and my own cases). It is not unreasonable to suppose that many of the cases of family spastic paralysis, described at a period when syphilis was not carefully considered, are really due to heredosyphilis. In the cases of Tooth and Tambroni,¹² in which syphilis was not even mentioned as an etiologic factor, family spastic paralysis began in infancy or in the early juvenile

12. Tambroni: Sopra un caso di tabe spastica famigliare, Riv. sper. di freniat., 1897.

period; at present there are cases of this syndrome based on heredosyphilis (Luzenberger, Vizioli, Finizio and my cases).

It is hard to say under what conditions heredosyphilis will produce spinal spastic paralysis rather than some other type of cerebral or spinal disturbance. The syndrome, or to be more exact, the various disturbances more or less constantly associated with the dominant symptom of spastic paralysis, suggest the conception of a primary morbid process of the encephalon, particularly of certain cortical areas and of their associated fibers. In some cases it is difficult to separate these types from Little's syndrome, because the pareticospastic motor disturbances only appear after the child has begun to walk (Vizioli, Nonne's second, third and fourth cases). Nevertheless, even in the cases of late development, in which the first symptoms appeared at the ages of 4 or 5, or even at 12 to 14 years of age, it does not seem to me that one should deny a close similarity of the syndrome in question and that of the congenital spastic paralysis of Little. The fundamental pathology of Little's syndrome lies in a deficient development of the pyramidal tracts which, arrested in the cervical segment or in the dorsolumbar segment, produce, respectively, a spastic tetraplegia or a paraplegia. One can extend this conception (the possibility of Little's syndrome being the same as that of family spastic paralysis) and admit that in the late cases of the disease in question the pathogenic element (syphilitic poison) affects the extreme end of the corticospinal protoneurons, pyramidal neurons, which are already developed, in order to understand that this will lead to an early decay—apparently primary—of the distal portion of these neurons. But in order that this may occur, it is necessary to fall back on a postulate, that is, that the neurons arising from the cerebral cortex, and especially those forming the corticospinal bundles, are so delicate that slight disturbances may result in degeneration. One can thus understand why, owing to the precocious onset of the disease, there are to be found areas of degeneration or absence of other anatomic structures which were developed prior to the disease, such as the optic reflex tract, nuclei of the abducens and optic nerve fibers. This would explain why in a third of all cases the disease began at the same period of life, especially at the time of puberty when the loss of balance of the endocrine glands is so easily acquired. One finds the same parallel in other organic neuroses, namely, muscular pseudohypertrophy and spinocerebellar aplasia (*morbus Friedreichi*). One might object that in some cases of spinal spastic paralysis the mentality was almost normal, but it is known that even in pure types of Little's syndrome the mental symptoms may be slight. This harmonizes with the fact that aplasia of the Rolandic cortical cells is sufficient to produce Little's syndrome; on the other

hand, pupillary disturbances, convergent strabismus and congenital psychic arrest are met with rather frequently in this disease.

It is only the cases of Artigas and of Friedmann, in which the patients were more or less permanently cured under specific treatment, that can be interpreted as the result of strictly medullary localization, caused by syphilitic and not parasyphilitic processes, as one must presume to be the case in all of the other patients.

I feel justified in making the foregoing statements because, instead of speaking (according to Nonne) of "spinal spastic paralysis with or without cerebral symptoms," I prefer to call the syndrome "spastic paralysis of spinal type." This follows the tendency of modern neuropathology of placing in the cerebrospinal class certain disturbances hitherto classed as purely spinal. This classification is, up to the present day, applied to tabes, amyotrophic lateral sclerosis and spinocerebellar aplasia (Friedreich and Pierre Marie); at present it should be applied also to the spastic-spinal syndromes on a heredosyphilitic basis.