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PARALYSIS AGITANS SYNDROME WITH SYPHILIS OF THE NERVOUS SYSTEM¹

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The chief points to which I would call the attention of the society are: First, a review of the cases reported as combinations of Parkinson's disease and tabes dorsalis with a more rational explanation of their occurrence than as merely a coincidence. Second, a report of a case in which the typical symptoms of Parkinson's disease were present but which clinical examination, as well as study of the blood and spinal fluid, showed to be a case of tabes. The rapid relief of the pseudo-Parkinson syndrome following the use of salvarsan shows that these symptoms were probably due to the syphilitic process. Subsequent study of the cerebrospinal fluid in ten cases of Parkinson's disease showed no changes similar to those found in the above case and the intravenous injections of salvarsan in six of these cases caused very slight improvement, if any.

Heinmann (according to Placzek) was the first to report, in 1888, a case of tabes dorsalis and paralysis agitans in the same patient. Placzek reported the second case, in 1892, in a male, age 52 years, who first complained of lancinating pains and three years later developed a typical paralysis agitans tremor. The

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examination of the patient showed Argyll-Robertson pupils, impotence, positive Romberg sign and lost knee jerks. In 1898, Weil reported a case of paralysis agitans in which there was absent pupillary and patellar reflexes but no luetic history and no bladder symptoms or pains. He regarded the case as most probably one of a combination of the two diseases. In 1900, Hess and also Wertheim-Salomonsen reported similar cases although the latter objected to calling it a coincidence of the two conditions and preferred to describe it as a new clinical entity, "Tromoparalysis tabioformis (cum dementia)." His case differed from the others recorded in the fact that there was a history of apoplectiform attacks and a marked dementia. In the same year Seiffer again reported the cases previously reported by Placzek and by Weil. In 1902, J. H. W. Rhein reported a case of tabes dorsalis in which there was a tremor resembling that of paralysis agitans. The other symptoms of paralysis agitans were not noted. Additional cases have been reported by Koddermann, Penzoldt and Stintzing, Bychowski, L. Bruns, Eichorst, Eshner, and Kurt Mendel (four cases). In the last two reported by Mendel the Wassermann reaction was positive.

It seems likely that some of these cases have been cases of tabes dorsalis or cerebrospinal syphilis in which there occurred, as a symptom, a tremor like that of paralysis agitans; the cases reported by Eichorst, Rhein, and Wertheim-Salomonsen may have been of that kind; excluding such cases, there still remain a number in which there is apparently a complete clinical picture of that disease. They are usually explained as due to a simple coincidence though such an explanation is not entirely satisfactory. It seems certain that there is nothing in common in the etiology of paralysis agitans and tabes for it is very unusual to obtain a history of lues in cases of paralysis agitans. The suggestion of Dupre (quoted by Mendel) that the tabes like symptoms in some cases of paralysis agitans may be due to sclerosis of the posterior spinal artery causing changes in the posterior columns does not fit the facts for, while it might explain the ataxia and loss of reflexes, it would not explain the lightning pains or the spinal fluid findings. Furthermore, such an explanation would not be possible where it was demonstrated by examination of the blood and spinal fluid that syphilitic disease was present. Cases of

general paralysis in which there has been observed a tremor similar to that of paralysis agitans have been reported by Reuter, Maillard and by Krabbe, but in none of these cases were other symptoms of paralysis agitans present. Maillard attributes the tremor in the case reported by him to a localized lesion in the cerebral peduncle though he had no autopsy findings. Krabbe's case with necropsy and histological examination of the central nervous system, showed only the lesions ordinarily found in general paralysis and no localized lesion in the mid-brain (serial sections of the region of the red nucleus).

The case observed by me was as follows:

The patient was 53 years old when she was admitted to the hospital, complaining chiefly of difficulty in walking and in the use of her right hand, in which there was a constant rhythmic tremor. There was nothing especially noteworthy in the family history except that she came of a very long lived family. She had had no serious illness except that at the age of 50 years she had had her uterus and ovaries removed on account of fibroid tumor. Her first husband had been killed in an accident. Her second husband was living and well. She had no children. About two years before her admission to the hospital she had an attack which she described as a burning, distressed feeling in the stomach which lasted several days, and while in bed with this trouble the right arm began shaking. This tremor gradually became more severe and also involved, slightly, the left hand. About six months after the onset of the tremor she began having pains in the legs which she said felt as if a knife were occasionally "jabbed" into it and at about the same time she noticed that her legs were numb and weak. These pains gradually extended to other parts of the lower extremities and she said that during the pains this flesh was very tender but as soon as the pain stopped the soreness disappeared. She had had several attacks of epigastric pain. She had noticed a paresthesia as if a fly were walking on her head and had frequently had a feeling of intense heat with an outburst of perspiration. She also described a feeling in the face as if the part had been washed in soapy water and it had been left to dry on the skin, and as if cloths wrung out of hot water were wrapped about her arms. She also said that she choked very easily but never had any regurgitation of fluids through the nose on attempting to swallow them. An examination made on admission to the hospital showed that the patient was decidedly over stout. Her face was expressionless and smoothed out. The eyeballs were prominent and there was rare winking. There was a decided hypertrichosis on the upper lip and chin and wide cheek bones

but no signs of acromegaly. There was no enlargement of the thyroid gland. The right arm and hand were held flexed and showed a rhythmical tremor of the pill-rolling type, typical of paralysis agitans. It was made worse by excitement and was absent when she was asleep. It stopped momentarily when she made a voluntary movement of the hand. The head showed a lateral oscillatory tremor at least in part transmitted from the arm. It was made worse by holding the arms. Occasionally there was a tremor in the left arm and in the right leg. Speech was somewhat monotonous but was otherwise normal and deglutition was not interfered with. The right pupil was larger than the left, neither reacted to direct or consensual light stimulation but both reacted promptly in accommodation. The extraocular movements were normal and there was no nystagmus, but at times in following the finger the eye would give several irregular movements. She said that for a moment the finger would go out of sight and that then she would see it again. The visual form and color fields were normal except for some interlacing of the red and blue. The ophthalmological report (Dr. Walter Parker) was that both eyes showed changes similar to those in retinitis circinata. There was no paralysis of the face, tongue or soft palate. There was no paralysis and no atrophy or deformity of the upper extremities. The grip was equal in the two hands. There was no ulnar deviation of the fingers. The right arm was more rigid to passive motion than the left but the biceps and triceps reflexes were prompt and equal on the two sides. Her station with the eyes open was good although she stood with her body bent slightly forward; when she closed her eyes she swayed markedly and would fall if not supported. In walking the right leg was held rather more stiffly than the left. The toes scraped the floor in the arc of a circle, her steps were short and there was some tendency to propulsion. If her eyes were closed, she staggered and could not walk at all. There was no weakness in the movements of the lower extremities. Both legs were somewhat rigid on passive motion, the right slightly more so. The legs were very stout and the flesh tender on pressure. The knee jerks were prompt and equal on the two sides; the Achilles jerks were not obtained. Plantar irritation caused flexion of the toes on both sides. The chest and abdomen were excessively stout but the umbilicus reflex was prompt on both sides.

Sensation to touch and pinpoint was normal in the face and upper extremities and localizing sense and the sense of motion and position were normal in the fingers. Sensation to pinpoint was delayed over the outer portions of both thighs and in the right leg below the knee. Light touch was not felt in the right leg below the knee except in the sole of the foot. The sense of motion and position of the toes was lost on both sides.

An examination of the urine was negative; the specific gravity, 1.025.

The examination of the blood showed: hemoglobin, 55 per cent.; red blood corpuscles, 3,600,000; white blood corpuscles, 9,950. A differential count of five hundred white blood cells showed: polynuclears, 56.6 per cent.; small lymphocytes, 18.2 per cent.; large lymphocytes, 14.2 per cent.; eosinophiles, 4.4 per cent.; and transitionals, 6.6 per cent. The Wassermann reaction was positive (XX). Blood pressure, 95 mm. Hg.

Aside from the marked adiposity, a general physical examination was negative. The heart and lungs were normal.

A lumbar puncture was done May 1, 1913. The cerebrospinal fluid was under increased pressure and 25 c.c. were removed. It was clear and colorless. There were 130 cells per c.mm. which stained slides show to be all lymphocytes. The Nonne-Apelt reaction, phase I, was positive though not marked; phase II was also not marked. The Noguchi butyric acid reaction and the carboic acid reaction of Pandey were present. The Wassermann reaction on the spinal fluid was positive (XXXX). Reducing substance was present in the spinal fluid in about the normal amount.

A pelvic examination showed only the cervix remaining, but no demonstrable pathology.

She was then given three intravenous injections of neosalvarsan. She said that she noticed a lessening of the tremor after the second injection and after the third it was much better. An examination showed practically no tremor, considerably less rigidity, and an improvement in sensation in the feet. She could walk much better.

She returned to the hospital three months later, at my request, for a reexamination. She said that she felt practically well and her weight was 200 pounds. Her facial expression was masklike and there was rare winking. There was slight rigidity in the right arm on passive movements but there was no tremor and it hung by her side. The Argyll-Robertson pupil and other signs of tabes were about as before. The urine was negative. The Wassermann reaction on the blood and also on the spinal fluid was strongly positive. The pressure of the spinal fluid was increased and it contained 110 lymphocytes per c.mm. Nonne-Apelt reaction, phase I, was positive; phase II, positive; and Pandey's carboic reaction was positive. Three months later she was again examined with practically the same findings.

So far as I am aware, a study of the spinal fluid has not been made in any considerable number of cases of paralysis agitans. In ten cases, typical in all respects, I found that it was normal.

The cell count in all cases averaged under four, the globulin and albumin were not increased and reducing substance was present. Six of these patients received intravenous injections of neosalvarsan. One patient, age 45, having the disease about two years, claimed that the tremor was lessened, that his appetite was better and he had less stiffness; objectively, there was a slight lessening of the rigidity but the characteristic tremor persisted. In two other cases there was an improvement subjectively but no objective change. One patient developed severe nausea and vomiting after the second injection and refused further treatment. In two, the treatment had no effect whatever.

In view of all the facts presented, I would conclude that a typical Parkinson syndrome may coexist with and be caused by a syphilitic lesion in the central nervous system but that this is not the etiology of the disease paralysis agitans and, furthermore, that any lesion in the central nervous system causing a Parkinson syndrome is likely an irritative rather than a destructive one.