PROGRESSIVE CEREBRAL HEMIPLEGIA: ITS PATHOGENESIS AND DIFFERENTIAL DIAGNOSIS*

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Under the term progressive cerebral hemiplegia is understood a condition in which a unilateral paralysis of cerebral type is established slowly and progressively but in sections, commencing either in the upper or in the lower extremity or else in the face. The pathological material in such cases has been found to present a great variety. Edema of a whole hemisphere, large areas of softening, obliteration (partial or complete) of the internal carotids, thrombosis of the Sylvian arteries, multiple foci of softening through arteritis of small bloodvessels in the cortex or centrally located; finally tumor of the brain—these are the anatomical conditions which have been observed by various authors in the form of hemiplegia under consideration. Before discussing further the pathogenesis of the condition the following cases are presented for consideration:

CASE I.—Man aged fifty-five, with a history of a syphilitic infection at the age of thirty-five, commenced to suffer from attacks of vertigo several months previously. His heart was somewhat enlarged, the aorta presented a marked accentuation of the second sound; the blood pressure was 170 systolic. Urinalysis showed a faint trace of albumin. One morning after a more violent attack of vertigo than usual, he felt the left arm and hand become numb, he rubbed the limb with vigor to make it react; on the following day the arm became paretic. Gradually the paralysis became more and more pronounced. On the tenth day the left lower limb became similarly involved and finally the left side of the face became deviated to the right. The speech was at no time affected except toward the end, when the patient's mentality became greatly weakened. In addition to the hemiplegia there was also a total hemianalgesia in the left arm and leg but only a hypalgesia in the lower half of the face on the same side. The reflexes were all typical of a cerebral attack. The eyes were normal, with the exception of a slight limitation of their movement toward the left. The tongue was deviated to the left side. There was no difficulty of deglutition. The sphincters were normal. The patient's condition gradually grew worse, the paralytic state became deeper, the sphincters ceased to functionate, the patient became mentally dull and gradually a comatose state made its appearance. Death occurred on the twenty-eighth day.

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At autopsy considerable edema was present over the Rolandic area in the right hemisphere. Both ascending convolutions were pale and on palpation a softness of the tissue was felt. The soft consistency was particularly marked in the middle portion of the precentral convolution, but less marked in its upper third; the lower third presented the same consistency as the surrounding tissue. Transverse cuts revealed that the entire cortical layer was correspondingly softened and contrasted sharply with the normal tissue but the softening did not penetrate deeper and left the white substance untouched, except over a very thin layer immediately close to the gray matter. The basal ganglia remained intact. The most interesting finding is the condition of the right middle cerebral artery; at its origin where it leaves the internal carotid, it was found to be hard on palpation; a section of it revealed an almost complete obliteration of the lumen. Dilatation of the basilar artery was another interesting feature. A microscopical study of these arteries showed distinct evidence of endarteritis, which was probably of luetic origin, but on the other hand the small arteries distributed over the Rolandic area did not show any material changes. A histological study of the nerve tracts showed distinct degenerative changes with the Marchi method but very slight with Weigert's stain. Special emphasis is laid on the fact that the most pronounced softening of cortical tissue was in that portion of the Rolandic area which corresponds to the center of the arm, and it is to be recalled that the paralytic condition commenced in the arm.

CASE II.—Man aged sixty-three, an inveterate alcoholic, had had attacks of vertigo with severe headache for a number of years. After one of these attacks in which the vertigo was unusually severe he became unconscious. Fifteen minutes later he commenced the regain consciousness and it was noticed that the left leg was paretic. The reflexes showed an involvement of the motor pathway. Sensations to all forms were greatly diminished. The paralysis of the leg became more and more pronounced. Ten days later there was observed a gradual diminution of power in the left arm with an accompanying flaccidity. Shortly afterwards a slight deviation of the face to the right was noticed. The patient was under observation until he expired, thirty days later. During that period he presented a typical left motor hemiplegia with a diminution of sensibility to all forms on the same side. Gradually the sphincters became involvement. The mentality remained clear until the end. The patient's blood pressure oscillated from 200 to 160 systolic. The left cardiac ventricle was dilated.

At autopsy the findings were as follows: Both hemispheres were edematous with predominance over the right sensorimotor area. At this level a pronounced vascularization could be seen. The lower portion of this area was somewhat softer than the neighboring tissue. Inspection and palpation revealed a more or less solid consistency of the arteries over the convexity and at the base of the brain. Only in one place a nodular condition was detected, it was at the level of

origin of the right Sylvian artery. A section of the latter showed an almost complete obliteration of the lumen. Further examination showed that the ascending frontal convolution was more softened than the ascending parietal, a fact which probably has some bearing upon the difference of the degree of involvement of objective sensibility and motor disorder during the patient's life. The softening was only peripheral but it gained also in depth. A gross section showed an edematous state in the right hemisphere between the periphery and the basal ganglia. However a certain portion of the internal capsule and of the adjacent lenticula was found softened, but the knee and the upper part of the capsule, as well as the thalamus were intact. Histological examination showed marked atheromatous changes in many of the cerebral arteries, which were particularly evident in the internal layer of the vessel wall. It was unusually pronounced in the Sylvian artery of the level of the above mentioned obliteration. The cortical cells showed marked deformities with a certain degree of chromatolysis. As to the nerve fibers some degenerative changes were traced only with the Marchi methods.

CASE III.—Man aged sixty-five, during the last ten years of his life severe attacks of mild subjective sensory disturbances in his left hand and foot. They consisted of tingling with numbness followed by weakness in the left arm and leg. The first attack lasted a few hours, but each subsequent attack was longer in duration so that the duration of the last one was a whole week. While he always recovered from each seizure, the fifth which was the last left his left arm somewhat weak. Nevertheless this weakness did not interfere with his regular occupation which was clerical. About fifteen days later his left arm became totally paralyzed and this occurred after an attack of vertigo without loss of consciousness. Five days later the left leg became paralyzed and at the same time a deviation of the face toward the right was noticed. The paralysis became gradually deeper and deeper and presented the typical symptoms of cerebral motor hemiplegia. Sensations were but slightly involved. There were no eye manifestations and the sphincters were intact. The patient presented a high blood pressure 220 systolic. The heart was dilated and the second aortic sound was loud and musical. Arteriosclerosis was pronounced. On the twentieth day the patient's condition became suddenly worse, a comatose state set in and he expired.

Autopsy revealed a marked edema in the right hemisphere. The pia was thick and infiltrated so that when after a puncture the edematous fluid escaped, it collapsed on the surface of the brain. There was distinct softening on the Rolandic area and particularly in its median portion, more in the precentral than in the postcentral convolution. The softening encroached somewhat on the prefrontal lobe in the middle portion. The arteries of the convexity of the entire cerebrum as well as of the base appeared thick and on palpation they felt hard. A specially hard area was found in the Sylvian

artery at the level of its origin. A cut of that area showed an almost complete obliteration of its lumen. A number of cuts were made over other arteries as well as those of the base. Thickened walls were in evidence in the majority but no obstruction of the lumen was seen. Transverse cuts of the brain revealed that the softening penetrated through the entire thickness of the cortex. An area of softening was found in the right internal capsule back of the knee. The thalamus opticus was intact but a small area of the lenticula adjacent to the capsule was also softened. Microscopical examination showed besides cellular changes in the cortex also degenerative changes in the motor pathway more with the Marchi than with Weigert's method of staining.

CASE IV.—Man aged thirty-eight, complained of severe headache, vertigo and had frequent vomiting spells. The tendon reflexes were increased in the lower extremities, but no other abnormal reflex was present. The eyegrounds were negative for a long time. The condition remained unaltered during a period of four months. At that time the patient observed a gradually oncoming weakness in the right arm with a mild difficulty of speech. Examination revealed also a sensory disorder in the affected arm, consisting of a pronounced hypesthesia to all forms of sensations; the patient complained of a numbness in the fingers and of inability to appreciate objects; astereognosis was present. The paretic condition grew deeper and deeper when on the sixth day the right leg became paretic. Gradually a complete paralysis of the leg developed. The face was but slightly involved. The speech became more and more typical of motor aphasia. Soon the patient's eyes commenced to show symptoms. Diplopia occurred frequently. The eyegrounds showed a haziness of the media, optic neuritis rapidly developed on both sides. The headache became more severe, vomiting more frequent. Convulsive seizures appeared on the right side. A comatose state gradually set in and the patient expired in one of the convulsive attacks. At autopsy a gliomatous neoplasm was found in the left hemisphere involving the ascending parietal convolution and partly the ascending frontal close to the Rolandic fissure affecting their median portion.

CASE V.—Man, sixty-four years of age, after a prolonged period of vertigo there developed five months ago a sudden paresis, with a sensation of numbness in the right leg. At the end of seven days he regained considerable power. Nevertheless a certain awkwardness of gait remained. Two months later a similar condition appeared in the right arm and leg. The face was also seen to deviate towards the left. The paresis became gradually more and more pronounced so that at the end of the fifth month the patient presented a complete right hemiplegia. Presently there was not only paralysis on the right side but also motor aphasia. The sensations were diminished on the right side. All the reflexes were typical of cases with a cerebral damage. The patient gradually grew weaker. Diarrhea made its appearance. Coma and death followed. At

autopsy was found a softening of the entire posterior limb and posterior portion of the anterior limb also of the knee of the left internal capsule. The large arteries of the convexity and base of the brain were found hard on palpation.

The clinical aspect in the five cases presents common characteristics, viz., the paralysis developed slowly and gradually at first as a mere weakness in one limb, but this imperceptibly grows more and more profound until a complete and total hemiplegia is fully established. The paralysis may commence in one or another extremity. In three cases the onset was in the arm, in two in the leg. The face as a rule becomes involved simultaneously with the arm. Sensations were invariably involved. In the first case there was a total analgesia on the affected side, in the second case—great diminution, in the third and fifth hypesthesia to all forms of sensations and in Case IV almost complete abolition of the superficial sensations and astereognosis.

Special emphasis deserves the onset of the condition: In all the cases except Case II, there was no suddenness which is so frequently the case in apoplexy; a mild paresthesia with a weakness in one limb is most characteristic of the invasion of progressive hemiplegia. The prodromal period in the five cases presented a state of vertigo and the onset of the unilateral paresis was invariably preceded by an unusually severe vertiginous state.

The anatomical findings require special consideration. In the first three cases we see a marked edematous state of the cortex with deep softening. Total obstruction of the main artery supplying the cortex was in evidence. The area of softening and the apparently anemic appearance of it is in sharp contrast with the neighboring tissue which remains firm. Edema and softening penetrate the brain through the entire thickness of the cortex and involve partly the white substance. The edema uniformly found in each of the three cases over a limited region naturally suggested a vascular obstruction. Indeed in each of these cases obliteration was found in the Sylvian artery although at a different level, but near its origin. The obstruction was complete. In Case IV we deal with a gliomatous tumor which for a long time (many months) did not show marked symptoms of increased intracranial pressure, as evidenced by the absence of focal symptoms and eye manifestations. No edema of the surrounding tissue was observed. In Case V there was a softened internal capsule and again no edema. Except Case IV with tumor, in all the other four cases there was distinct and pronounced degenerative changes in the blood vessels.

In view of these anatomical data let us consider the pathogenesis of progressive cerebral hemiplegia.

In Cases I, II, III, and V we find during the patient's life evidences of endarteritis, viz., frequent attacks of vertigo with peripheral arteriosclerosis. Arteritis, symptomatic of localized edema, is a well known fact in pathology. The first consequence of interference with arterial circulation in complete or even partial closure of the lumen of an artery is a more or less pronounced stasis in the neighboring veins and a passive dilatation of the latter. The passive dilatation is facilitated by a vasomotor paralysis which is usually present in such cases. In the cases under discussion by virtue of arterial alteration partial obliteration was being produced very slowly and over a long period. The edematous state of the nervous tissue was consequently equally slow and according to the intensity and the seat of the edema corresponding symptoms made their appearance. When an obliteration becomes complete in an artery which irrigates a given region, edema is pronounced and total suppression of function in the affected area will be the result. In the case of tumor the mechanism of progressive hemiplegia finds its explanation first in the consistency of the mass and secondly in the gradual encroachment of the latter on the various centers of the Rolandic area. The softness of the neoplasm prevented a stormy onset with its usual abrupt or sudden development of paralysis, with or without loss of consciousness; it produced slight but progressive pressure on the important sensorimotor area with the result of a gradual and mild interference of function dependent on the involved portion of the brain. A paretic condition was the first and a complete extensive paralysis was the final step.

The five cases described here permit to draw this conclusion that progressive cerebral hemiplegia may be encountered in endoarteritis from arteriosclerosis, in obliteration of a large artery such as the middle cerebral and in tumors especially of gliomatous type. But in Case I, in addition to the arteriosclerosis incidental of the patient's age there was also a history of syphilis. The sclerosis of the blood-vessels may be considered in a causal relationship with lues. That syphilitic infection at any of its periods may produce paretic conditions in one limb or partial paralysis is a well known fact. It may also cause a paralysis in a portion of the body on one side and later on produce identical conditions in the remaining portions of the same side. I have in my possession facts of this order of a clinical order. I observed in the tertiary period of syphilis, as well as in

tabes, progressive cerebral hemiplegia precisely with the same characteristics as those seen in the cases described above. In fact in all instances of partial paralysis lues should be suspected and if the laboratory tests are all confirmatory, they should be managed accordingly. The recognition of this possibility is preeminently important as cerebral ischemia may be prevented if the preobliterating phase of arteritis can be handled successfully. Syphilis of the nervous system as a rule presents no difficulty of diagnosis. The accompanying changes in the reflexes, in the reactions, dimensions and form of the pupils, in the function of the ocular muscles, in the function of the sphincters—are all adequate enough in presuming a luetic state. Particular reliance however should be placed on the Wassermann reaction of the blood and spinal fluid, on the cytological state of the latter, namely lymphocytosis.

Partial and incomplete hemiplegia occurring abruptly without loss of consciousness, mostly bilaterally and commencing frequently in the upper extremities, soon showing improvement (but not recovery) and remaining flaccid, if ever reaching the state of contracture, is observed in aged individuals and it is due to a successive formation of lacunar foci in the cerebrum. This condition was first described by Marie, Ferrand, and Léri. The successive appearance of the symptoms may give the impression of a progressive hemiplegia as described above, but the occurrence in old age together with other evidences of pathological senility, such as speech disturbance and psychic changes, namely flaccidity, also the tendency to amelioration, the cilaterality of the motor disorder,—all these features will aid in building up the correct pathogenesis of the condition.

Progressive hemiplegia may occur in cases of aneurisms of the basilar artery. Such a case is reported by de Massary and Carton (*Bulletin de la Société Anatomique*, Juillet, 1901). The patient, aged forty-three, had at first a slight hemiparesis on the left side, face including. There was some dysarthria. The latter rapidly improved. Soon the paralytic condition commenced to increase and gradually became complete and profound. The speech disorder reappeared. A comatose state set in and the patient expired. At autopsy an aneurysm was found on the right side of the basilar trunk which was compressing the upper and internal third of the right side of the pons. A section through the latter showed that the aneurismal sac produced not only pressure but also and more so a disorder through the disturbed vascular supply causing a focus of softening in the middle cerebellar peduncle and in the right pyramid. The ad vitam

diagnosis was very difficult in view of absence of cranial nerve involvement: The oculomotor, facial and trigeminal nerves were all intact.

Progressive hemiplegia has been observed in cases of uremia and at autopsy an edematous state of the cortex with a localized predominance over the motor area was found. Whether the edema is due to the accompanying endarteritis frequently found in such cases or to the toxic action of the pathological urine, or else to both elements, one cannot be certain. At all events one must bear in mind the possibility of occurrence of a progressive paralysis in uremic states. However, the distinguishing features will be found, in brief, alternating phases of aggravation and retrogression of the paralytic phenomena.

A similar tendency to retrogression in paralysis is observed in diabetes in the course of which progressive hemiplegia is sometimes observed. It is very probably due to a vascular lesion produced by intoxication of diabetes. Analogous condition and pathogenesis have been observed in intoxications from other sources, such as saturnism, carbon monoxide, alcoholism.

Tuberculous meningitis is another affection in which a slow and progressive paralysis of the limbs on one side of the body is sometimes observed. Chantemesse was the first to call attention to it (*Thèse de Paris*, 1884). Here again a vascular disorder caused by the tuberculous process might be incriminated. It must be pointed out that here like in uremia the intensity of paralysis is liable to alternate in the course of the disease, but invariably it terminates by a complete and permanent hemiplegia.

In 1846 (*Medical Times*, London) Fletcher described a symptom-group under the name of ingravescent apoplexy which was subsequently studied among many other observers by Broadbent (*Medico-Chirurg. Transactions*, V. L, IX, 1876), Abercrombie, Thomas Watson, Mossé (*Gaz. hebdom. de Montpellier*, 1889) and Peuch (*Progrès Médical*, 1889). Pathologically the affection is characterized by a hemorrhagic focus situated between the striate body and the external capsule. The clinical picture is as follows: In the beginning there is usually a sudden and severe headache; the patient is pale, the body becomes cold, the pulse is feeble and the individual falls in syncope. Sometimes the initial symptom is mental confusion. The syncopal state is usually brief. After having regained consciousness the patient continues complaining of headache. After a more or less brief period the patient becomes incoherent and suddenly

coma sets in from which he never recovers. Paralytic symptoms may be or may not be present. If it does appear it usually develops slowly and by gradation so that eventually complete hemiplegia will ensue. In the latter case the resemblance to the clinical picture of progressive hemiplegia described above is striking. A careful analysis however will show fundamental differences between the two affections. In ingravescent apoplexy the paralytic symptoms may sometimes be entirely wanting. In cases in which they are present, there are always stormy manifestations characteristic of an apoplectic insult, viz., sudden loss of consciousness. Coma appears soon after consciousness has been regained and increases progressively. In progressive hemiplegia on the contrary, a comatose state appears as a terminal phase a long time after the hemiplegia has been established. Besides, the state of malaise with headache and vomiting described above as immediately preceding the syncopal state of ingravescent apoplexy is entirely absent in the syndrome of progressive hemiplegia. The difference between the clinical pictures of one and the other cerebral condition finds its corroboration in the anatomical findings: while in ingravescent apoplexy there is always a hemorrhage, in progressive hemiplegia there is an area of softening which may be caused by a multiplicity of lesions, such as we have seen in the beginning of this work, viz., endarteritis, obliteration of middle cerebral arteries, pressure from a tumor, aneurism of the basal arteries, a degenerative state of the arteries of syphilitic nature.

In 1899 Thomas and Long (*Comptes rendus hebdomadaire des séances de Société de Biologie*, No. 28, 1899, p. 768) report an anatomoclinical case resembling to some degree progressive hemiplegia under discussion. It was a case of a man aged forty-seven, with a history of syphilis at thirty-six. Seven years before his death there gradually developed a paresis of the right leg which eventually became permanently paralyzed. There was also a complete anesthesia in the leg. About a year later a paresis of the right arm made its appearance. Gradually spincter disturbances made their appearance and a year later the patient died, after an acute attack of pleurisy. At autopsy several plaques of sclerosis were found in the spinal cord. The condition in the opinion of the author reminds one of some of the lesions in spinal syphilis. Another similar case but without autopsy findings was described by Mills (*Journal of Nervous and Mental Disease*, 1900, 27, p. 95). In a man of fifty-two a gradual weakness and awkwardness developed in the right lower extremity. Eighteen months later an identical condition appeared

in the right arm. The face was also involved in its right half. The paretic condition of the two limbs did not increase for a long time and there was no spasticity. The condition of the reflexes pointed to an involvement of the motor tract. The sensations were normal but atrophy of the musculature was present on the right side. The author expresses the opinion of the case being probably an example of a slowly increasing degeneration of the cerebral motor neuron system.

The two cases just reported present a progressive development of a unilateral paralysis, but the mode of onset and the course of the disease—the character of the paralytic symptoms, the termination and the absence of arterial manifestations, finally the autopsy findings in one of them,—all prove a fundamental difference with the syndrome of progressive cerebral hemiplegia described in the present contribution.