A CASE OF PROGRESSIVE LENTICULAR DEGENERATION

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The similarity of certain symptoms occurring in paralysis agitans to symptoms found in the syndrome ascribed to disease of the corpus striatum is such as to suggest as a causative factor merely a difference in degree of intensity of involvement of that body. The pathological findings as far as known in both conditions, uncomplicated, are essentially those of an extrapyramidal disease.

Of the pathology of paralysis agitans there is at present great uncertainty. Dr. C. D. Camp (1) found in a series of nine case changes in the muscles. Redlich, Dana and Oppenheim have each reported changes in different spinal paths. Jelgersma (2) found bilateral degeneration of the striothalamic fibers. R. L. Thomson (3) from a study of the parathyroid glands in nine cases of paralysis agitans finds "no change in number, size, position or histological structure that would serve to distinguish them from the parathyroid glands found in individuals dying from other diseases." G. Gjestland (4) recently reported a case of paralysis agitans with enlargement of the parathyroid glands. Leriche (5) regards the disease as a result of a sensory excitation, ignored but incessant, keeping the muscles constantly on the strain and with this in view has performed the Foerster operation severing the fifth, sixth and eighth cervical roots on each side with unmistakable beneficial results. Gowers refers the tremor to a derangement of cortical centers concerned in movements that are naturally brought about by fear but offers no explanation of the characteristic rigidity found in paralysis agitans.

Our knowledge of the pathology of progressive lenticular degeneration is based chiefly upon findings of disease of that body after death. Mingazzini (6) from a study of thirteen cases

claims that the lenticular nucleus is a motor center disturbance of which is manifested by disassociated paresis and sometimes tremor. Mills and Spiller (7) from an analysis of eleven cases describe in lesions restricted to the nucleus absence of sensory symptoms and paresis of the limbs and face. Raggi (8) reports a case with complete destruction of the left lenticular nucleus in a right hemiplegia with a slow, monotonous, scanning speech but without aphasia which is not in accordance with the theories of P. Marie, Mingazzini and v. Monakow. Demang (9) reports the case of a man aged seventy-five with a right hemiplegia who later developed a progressive left hemiplegia. There was present a rhythmical tremor of the right hand and later of the left. At autopsy a bilateral degeneration of the lenticular nucleus was found. Dejerine denies that the lenticular nucleus has any connection with the motor and other portions of the cortex or that it plays a part in the control of movements in the functions of speech. S. A. Kinnier Wilson (10) in his brilliant paper was the first to ascribe a definite symptom complex to disease of the corpus striatum. From deductions drawn from a series of twelve cases, four of which were self-observed, he defines progressive lenticular degeneration as "a disease which occurs in young people, which is often familial but not congenital or hereditary; it is essentially and chiefly a disease of the extrapyramidal motor system and is characterized by involuntary movement usually of the nature of a tremor, dysarthria, dysphagia, muscular weakness, spasticity and contractures with progressive emaciation; with these may be associated emotionalism and certain symptoms of a mental nature. It is progressive and after a longer or shorter period fatal. Pathologically it is characterized predominately by bilateral degeneration of the lenticular nucleus and in addition cirrhosis of the liver is constantly found. The latter morbid condition rarely if ever giving rise to symptoms during the life of the patient." J. E. P. Swayer (11) reports the case of a male age thirty-six in whom the first symptoms of extrapyramidal disease appeared as a tremor of the right hand at the age of nineteen. At present he has constant tremor and muscular rigidity of varying intensity, dysarthria and a marked retardation of speech and movement; no true contractures or dysphagia, no evidence of cirrhosis of the liver and no familial history.

CASE. Patient is a female, born April 16, 1854, and admitted to the Kalamazoo State Hospital March 20, 1906.

Family History.—Her father, of whom little is known, is thought to have died of heart trouble. Her mother died at sixty-eight of unknown cause. The following letter from a son is all the history that can be obtained. "I don't remember the exact time the tremor (St. Vitus Dance) began but think about two days before she died. She never was paralyzed nor had any stiffness of muscles to my knowledge nor apoplexy, had some headache I don't think she vomited when she had them. She had bilious spells and at times she would be very sick and vomit. I never knew that she had any nervous trouble unless it was at the last with the St. Vitus Dance. My mother's stomach and liver troubled her for a long time. Before she died the doctors told her that she had hardening of the liver. She was also bothered with diarrhea. At her last sickness they told us she had a complication of diseases." The maternal grandmother was insane and the patient had one sister and one son temporarily deranged.

Previous History.—Her early development was uneventful. Menses were established at the usual age, were always irregular and ceased at forty-eight years of age. She had three children but was never very strong and could not care for her children as well as most mothers. She had in her early life severe headaches of the nature of a migraine; previous to the onset of the headaches she would see "black spots" and the headache usually ceased after vomiting. She always complained of "liver trouble, of the yellow color of her face and of liver spots."

History of Present Illness.—In the year 1898 the patient noticed that the big toe of her left foot was drawing up and interfered with the wearing of her shoe. During the year 1901 a tremor appeared in the left hand. At this time her arm felt stiff and she had some difficulty because of this stiffness in combing her hair. It was thought at that time that in walking she dragged the left foot somewhat and brought it a trifle more quickly to the floor. In the year 1904 she had a swelling of the lower limbs, the left being the more severe of the two. The left foot had already begun to turn in. The limbs were very tender to pressure and she complained continually of cramps in her limbs. Early in the year 1905 a tremor appeared in the right arm, later the right leg became stiff. The limbs and left arm were already somewhat contracted but she was able to walk with assistance until the year 1906. At this time the patient was so irritable and exacting that life was intolerable for those about her. The rigidity of the face and a speech defect were observed in 1906. From the onset of her illness the patient complained of abdominal pain and always held the abdominal muscles perfectly rigid. With but slight increase in the speech defect, the degree of contracture of the limbs and the bodily rigidity there has been no

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change in her condition since 1907. As early as 1906 retardation in originating speech and movement were noticed. In February, 1909, the patient had an attack of nausea and vomiting and at this time vomited considerable "bile." She has always been constipated and complains of intestinal cramps. She has had no headaches since the year 1909 and there is no history of alcohol, drugs or venereal infection.

Present Examination.—The patient lies either in bed or in a wheel chair in a fixed rigid attitude and can only change her position with assistance. When moved, she is carried "en bloc." The facies is smooth, stolid and mask like. The lines about the mouth are not completely obliterated and voluntary movement of the lips is possible but very slow and rigid. She occasionally breaks into a broad stiff smile which recedes very slowly. There is no difficulty in moving the eyelids. She is able to partially open the mouth but unable to move the jaw from side to side. The eyes are keen and the expression alert, contrasting markedly with the facial expression. The pupils are round, central, equal, regular, react to light direct and consensual and in accommodation. Extra-ocular movements are normal. There is no nystagmus, no contracture of the visual field. Fundus examination shows a marked central choroiditis. Her hearing is normal. Swallowing is very difficult, more so with liquid than with solid food; after much delay the patient swallowed on request. In drinking the water is held in the mouth until it can be swallowed and frequently saliva and liquid foods may be seen running out at the corners of her mouth. Contraction of the masseters is very slight. When asked to protrude the tongue she after much effort moved it to the edge of the lips and was unable to move it further. There is a pronounced to and fro tremor but no atrophy. She is able to slowly move her head from side to side but is unable to bend the neck forward or backward. The general muscular nutrition is fair but there is generalized atrophy. On palpation the muscles feel firm. There is marked hypertonicity of the muscles of the neck, trunk and limbs; most pronounced in the lower limbs. On passive, alternating flexion and extension of the forearm and arm one meets with great resistance, rapid movements being impossible. Both arms are held in adduction at the shoulders. The forearms are flexed, the left more so than the right. The right wrist is slightly flexed. The thumb of the right hand is bent under to the palm and the fingers are slightly hyper-extended. Voluntary flexion is impossible and passive flexion very slight. The left wrist is more flexed than the right. The fingers of the left hand are flexed tightly onto the palmar surface and the thumb fixed in extension. Voluntary or passive movements are impossible. The elbow contractures can be somewhat overcome by passive movements. There is contracture of the hips which cannot be reduced. The knees are contracted at

right angles but this contracture can be partially overcome. The left foot shows complete equinovarus with dorsal flexion of the first phalanx of the big toe and plantar flexion of the terminal phalanx. The left ankle is fixed. The left foot inclines to the position of equinovarus. The right ankle is partially movable. The abdominal muscles are rigid. She exhibits a constant tremor of the entire body, which is aggravated by any volitional effort and ceases during sleep. It is a true tremor of varying intensity and excursion and in the arm the most pronounced movement is from the elbow and wrist. There is at times a marked rotatory interosseous tremor of the third and fourth fingers of the right hand. When the mouth is partly open there is a rythmical up and down movement of the lower jaw. There is a constant up and down tremor of the lower limbs.

The speech is slow and there is slurring of the words. Her voice comes from deep in the throat and there is only very faint movement of the lips.

Corneal and pharyngeal reflexes are present, the umbilical reflex is not obtained. The biceps, triceps and wrist jerks are present, prompt and equal. The knee jerks and Achilles jerks are not obtained because of the contracture. There is no ankle clonus, Babinski or Oppenheim.

There are no sensory symptoms except the cramps of which she frequently complains.

During the greater part of her illness the patient has been exceedingly irritable and fault-finding but of late years has become more cheerful and frequently smiles spontaneously. She has never had any hallucinations or delusions but there is some mental deterioration and her memory is not reliable. She is fully aware of requests and responds in so far as her physical condition will permit.

The Wassermann tests, blood examination and urine analysis are negative.

The examination of the thorax is negative; of the abdomen shows pronounced dilatation of the veins about the umbilicus and the liver to be enlarged toward the umbilicus and the right iliac fossa a hand's breadth below the rib margin.

This case is of a special interest in as much as occurring as it does at the possible age of onset of paralysis agitans, it presents the comparatively rapid progress, continuous tremor, constant rigidity, true contractures, dysphagia, dysarthria, possible familial hepatic disturbance and evidence of disease of the liver found in progressive lenticular degeneration.

In all the cases reported with the exception of that of Sawyer's, cirrhosis of the liver has been observed. Kinnier

Wilson suggests that it is a primary feature of the disease and that the toxin producing the degeneration in the brain may be generated in connection with the hepatic cirrhosis. Sawyer considers the possibility of an enterogenetic toxin producing primarily the lenticular degeneration and subsequently the hepatic cirrhosis. From the history of the above case and that of her mother the hepatic disease evidently preceded the nuclear degeneration, but whether the toxin is primarily of hepatic origin or primarily of other origin the fact is evident that it has a selective action on the lenticular nucleus.

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