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CEREBRAL SYPHILIS

WITH REPORT OF A CASE OF CHRONIC MENINGO-
ENCEPHALITIS SECONDARY TO SYPHILIS

BY

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601

CEREBRAL SYPHILIS.

WITH REPORT OF A CASE OF CHRONIC MENINGO-
ENCEPHALITIS SECONDARY TO SYPHILIS.¹

BY CHAS. NORTON BARNEY, M.D.

THE patient whose case I am about to report entered the Massachusetts General Hospital first in July, 1895, on the service of Dr. Gannett, having been recommended for admission by Dr. James Putnam. She was at that time twenty-five years of age, and had already borne four children, the youngest of which was four years old. For a year she had had ulcers on the left leg. For two months before entrance these ulcers had been treated with mercury and iodide and had improved much, but were not yet entirely healed.

Since February, 1895 — that is, for five months — she had complained much of severe frontal headache, and at intervals, for periods of a week or more at a time, had appeared very dull and stupid. Some vomiting without relation to food, and occasional dizziness, but no paralyses nor twitchings. Headache, vomiting, dizziness and stupor, then, through the spring.

In July, nine days before entrance, mental dulness became more marked, headache very severe, her speech became "thick," and she often used the wrong words. The next day tingling in the little finger of the left hand, then of the whole left arm, and, later, paralysis

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601.

of the arm. In two or three days more her face became drawn to the right, and the next day the paralysis involved the left leg. During the week, then, there were added to the previous symptoms of headache, vomiting, dizziness and stupor, a degree of motor aphasia, and left hemiplegia of gradual onset. There were also incontinence of urine and constipation.

Examination showed a fairly well-developed and nourished woman of small build, with dull expression and dull mental condition. She could answer questions, however, and recognize objects. There was slight internal strabismus at times. Movements of eyeballs were good. Pupils equal and reacting to light. Left facial paralysis not involving the upper segment. Paralysis also of the left arm and leg. There was some enlargement of glands in the neck, and scars of former abscesses. The knee-jerk was present on the right, but was not tested on the left because of the ulcers which extended from knee to ankle. No ankle-clonus on either side. Temperature, pulse and respiration were normal. Examination of the fundus of the eyes was negative. Examination of the urine negative. Blood examination showed no anemia and no leucocytosis.

During her stay in the hospital there were noted dulness, involuntary micturition, constipation, occasional headache and vomiting, aphasia, paralysis, and labial herpes. These symptoms rapidly cleared up under iodide administered by mouth and by rectum in doses pushed to 180 grains a day. The mental condition was the first to improve, becoming normal in four days. The paralysis disappeared first in the face, then in the arm, and last in the leg. The patient was under treatment one month. At the end of that time her intellect was perfectly clear, and she was able to walk several blocks with no help but a cane.

All the late summer and early autumn her husband considered her practically well. Catamenia did not appear in October nor in November. At this time she began to show slowness of thought and speech, failing memory, and "said odd things." Early in December she had an abortion and her uterus was curetted. Under iodide her mental symptoms improved. Later, however, headache became continuous and especially severe at night.

At the end of January, 1896, about a year after the first appearance of cerebral symptoms, she entered the hospital a second time, on the service of Dr. Fitz. She was unable to give any account of herself, but her husband stated that the chief symptoms had been mental confusion, thick speech and headache; and also, that for two days she had complained of tingling in the arm and lip, the symptoms which preceded the hemiplegia of six months before. This time, however, the tingling was on the right, whereas before it had been on the left. There had been no vomiting, and the eyes had been straight.

On examination, temperature, pulse and respiration were found normal. Nutrition and color not as good as at the examination six months before. Expression very dull. Questions were answered slowly or not at all. The eyes showed nothing wrong. Chest and abdomen were negative. There was ankle-clonus on the left, and a tendency to clonus on the right. Both knee-jerks were active. The patient moved the left foot only very slightly, and the right foot not at all. All movements were weak and sluggish, but possibly partly on account of hebetude.

The night of entrance she became noisy, was transferred to a private room, tied in bed, and given trional. Antisyphilitic treatment was begun. Mercurial ointment half a drachm by inunction every night, and

iodide forty grains a day, to be increased fifteen grains each day. For the next week she was delirious most of the time. These days are perhaps worth reporting in detail.

February 1st. The temperature, previously normal, was now ranging between 99° and 100° , with a corresponding rise in the pulse. Patient shrieking when nobody was in the room, and crying when anybody entered. She was emotional and forgetful. Got out of bed unless tied. Slept only after hypnotics. Trional fifteen grains, and sulphonal ten grains were ordered to be given every morning and night if the patient were noisy.

On the second day she was in the same condition, shrieking and trying to get out of bed if alone and not under hypnotics. Hallucinations and delusions. Emotional. She shrieked all night, in spite of trional and sulphonal, and of hyoscine given subcutaneously.

On the third day she slept in naps, but would shriek as soon as she awoke; and when anybody would come into the room she would cry out that she was being killed. She heard the voices of her friends and children, and had various other hallucinations. The daily dose of mercurial ointment given by inunction was increased to one drachm, and the order for trional and sulphonal changed to ten grains of trional and five of sulphonal every six hours if necessary. Three doses were required on this day.

On the morning of the fourth day she was awake and quiet. She screamed very little and got along with only one dose of the hypnotic.

On the fifth day she was crying, but not yelling. This day also she required only one dose of the trional combination.

The sixth day she was perfectly quiet and required

no hypnotic. Headache was complained of and relieved by phenacetin.

On the seventh and eighth she took trional and sulphonal twice.

By the tenth she was rational, bright and quiet. She took a hundred and sixty-five grains of iodide, and showed no toxic effects from either iodide or mercury.

On the thirteenth she was taking one hundred and eighty grains of iodide and was kept at this dose from this time on. She had not required trional for two days and did not take it again. She was not noisy, and only occasionally had to be tied in bed.

By the eighteenth she had still shown no toxic symptoms from the mercury and iodide. It is interesting in connection with the question of the toxic effects of trional that this patient took considerable doses without evil result. She had improved remarkably and was discharged from the hospital to go home in care of her husband.

After this iodide treatment was neglected. She was thought to be pretty well, though all her mental faculties were still slightly dulled, and before long was trusted to walk alone daily to the house of a friend. One day she started out on her walk as usual, but got lost and turned up in Tewksbury. This was about three months after she left the hospital. Then a woman was hired to look after the house and children. The patient's mind was becoming more dull again; her memory was very poor; she was emotional and occasionally irritable. There were no hallucinations. Ptosis appeared, and disappeared. At one time she was jealous and threatened to kill her husband; but when he put a carving-knife in her hands, she broke out crying. After this he slept with her wrist strapped to his, so she should not get out of bed

without his knowing it. At this time defecation and micturition were involuntary. She could not dress herself. She could not read. Her eyesight was good, but she could not tell one word from another. She had been getting distinctly worse all summer, but in August she was delivered of a living child, and then improved. The same improvement was noticed for a while after she had the miscarriage the previous December. The baby died in one month. The cause of death was stated as "cholera infantum."

October 3d I saw her for the first time since she had left the hospital. She was slow of comprehension, but talked fairly well; was polite, and apologized for the boisterousness of her children. She remembered nothing about the birth of her last child till certain circumstances were called to her mind; and after I left she remembered nothing of my visit. She had no delusions of grandeur, and no tremors of tongue and lips in talking. The only remains of the old hemiplegia were seen in a weakened grasp on the left side.

The next day she had a general epileptiform convulsion — the first. There was no warning. There was no cry. Her husband heard the thud of her fall, and, running in, found her unconscious, stiff, blue and twitching. The convulsion lasted about a minute and left her sleepy. Iodide treatment was now begun a third time.

Long before any nervous symptoms developed in this case the diagnosis of syphilis was made by the dermatologists. Syphilis was diagnosed at the patient's first stay in the hospital, from the irregularity and remittance of the symptoms and from the effects of antisiphilitic treatment. Again, at her second stay in the hospital the diagnosis of syphilis was made by independent observers. Her subsequent progress has

confirmed the diagnosis. It is interesting that no history of infection can be obtained. The husband has denied secondary as well as primary symptoms, and presents no evidence whatever of syphilis on the most careful examination. The children are, and have been, perfectly healthy. The following data of past history and of family history are noteworthy. The patient has never been perfectly well. Throughout childhood she presented glandular and skin lesions. All her brothers and sisters died in infancy or childhood—most of them in convulsions. Her father and mother led very dissolute lives, but the causes of death could not be learned. In considering the possibility of hereditary disease the question comes up whether these facts are admissible as evidence.

ETIOLOGY OF CEREBRAL SYPHILIS.

Cerebral involvement in syphilis may occur at any time from a few months to twenty or even thirty years after infection. It is not necessarily a late manifestation. Over half the cases occur within three years, and as syphilitic infection is most common in young adults, so is the cerebral involvement. It is more likely to develop in cases where the skin lesions have been insignificant, perhaps because anti-syphilitic treatment in these cases is more commonly neglected. Causes weakening the nervous system are thought to predispose to the localization of the disease in the brain. Such are trauma, meningitis and sunstroke, in addition to excesses and the other usual causes of neurasthenia.

PATHOLOGICAL ANATOMY.

Brain syphilis may result by extension of the disease from nodes in the bones of the skull, but is usually secondary to lesions of the membranes and

vessels. These lesions consist of circumscribed gummata, large or minute, single or multiple, or of a diffuse gummatus infiltration in single or multiple patches. They extend from the membranes and involve the cortex, especially the anterior and middle portions of the base. The gummata are at first red and soft, vascular and diffuse, but later become yellow and hard, fibrous, shrunken and more definitely circumscribed. Gummatus infiltration of arteries results in stenosis and thrombosis. Secondary results are hemorrhage, softening, or atrophy of the brain tissue.

SYMPTOMATOLOGY.

The symptoms of brain syphilis are variable on account of the variety in the nature and situation of the lesions, but there are certain general symptoms common to most cases and there are certain characteristics in the mode of development of symptoms.

Headache is ordinarily the first complaint. It is constant for considerable periods of time, and grows progressively more severe, though having remissions and exacerbations from day to day or week to week. It is, as a rule, more severe toward evening, and at night may be unbearable. *Insomnia* is another early symptom. *Torpor* comes next. The patient sleeps little, but is dull and stupid much of the time. He loses power of concentrating attention. *Memory* fails. *Dizziness* may be troublesome. These symptoms, like the headache, are progressive and remittent.

Finally, after days or months, aphasia, convulsions, paralysis, coma, delirium or dementia develop, and the other symptoms become less prominent. These symptoms also are slowly progressive, continuous or intermittent.

The *disturbances of speech* vary according to the

centres involved. Any form of aphasia may be present, and may be the only localizing symptom. Motor disturbances are perhaps more frequent than sensory.

Paralysis also is more likely to be motor than sensory. The same lesion which produces the aphasia may extend and involve the adjoining motor area. The paralysis, unless due to hemorrhage, advances rather slowly, and has its exacerbations and remissions like the other symptoms. Loss of power is slight or complete. Hemiplegia and paralyzes of cranial nerves are the usual forms. Optic neuritis is common.

Disturbances of cranial nerves and convulsions are not often prominent symptoms in the same case, for the former are due to gummatous growths about the base of the brain, and convulsions, as a rule, to growths on the convexity.

Epilepsy, secondary to syphilis, may take the form of convulsions without loss of consciousness, loss of consciousness without convulsions, or both convulsions and loss of consciousness. It is to be distinguished from idiopathic epilepsy by the recognition of the existence of syphilis, by the precedent paroxysmal headache, by precedent or coexistent mental disturbance, optic neuritis, aphasia, hemiplegia or other paralysis, by the results of treatment and by the age of the patient at the first onset. Excluding uremia, hysteria and alcoholism the onset of epileptiform convulsions in a patient over twenty is probably due to syphilis. The aura and cry are not so often present as in idiopathic epilepsy.

Changes in mental condition due to syphilis are hebetude, delirium, coma, confusional insanity, hallucinations, or a dementia resembling general paralysis of the insane. There has been much dispute as to the nature of the dementia following syphilis. Mickle and Fournier conclude that syphilis produces a de-

mentia resembling general paralysis, but that the two are not identical. A tenable view is that general paralysis is not necessarily gummatus, but may be secondary to gummata as well as to other causes. The lesion is a chronic meningo-encephalitis resulting in atrophy of the cortex, or encephalo-malacia. Perhaps the division of general paralysis into an idiopathic and a secondary variety, as with epilepsy, would clear up the confusion. The characteristics of the dementia secondary to syphilis, as opposed to the idiopathic general paralysis of the insane, are as follows: Precedent syphilis; precedent headache, continuous, intense, worse at night; precedent or coexistent hemiplegia, aphasia, or paralyse of cranial or other nerves, having the irregular characteristics of development and course already described. Greater variety and more fantastic combination or sequence of symptoms. Absence or inconspicuousness of the delusions of grandeur and of the tremors of face and tongue. And, finally, the beneficial effects of thorough antisiphilitic treatment upon many of the symptoms.

PROGNOSIS AND TREATMENT.

As prognosis in cerebral syphilis depends largely upon treatment these two will be considered together. Gummatus lesions are favorably influenced by iodide and mercury; secondary lesions are not. Treatment of the gummatus lesion is so effective that a case may present the very gravest symptoms of severe brain disease — delirium, convulsions, coma — and yet get well. Whereas treatment of the secondary changes is ineffective, though these are usually associated with syphilitic lesions susceptible to iodide. Prognosis, therefore, depends not so much upon the extent and situation of the syphilitic lesions as upon their age at the beginning of treatment and conse-

quently the nature and extent of the secondary changes which they have set up. Earliness of treatment, then, is a most important consideration in prognosis. And as this depends upon the earliness of diagnosis the importance of making a correct diagnosis at the earliest possible period is supreme. As these patients are at any time liable to become blind, or to die in an epileptiform attack, or of hemorrhage from a gummatous artery, it is important also to get them under the influence of antisyphilitic drugs as soon as possible. For this reason mercury should be given as well as iodide, and both in doses rapidly increased to the limit of toleration. The mercury may be given in the form of mercurial ointment by inunction, in doses of half a drachm to a drachm or more every night, unless toxic symptoms appear; and at the same time iodide, beginning with thirty drops of the saturated solution a day, and increasing at first rather slowly to a drachm, and later, if toxic symptoms are not conspicuous, increasing very rapidly. If the patient is vomiting, the iodide may be given by rectum. Two hundred grains a day by mouth is not at all an extraordinary dose. Ten grains, three times a day, constitutes neither a reliable therapeutic test nor efficient treatment.

The practical points of this paper are the importance of the early diagnosis of cerebral syphilis, the importance of early treatment, treatment by mercury and iodide together, and both increased as rapidly as possible to the limit of toleration.

