

CHRONIC ENCEPHALITIS

A 20 Year Clinical Study — Case Report

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In major epidemics of encephalitis lethargica it is estimated that about 25 per cent of the patients recover completely without sequelae.¹ Of the remainder some die, and the others develop neurological disorders of one sort or another. Not all of these patients show progressive degenerative changes in the striatal and parastriatal nuclei of the mid-brain to the extent that severe parkinsonism develops. Furthermore, when one considers the diffuse damage to the gray matter of the brain, it is not surprising that certain patients develop bizarre disturbances of the sensory and motor functions of the body.

If the symptoms of the acute onset are characteristic of the lethargic or epidemic type of encephalitis, the neurological changes may be attributed to this disease with reasonable certainty. On the other hand, even though great care is used in the clinical study of any particular case, a long period of observation may be required before a proper diagnosis can be made. The following case is of a patient whose symptoms and physical signs were puzzling and who at first seemed to be headed for postencephalitic parkinsonism.

CASE REPORT

This case of chronic encephalitis is of considerable interest not only because it has been followed clinically for 20 years during which time the patient has married, raised three children, and has earned his living; but also because he has not showed the usual progressive changes in the basal ganglia and hypothalamic areas of the brain. Another important feature of this case has been the recent occurrence of spells of unconsciousness which have resembled the grand mal seizures of idiopathic epilepsy. Furthermore, the clinical differentiation between encephalitis, encephalomyelitis, and disseminated sclerosis has presented an interesting and difficult diagnostic problem.

A young man, 22 years of age, was admitted to the Clinic in September, 1922. His chief complaints were marked generalized fatigue, dizziness, fullness in the head, difficulty with speech, and moderate ataxia. In February, 1920 he had had an attack of "influenza" characterized by moderate fever, malaise, and muscular aching. He had recovered from the acute illness within a few days, but had complained of continued fatigue until July, 1920. At that time symptoms of encephalitis had developed with diplopia, somnolence, and dizziness. His speech became disturbed by difficulty in use of the tongue, although swallowing was not affected. He improved in the next two months, but he continued to experience moderate ataxia, dysarthria, and excessive salivation.

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At the initial examination at the Clinic in September, 1922 the dysarthria was still present as well as dizziness, moderate ataxia, generalized fatigue, and marked constipation.

Neurological Study: An examination of the eyes revealed that the optic discs were normal. The pupils were equal and regular and reacted to light and convergence. There was no ocular palsy. Nystagmus was present on looking to the right.

The patient had a moderate mask-like expression with infrequent winking. Weakness was especially marked on the left side of the face when in repose, and there was hypesthesia of the left face.

The reflexes of the upper extremities were normal. In the lower extremities there were hyperactive patellar reflexes and a positive Babinski on the right.

The abdominal reflexes were present but sluggish; the cremasteric reflexes were also present.

The muscles of the right arm and leg were slightly spastic. There was no atrophy nor tremor. The finger to nose and heel to knee test was awkward on the right. The patient's gait was slightly ataxic from spasticity of the right leg.

Laboratory Studies: The blood Wassermann and Kahn reactions were negative. Blood counts revealed no macrocytosis. An Ewald test meal revealed a free acid of 42 and a total acid of 56 in one hour. The patient refused spinal puncture.

The clinical diagnosis was a diffuse lesion of the cortex and basal ganglia from acute encephalitis and probably early parkinsonism.

Treatment: With the use of Fowler's solution and potassium iodide the patient gained 21 pounds in two months and was able to return to work.

Progress Notes: In June, 1923, nine months later, the patient complained of numbness of the right side of the body and of marked numbness and hypesthesia of the palm of the right hand. His general condition was good.

In May, 1924 examination revealed numbness in the toes of both feet with diminution of sensation to touch, pin prick, and vibration.

In April, 1929 examination revealed numbness of the left side of face and the left half of hard palate and tongue, with loss of the sense of taste over this area. A speech difficulty was again present. There were hyperactive reflexes in the legs with bilateral ankle clonus and a right Babinski. Abnormal reflexes were absent, and there was no ataxia.

At this time the patient submitted to a spinal puncture. The subarachnoid fluid was clear and colorless, and the pressure was normal. Spinal fluid analysis revealed 2 cells; the globulin and Wassermann reactions were negative; colloidal gold, 3-2-2-1-1-1-0-0-0-0.

In October, 1937 the patient had numbness in the right leg, ataxia in the right leg on walking, and dysarthria.

In July, 1940 examination revealed a slight slurring of speech, a moderate intention tremor of right hand, weakness, and a sense of fatigue.

In 1938 the patient had a convulsion which was nocturnal and associated with urinary incontinence, but the tongue was not bitten.

In June, 1942 the second convulsion occurred. The patient felt himself pulled to the left and fell unconscious. When he recovered his senses after two hours, he was standing at his desk dressed in his overcoat and hat. No head injuries were noted, nor was the tongue bitten.

On February 5, 1942 at 9:30 p. m. while at home, he became unconscious and fell off of his chair. Convulsive movements involved the left arm and leg and lasted from five to six minutes. The inside of the right cheek was bitten. Upon recovery of consciousness within a few minutes, he experienced no pain nor headache, but felt dazed for an hour.

A neurological examination in February, 1942 revealed the following: There was no nystagmus nor ocular palsy. The pupils were equal and reacted normally; the optic discs were normal. There was slight weakness of the left side of face. The other cranial nerves were normal, and there was no pallor. Sensation to vibration was poor in the feet and absent in the toes. Reactions to pain, heat, and cold were normal.

Reflexes: The patellar and Achilles reflexes were hyperactive bilaterally. The Babinski reflex was present, the sustained ankle reflex was clonic on the right. The abdominal reflexes were active in the upper areas and sluggish in the lower. Cremasteric reflexes were absent. Ataxia was noted in the left finger to nose test. There was slight muscular hypertonus of the right leg. The Romberg was to the right and backwards. The patient's gait was disturbed as a result of spasticity of the right leg.

Upon spinal puncture the fluid was found to be clear and colorless; there were normal dynamics of fluid, 7 cells, a faint trace globulin, 45 mg. total protein; the Wassermann and Romberg colloidal gold reactions were negative. Electro-encephalography showed abnormal waves in the frontal area of the brain.

Discussion: Although the acute stage of encephalitis lethargica may show the characteristic signs of fever, somnolence, diplopia, and ocular palsies, postencephalitic sequelae often appear in the absence of such early disturbances. Likewise, postencephalitic or chronic encephalitic symptoms may be manifested by disabilities other than chronic parkinsonism or oculogyric crises. Changes in personality, emotional instability, and various sensory or vasomotor symptoms indicate that the lesions are diffuse, widespread, and usually progressive. Although the "virus" shows a specific affinity for the gray matter of the cortex and basal ganglia, the damage is by no means confined to these areas. Toomey² has emphasized the existence of many varieties of encephalitis and has drawn attention to their atypical manifestations.

Although the case reported illustrates certain rather bizarre and confusing nervous symptoms, the history clearly indicates an acute onset of encephalitis followed by more or less permanent disability. Even though the nervous system changes have been slowly progressive, the occurrence of remissions and exacerbations suggested that actually the patient might be suffering from chronic encephalomyelitis or from disseminated lesions of multiple sclerosis. However, although the spinal fluid showed a mildly positive colloidal gold reaction to the first test in April, 1929, the fluid obtained in February, 1942 was normal. Positive colloidal gold reactions have been reported in some cases of encephalitis, but are more common in disseminated sclerosis and cerebrospinal syphilis. Also, the abdominal reflexes have not been consistently absent in this patient, although they usually disappear in multiple sclerosis. Furthermore, it is unlikely that demyelination of the white matter would persist and progress for 20 years without showing typical lesions of disseminated sclerosis.

The appearance of convulsions 18 years after the acute onset of encephalitis is an unusual feature of this case. The electro-encephalography showed abnormal waves in the frontal area of the brain. Such irritative foci indicate that brain damage has occurred here. From a

clinical standpoint the convulsions were Jacksonian in character and seemed to originate in the right motor area. As yet an encephalogram has not been made, but may be indicated if the attacks are not controlled by dilantin and phenobarbital.

CONCLUSION

The progress of this patient during the past 20 years has manifested signs and symptoms of a rather diffuse and progressive lesion of the central nervous system, yet he has not shown any clinical manifestations of degenerative changes in the basal ganglia. The neurological changes here have been difficult to place in any definite classification. If a history of acute encephalitis had not been obtained from this patient, we would have regarded him as having atypical multiple sclerosis.

It is of further interest to record that he has worked quite steadily during this long period with little loss of time, and it appears that he will lose only five or six weeks of working time in this present episode. With reference to treatment we doubt that the administration of arsenic and iodides had any definite influence upon the course of the disease.

However, in the past month he has improved markedly with the use of dilantin not only in the control of his convulsions but also in a general feeling of well-being.

Regardless of what the future has in store for this patient, it is fortunate that his defense mechanisms were adequate to keep this disease in abeyance for so many years.

REFERENCES

1. Brain, W. R.: Diseases of the Nervous System. 2nd ed. (London: Oxford University Press) 1940.
2. Toomey, J. A.: Differential diagnosis of various forms of encephalitis. J.A.M.A. 115:1985-1989. December 7, 1940.