Chronic progressive softening of the brain : report of cases, with autopsies simulating cerebral tumor / by J. Ramsay Hunt.

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CHRONIC PROGRESSIVE SOFTENING OF THE BRAIN: REPORT OF CASES, WITH AUTOPSIES SIMULATING CEREBRAL TUMOR.[1]

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Wernicke, in his *Lehrbuch der Gehirnkrankheiten*, published in 1881, describes a form of chronic progressive softening of the brain not dependent upon and having no evident relation to vascular disease. This form he distinguishes sharply from the encephalomalacia of embolic or thrombotic origin.

The importance of this group of cases is emphasized because of the resemblance which they bear in their clinical course to that of abscess and tumor of the brain. The affection is rare, and while not a few of the older cases described under this head are relegated to the domain of tumor or abscess there still remain five cases—four the observations of Andral and one of Birch-Hirschfeld—which he regards as unassailable.

Clinically the cases are characterized by the gradual development of hemiplegia accompanied by irritative sensory symptoms and more or less anæsthesia, but without apoplectiform manifestations or very severe general cerebral symptoms.

The focal symptoms continue to progress slowly for months, a year, or even longer, then ceasing to progress and remaining stationary. It is during the progressive stage that the resemblance to tumor is most striking.

Postmortem, areas of white and yellow softening were found in the centrum semiovale, invading the basal ganglia and rarely extending to the cortex. The vessels of the circle of Willis, while perhaps showing some arteriosclerotic changes, were patulous and free from occlusion, in the distribution corresponding to the broken-down and softened brain tissue.

This type of brain softening occurring without apparent relation to vascular obstruction, was described by the older writers on

[1] Read at a meeting of the New York Neurological Society, April, 1906.

encephalomalacia, and since its resurrection by Wernicke has received but little attention.

It is not included in the descriptions of this subject in the modern systems of medicine and pathology.

Gowers, however, in his *Diseases of the Nervous System* (1893), accords it a brief description and states that "although it is not probable that the condition is directly related to arterial disease, the latter is present in most senile cases and may have an indirect influence in determining the lesion."

Oppenheim also briefly outlines the condition under the somewhat cautious heading of "The So-called Chronic Progressive Softening of the Brain," and cites a case observed by him in which the circle of Willis was free, but the carotid artery in the neck was obliterated by arteritis and thrombosis.

Brissand and Massary, in 1890, recorded a somewhat similar case, in which an arteriosclerotic constriction of the upper portion of the carotid produced œdema and softening with progressive hemiplegia. Prevost and Cotard in their monograph on the *Ramollissement Cérébrale* (1860) record four cases of brain softening with hemiplegia without evident vascular lesions. By "evident" vascular lesions is meant in these cases, as in those quoted by Wernicke, an obliteration of one of the main trunks of the circle of Willis corresponding to the focus of softening.

In the light of the more recent investigations in encephalitis it is possible that some of these cases would fall into this group. In any event, modern literature is singularly free from observations which would justify the view of Wernicke that such an affection exists which is not of vascular origin.

A unique example of brain softening has been described by Henneberg, in a woman aged thirty-two years, the symptoms beginning during the puerperium. The duration of the affection was four months and severe general cerebral symptoms were present from the first and persisted throughout; a progressive hemiplegia gradually developed, later the leg of the opposite side becoming involved. In addition to extensive areas of softening in the brain, the spinal cord showed similar changes, but in a lesser degree. The examination of the bloodvessels was negative and the author concludes that the softening was of toxic origin, similar in nature to the pseudosystemic degeneration of the cord occurring in the anæmias and various toxic and cachectic states.

Two cases of chronic progressive softening of the brain have come under my own observation, both dependent upon obliteration of the central perforating arteries supplying the region of the basal ganglia and the internal capsule.

In one of the cases an exploratory operation for tumor was performed, and in the other an optic neuritis for a time rendered the diagnosis of tumor very probable.

SUMMARY OF CASE I.—(From Bellevue Hospital, I. Medical Division, service of Dr. V. H. Norrie.)

The gradual development of paræsthesiæ, objective sensory disturbances, and weakness, affecting the face, arm, and leg in succession, in a man aged fifty-three years. Severe paroxysmal headaches in the left parietal region and vertigo, no vomiting, no optic neuritis.

Diagnosed as a progressive lesion, probably tumor, beneath the cortex of the face and arm centres and exploratory operation performed. Operation was negative and was followed by complete right hemiplegia, stupor and incontinence of the sphincters. Patient died one month later.

Postmortem old areas of softening were found in the basal ganglia, internal capsule and the sub-cortex of the face and arm centres. In these sclerosed and degenerated areas obliterated vessels were present. The origin of the softening is to be referred to a localized arteriosclerosis at the junction of the internal carotid, middle cerebral, and posterior communicating arteries with progressive and successive obliteration of the small central perforating branches; the main arterial trunks were excluded by a recent thrombus. An abscess was situated beneath the leg centre invading the white matter, a sequela of the exploratory punctures.

HISTORY. The patient, a mechanic, aged fifty-three years, was admitted to the Bellevue Hospital July 27, 1902. He had been very moderate in the use of alcohol, and denied having had any venereal disease.

He had enjoyed unusually robust health and had not been ill since childhood until one and one-half years before admission to the hospital, when he was suddenly seized with vertigo, and lost consciousness, while working at a turning-lathe. He remained in an unconscious state for nearly two hours, but was unable to say whether or not he had had a convulsive seizure.

He remained in bed for two weeks suffering from weakness and vertigo on attempting to arise. At the end of that time he returned to his occupation, regaining his health and strength completely, which continued until six months ago, when he had another vertiginous seizure at the turning-lathe, but did not lose consciousness. He was obliged to give up his occupation and remain at home, owing to general weakness and vertiginous seizures. Two months later headaches made their appearance over the vertex and left parietal region. These would come on suddenly, were very severe, and would disappear as quickly as they came on. They would last from a few minutes to several hours, and at times were very intense. Between these paroxysms the head was comparatively free from pain.

After three months of parietal and vertical headaches and vertiginous seizures paræsthesiæ made their appearance on the right cheek and about the angle of the mouth. They consisted of prickling,

numbness, formication, and a thick sensation, as if the parts were swollen. These persisted and increased in extent until finally the whole right side of the face below the eye was involved.

A few weeks later a similar prickling and tingling was noted in the fingers of the right hand, later involving the thumb. These paræsthesiæ of the fingers gradually ascended the hand and the forearm, so that on admission there were present evidences of sensory irritation on the right side of the face and right upper extremity to the elbow, beginning in the cheek and progressing gradually. During this period the paroxysms of headache in the left parietal region continued and at times were very severe.

While some sensory symptoms were constantly present in the face and arm they were subject to acute exacerbations lasting from fifteen minutes to a half-hour, and this aggravation of sensory manifestations often connected with the paroxysms of headache. Several times the hand and fingers were fixed in prolonged involuntary extensor spasm, lasting a quarter of an hour, and on one occasion almost an hour.

There had been no speech disturbance and no aphasic attacks, and except for general weakness the legs were not affected. There had been no nausea or vomiting.

Status Præsens, July 28, 1902. A tall, robust, well-built man, florid complexion. Gait and station normal.

A drooping is present on the right side of the face at the angle of the mouth, both at rest and on voluntary innervation; on emotional innervation the two sides contract symmetrically. The tongue deviates toward the right side. The other motor cranial nerves are normal. No gross defect in the motility of the right upper extremity is present, excepting in the hand, the grip of which is distinctly reduced in power. There is no tremor and no ataxia.

The tactile and pain sensibility is diminished over the right side of the face, right hand, and forearm, while thermic sensibility and the articular sense are undisturbed. The localization of tactile impressions is also disturbed. While the stereognostic is not absolutely lost, objects are recognized with difficulty and some not at all.

There are no paræsthesiæ or sensory disturbances noted in any other parts of the body and the gross motor power of the lower extremities is normal. The arm jerks on the right side are exaggerated and more active than on the left. The knee jerks and Achilles jerks are present on both sides and equal in intensity. Plantar flexion of the toes on both sides (no Babinsky).

The cremasteric and abdominal reflexes are present on both sides. The pupils are equal and react normally.

The heart is normal in its outlines, the sounds are not accentuated and no murmurs are audible. The radial shows only moderate thickening and the pulse is full and regular, beating eighty times to the minute.

The urine is negative and the other viscera present no abnormalities.

The optic nerves are normal. Percussion of the head over the left parietal bone was tender, but no difference in the percussion note could be made out.

NOTE. August, 1902.—The patient was placed upon ascending doses of potassium iodide and for a short time the headaches improved but later returned with all their old severity in spite of vigorous antisyphilitic treatment. The weakness in the right hand and arm increased gradually and in September the right leg felt weaker, the right knee jerk became exaggerated, and pseudo-ankle clonus was elicitable, but no Babinsky. The temperature was normal and the pulse rate ranged between 72 and 90. The condition was evidently slowly progressing and the advisability of an exploratory operation was considered by Drs. Vanhorne, Norrie, and myself.

On October 22, 1902 the patient was examined by Dr. Dana, who concurred in the diagnosis of a progressive lesion beneath the centres for the face and hand and the advisability of surgical exploration. The case was transferred to the surgical ward for operation, in the service of Dr. Bern B. Gallaudet.

Operation, October 29, 1902, by Dr. Gallaudet. An oval flap of bone was removed uncovering the left Rolandic area over the face and arm centres. The brain was rather less prominent than is usual and pulsated normally. The cortical convolutions were normal in appearance. A needle was introduced four times into the suspected territory, in hope of puncturing a possible cyst, but with negative results. The dura was then sutured and the disk of bone replaced.

On the following morning, October 30, 1902, the patient had a convulsive seizure limited to the right side, without loss of consciousness, and was found completely paralyzed on the right side.

The wound was opened, the disk of bone removed and with it a a small blood clot on the outer surface of the dura. The bone was replaced and the wound sutured. No more convulsive seizures occurred but the complete right hemiplegia persisted, with ankle clonus and the Babinsky reflex and loss of the skin reflexes on the right side. Patient was dull and apathetic and passed urine and feces incontinently. Pain impressions are felt at both sides but less keenly on the right.

Because of the profound mental state of the patient and the completeness of the hemiplegia, it was thought probable that the exploration had been followed by hemorrhage. The man survived the operation one month, the right hemiplegia remained complete. The stupor, somnolence, incontinence of urine and feces continued.

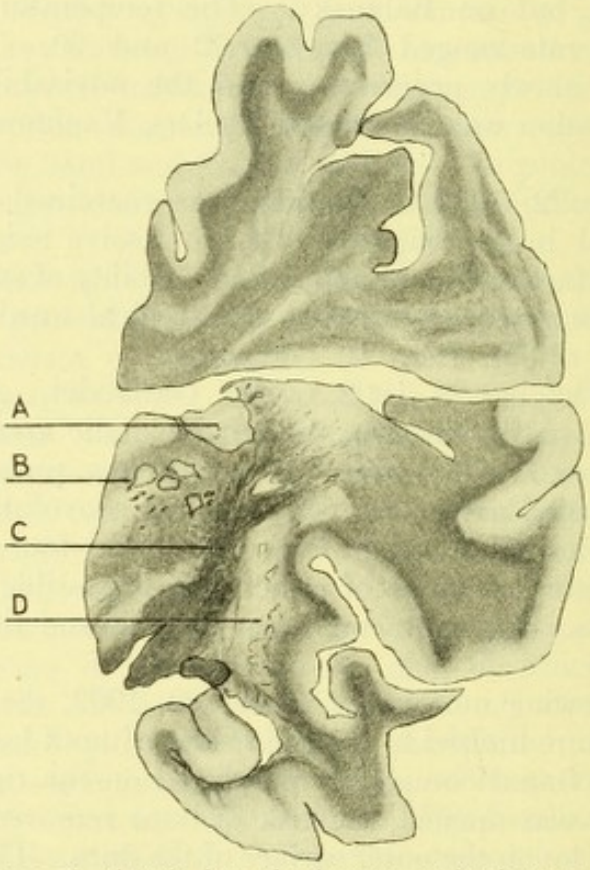
The temperature ranged from 100° F. to 101° F. and 102° F., but without marked remissions. No chills, no more convulsive

seizures. Pulse rapid and in proportion to the fever; no bradycardia; no rigors. The right leg gradually assumed a state of flexion contraction.

Death occurred on October 29, 1902, having been preceded by a rise of temperature to 104° F.; pulse, 150.

Postmortem Examination. (See Fig. 1.) A general autopsy was refused, but permission was granted to enlarge the skull and examine the brain. By this means it was found possible to remove the entire left hemisphere, with the parietal dura mater.

FIG. 1



Showing small areas of sclerosis, loss of substance, and minute cavities in the basal ganglia (*A*, *B*, *D*) and internal capsule (*C*), produced by obliteration of the central perforating arteries.

The dura was bound to the leptomeninges over the leg centre by inflammatory adhesions. On separating these it was found that the cortex of the posterior central convolution, over the leg centre, was broken down and gangrenous. This rupture in the cortex opened into a conical abscess cavity directed downward and inward, containing thick hemorrhagic pus.

The convolutions in the immediate neighborhood were enormously swollen and flattened out, but there was no general purulent meningitis, and the other convolutions presented a clean and unbroken surface. With the left hemisphere was removed also the left half of the

circle of Willis. These vessels present some general thickening and a few disseminated plaques of arteriosclerosis.

The internal carotid, however, and the adjacent portion of the middle central and posterior communicating at their origin show an extreme degree of arteriosclerosis, and on dissecting them out carefully the small perforating branches which pierce the anterior perforated space appear as the finest thread-like prolongations and are evidently atrophic.

The Sylvian artery and its branches beyond this point are patulous throughout and free from sclerotic plaques.

The hemisphere was then divided systematically by a series of transverse vertical cuts at right angles to its long axis. The abscess cavity was found to be pear-shaped, its base to the periphery, and extending downward and inward through the centrum semiovale. It is limited by a pyogenic membrane and the adjacent white matter is extremely œdematous and contains a few small hemorrhagic and purulent foci.

Beneath the cortex of the face and arm centres the white matter is softened, gray and firm, containing a few small irregular cavities

Very striking are the changes noted in the region of the basal ganglia. The caudate and lenticular nuclei, the optic thalamus and the internal capsule are the seat of small cavity formations, presenting sharp, irregular outlines and varying in size from a pin's head to a grain of wheat, a few even larger. The nerve substance between these cavities is firm and normal in appearance, and the abscess cavity, with its œdematous and inflammatory areola, does not encroach upon this area.

Histological Examinations. Bloodvessels. A series of sections of the sclerosed portions of the internal carotid, middle cerebral, and posterior communicating arteries were stained by the hæmatoxylin-eosin method.

All three of these vessels present an extreme arteriosclerosis of about one-half of the circumference of the vessel wall, with great proliferation of the intima.

At the junction of the media and intima the endothelial proliferation has undergone retrograde metamorphosis, and is broken down and degenerated; in some sections a moderate round-cell infiltration of the adventitia is also present A fibrin clot, containing a few leukocytes and red cells, is contained within the lumen of the vessels in the sclerosed areas and is adherent to the intima in a few of the sections; there are, however, no evidences of organization or old thrombus formations. The middle cerebral artery, distal to the sclerotic plaque, is entirely free from any thrombus formation.

Many of the small arterial branches, taking their origin from the sclerosed portions of the internal carotid and middle cerebral arteries, present various stages of obliteration, old thrombus formation, and organization.

Brain. Vertical sections of the whole hemisphere through the ganglia were stained by the Weigert-Pal method (see Fig. 1), and small blocks of tissue from various portions of the basal ganglia, the cortical face and arm centres were prepared by the Pal, Van Gieson, and hæmatoxylin-eosin methods.

The basal ganglia and the internal capsule contain small sclerotic areas, small irregular cavities without a trace of any nerve structure, or only a few remnants of myelin substance and swollen axis-cylinders. In or near these areas remnants of obliterated vessels, some containing organized thrombus, are demonstrable.

The sub-cortex of the face and arm centres has lost the larger portion of its myelin constituents and consists essentially of a glia and connective-tissue network. The cortex of the leg centre and adjacent white substance are the seat of active purulent inflammation, hemorrhages, and large accumulations of polynuclear round cells. No evidence of acute inflammatory change or abscess formation were present in the region of the basal ganglia or the face and arm ends of the cortex.

Summary of Case II.—(From the Montefiore Home for Chronic Invalids.)

Following a head injury, a man aged fifty-seven years had headaches and developed weakness and stiffness of the right leg, which persisted. Five months later appeared dysarthria and weakness of the right arm, and in a few months the left leg became involved. There was apathy, dulness, great mental deterioration, explosive laughter, and at times stupor. Beginning optic atrophy consecutive to a neuritis. No essential progression of the focal symptoms during eight months' observation, but progressive mental deterioration, later becoming bedridden and incontinent. Glycosuria was present throughout.

Autopsy. Obliteration by old organized thrombi of central perforating arteries supplying region of internal capsule and basal ganglia, with softening and sclerosis. In addition extensive breaking down of the white matter forming the walls of the lateral ventricles, extending even into its cornual prolongations, but without the usual histological evidences of softening or inflammation, the only reaction consisting of minute extravasations and infiltrations of red blood cells.

History. Owing to the mental deterioration of the patient and the ignorance of his relatives, it was impossible to obtain a very detailed history of the progression of the symptoms, but the following facts were obtained and confirmed by various members of the family:

The patient, a man aged fifty-seven years, a tailor by occupation, was admitted to the Montefiore Home October 14, 1902.

He had indulged excessively in tobacco, was very moderate in the use of alcohol, and denied any venereal disease. His wife had

borne four children, one dying in infancy the others living and well. No miscarriages. His health previous to the onset of the present trouble, which dates back two and a half years, had been excellent. At this time he was struck by a trolley car and dragged a short distance on the fender. He was cut about the head and considerably bruised, but did not lose consciousness, and was removed to his home in an ambulance. He remained in bed two weeks, during which time he had headaches. After recovering from the acute effects of this injury it was noticed that the right leg dragged, but he still continued his occupation of tailor, the stiffness and awkwardness of the right lower extremity becoming more and more apparent.

Four or five months later on arising in the morning he had some difficulty in articulating, the speech was thick and the right arm was weak. This condition persisted and continued to grow worse. Several months later the left leg also became weak and awkward, walking became very difficult and the speech more indistinct. He had had no convulsive seizures and had not lost consciousness, but complained frequently of headaches and vertigo and of pains and paræsthesiæ in the extremities.

While an inmate of the Home he was frequently examined by Dr. Joseph Fraenkel, Dr. I. Abrahamson and myself, the result of our several examinations may be summed up as follows:

Mentally the patient was dull, heavy, and apathetic. Memory was very defective and he could only grasp and answer the simplest questions frequently repeated. The simplest problems in arithmetic were answered incorrectly. He, however, recognized his surroundings and knew those about him, but evinced very little interest, and would stare vacantly before him.

The degree of mental apathy varied from time to time. During his better days he would sit in a chair holding his cane in the left hand, taking little or no notice of his surroundings, while on other occasions stupor and somnolence were marked. He was aroused with difficulty and passed the urine and feces in the bed.

He is able to stand alone with the eyes closed and can walk with the aid of a stick, but the gait is slow, labored, and stiff, especially the right leg, while the right arm is held in hemiplegic attitude. The steps are short and shuffling and the legs held apart. On lifting the arms, neither can be elevated in a normal manner, but the right is much weaker and slower than the left. The right side of the face at the angle of the mouth droops and the tongue on protrusion deviates toward the right side.

The pharyngeal reflex is active, but on voluntary innervation the right side appears weaker. The ocular excursions are normal, no nystagmus, and the movements of the jaw are normal.

The pupils are at times equal, at times the right is wider. The reactions to light and accommodation, while sluggish, are present. The papillary skin reflex is not elicitable.

There is no tremor and no ataxia of the arms. All tendon reflexes are exaggerated, including the jaw jerk. On both sides ankle clonus is elicitable, but more pronounced on the right side. Slight wrist clonus is also present on the right.

The skin reflexes are present and are less active on the right side. On both sides there is plantar flexion of the toes. (The Babinsky reflex was not elicitable during the whole time the patient was under observation.)

The gross motor power of arms and both legs is considerably reduced, but the right side is weaker than the left.

As far as the mental state of the patient would permit sensation was examined. There was no gross defect of the special senses, of hearing, sight, smell, or taste and coarse touch, pain, and temperature stimuli were recognized over the arms and legs.

Dr. Karl Koller, who made the ophthalmological examination, found both disks in a state of beginning atrophy consecutive to an optic neuritis.

The speech was thick, indistinct, dysarthric, and while speaking, paroxysms of explosive laughter were occasionally observed.

The left ventricle was hypertrophied, there was a soft systolic murmur at the apex, and the second aortic sound was markedly accentuated. The radials were moderately thickened, and under increased tension. The lungs were hyperresonant and a few moist rales were heard in auscultation.

The urine contained a variable trace of albumin and an occasional granular cast. Sugar was also present, but would disappear under proper dietetic measures. The highest specific gravity recorded was 1029.

An examination of the blood made October 24, 1902, reads as follows: red cells, 4,480,000; white cells, 4,500; hæmoglobin, 73 per cent.

NOTE.—During the eight months of observation in the Montefiore Home little or no change was noted in the extent of the paralyses. The right hemiplegia with characteristic attitude persisted, the left leg remained stiff and weak, the left arm retaining fairly good power of movement, and having less involvement than the left leg. The mental deterioration, dysarthria, and general weakness and apathy grew gradually worse. The patient became bedridden, with incontinence of urine and feces. For a considerable time polydipsia was pronounced, with a ravenous appetite.

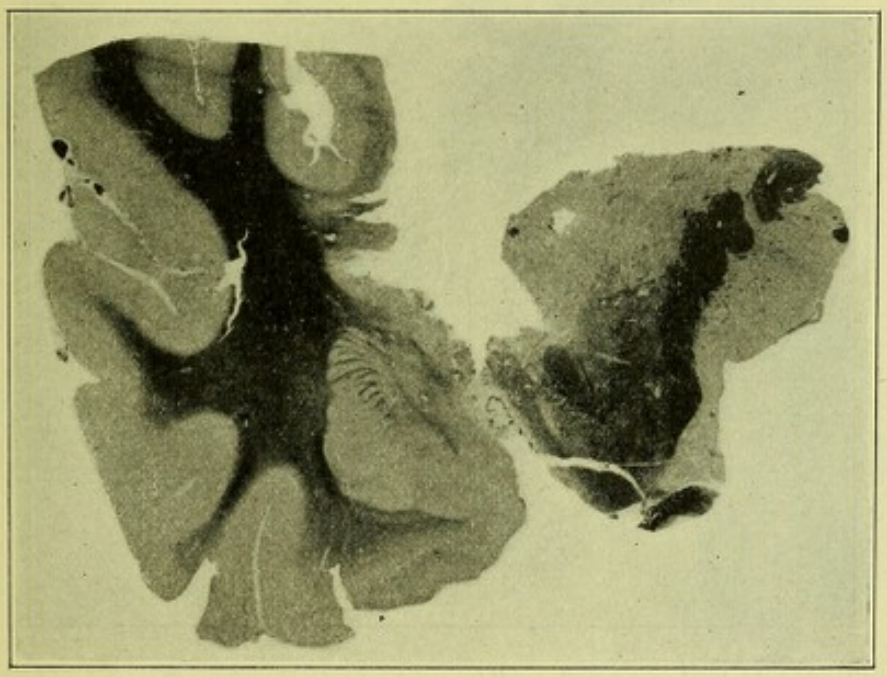
During almost this entire period there was a daily rise of temperature to 100° F., falling in the morning to normal; occasionally the temperature curve would rise to 102° or 103° F., but without chills or sweating. Evidences of old optic neuritis, with beginning atrophy, still persisted, and as far as the mental state permitted no gross defect in general or special sensation could be detected.

Sugar was present in the urine on repeated examination.

Postmortem Examination. Permission was granted to examine the brain only, which was removed within twenty-four hours after death and placed in a 10 per cent. formalin solution. The surface of the central convolutions show no areas of atrophy or breaking down. The leptomeninges, more especially over the frontal lobes, are thickened. The vessel walls of the circle of Willis are not sclerosed but are unusually thin and smooth, and contain a few disseminated plaques of arteriosclerosis. In both middle cerebrals a soft recent clot has formed, which is not present in the other vessels.

On turning back the corpus callosum, which is intact, the ventricular cavity presents a most unusual appearance. The cavity seems to be greatly enlarged, this having been produced by extensive

FIG. 2



Weigert-Pal method: left temporal lobe, basal ganglia, and internal capsule, showing softening of walls of lateral ventricle.

destruction and breaking down of the ependymal lining and neural structures forming the walls of the lateral ventricle and its cornual prolongations. (See Figs. 2, 3, 4 and 5.)

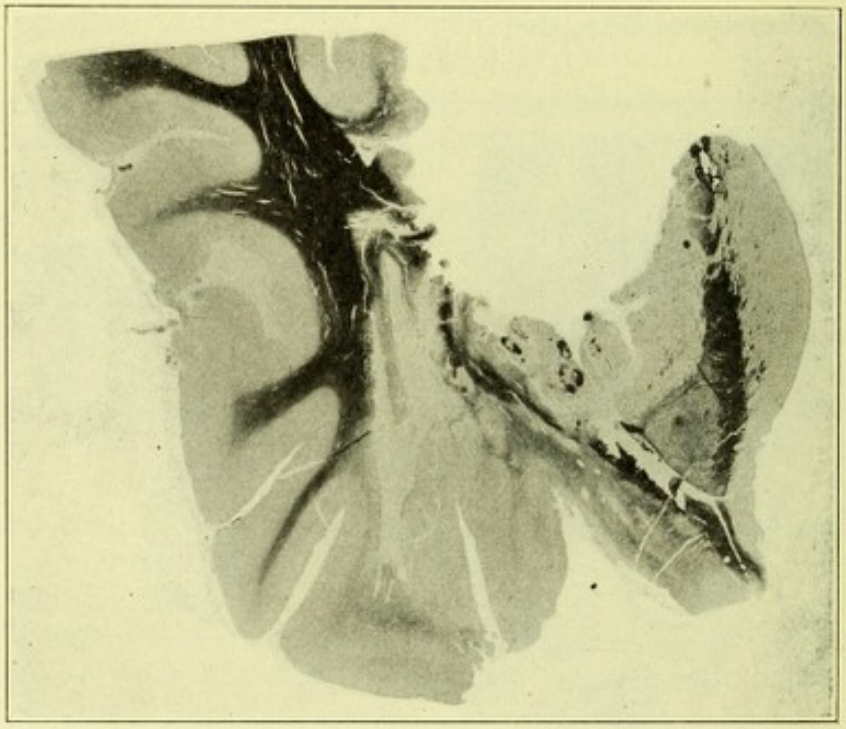
The walls of the third ventricle are well preserved, but the brain substance, external to the basal ganglia in the lateral ventricelly is undermined and eaten away, so that these structures are undue, prominent and stand out like mushrooms on both sides with ragged margins. (See Figs. 2 and 3.)

This destruction, or rather erosion, of the brain substance follows accurately the mould of the lateral ventricle, and diminishes in intensity but still quite apparent in its prolongations into the frontal temporal and occipital lobes. (See Fig. 5.) In a few areas the

ventricular lining is unbroken and in these portions the ependyma contains a few granulations.

The rough and uneven brain substance which largely forms the walls of this cavity is of a light-yellow tint and tapers off into normal white substance. Occasionally from the yellowish area are sent prolongations into the medullary columns of the cortex, and beneath the Rolandic area, where the defect is most pronounced, some of the medullary columns are involved to within a short distance of the gray matter. (See Fig. 4.) The gray substance of the cortex is intact throughout.

Fig. 3



Right temporal lobe, basal ganglia, and internal capsule, showing softening and breaking down of walls of lateral ventricles.

The interior of the cavity contains a cloudy fluid with crumbled tissue remnants, debris and several small recent blood clots.

The cerebellum, pons and medulla are normal macroscopically.

Both hemispheres from the frontal to the occipital poles were divided by a series of transverse cuts and histological studies conducted by the Marchi, Weigert-Pal, and hæmatoxylin-eosin methods. (Unfortunately, the medulla and pons were not preserved for histological examination.)

Histological Examination. Bloodvessels. The walls of the main trunk of the middle cerebral arteries are practically free from arteriosclerotic changes, except a very moderate degree of intima proliferation in certain areas.

Both middle centrals contain recent thrombi, composed of cor-

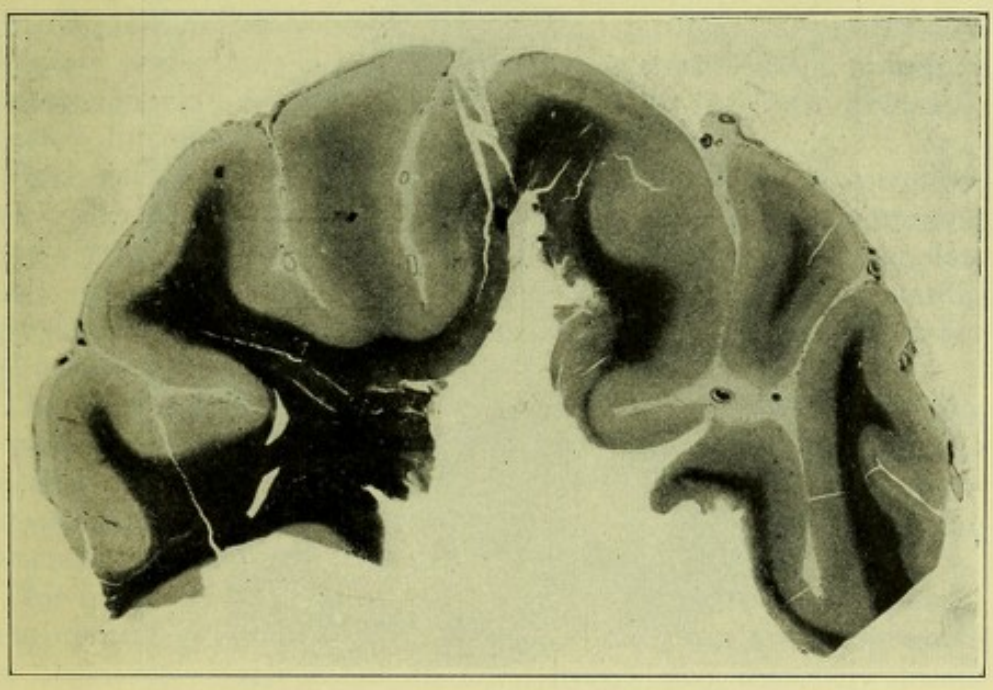
puscular elements held in a fibrin mesh, without evidences of proliferation or organization.

The vessels of the cerebral cortex passing through the meninges as well as the cortical perforating arteries show a high degree of arteriosclerosis, chiefly of the media and adventitia. In these smaller vessels the lumen is considerably reduced in size, but not obliterated.

The central perforating vessels supplying the ganglionic region and the internal capsule are not only greatly thickened, but many are included by old thrombi in various stages of fibrous organization.

Brain. These obliterated vessels lie on or near foci of old softening formation in the basal ganglia and internal capsule.

FIG. 4



Extensive breaking down of walls of lateral ventricle, extending to beneath the cortex of the parietal lobe.

Hemorrhagic foci, accumulations of blood pigment, perivasculai hemorrhages, numerous amyloid bodies, and granule cells (Marchr method) are demonstrable in these softened sclerosed areas.

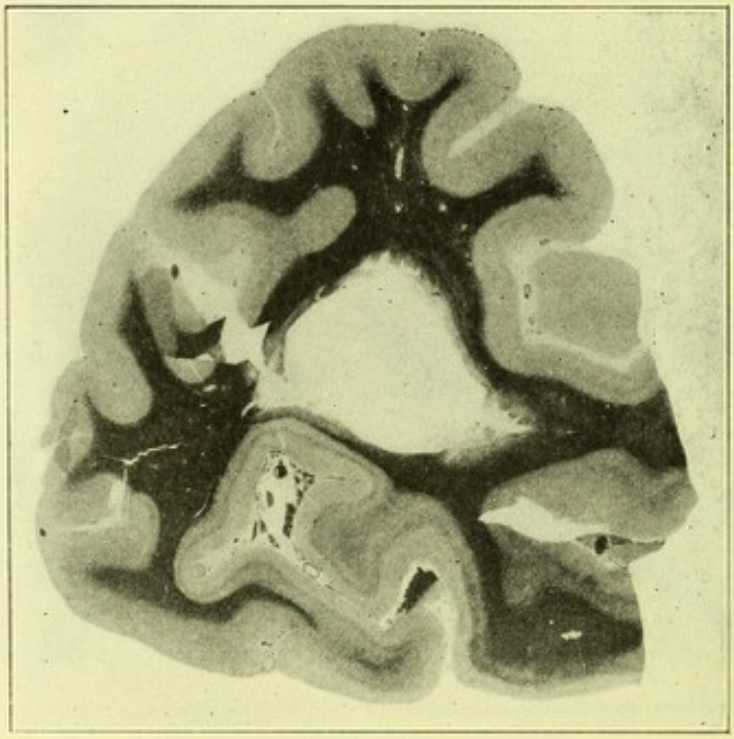
Of especial interest is the histological character of the softening in the ragged and crumbled walls of the lateral ventricle. This loss of the ependyma lining and subjacent white matter is extensive and beneath the parietal lobes is extreme, reaching almost to the gray matter, but nowhere in this marginal softening of the ventricles are granule cells or infiltrations of leukocytes to be seen. In the few spots on the lining of the ventricle where the ependyma has

been preserved the columnar cells are normal and pass abruptly into the ragged and eroded areas.

This ragged margin of eroded brain substance takes all the stains poorly, the sole evidence of reaction consisting of numerous microscopic hemorrhages, pericapillary hemorrhages, and infiltration of the layers of the tissue by red blood cells, but no leukocytes containing pigment or myelin droplets.

The tissue looks as if it had been macerated and eroded by the direct action of some substance, and except for the diapedesis and minute extravasations of the red blood corpuscles near the margin shows no other changes. The ependymal lining of the third ventricle is well preserved.

FIG. 5



Posterior horn of lateral ventricle in occipital lobe, with breaking down of the ependyma and subjacent white matter.

Optic Nerves. The sheath of the nerves, the endoneurium, and adventitia of the bloodvessels are thickened; no round-cell infiltration. In the centre of one of the nerves there is an accumulation of red blood cells. With the Weigert-Pal method the central fibres of the nerve trunks appear paler than normal; numerous corpora amylacea are present.

REMARKS. Case I. is of especial interest because of the clinical resemblance to brain tumor. The apoplectiform seizure which occurred one year before the appearance of any focal symptoms was of very doubtful nature; there was no paralysis at the time and the recovery was absolute and complete.

After the beginning of focal symptoms in the right side of the face the course was gradual and progressive, appearing next in the fingers, then in the hand and forearm, and later only slightly in the leg. During this entire period and even antedating the focal symptoms, severe paroxysmal headaches, usually unilateral, were present, with some variable tenderness over the left parietal region.

The absence of optic neuritis had given rise to serious doubts as to the real nature of the lesion, but this as is well known by no means excludes tumors, especially those situated in or near the cortex.

As all the internal measures employed had failed and as the condition was slowly progressing, surgical exploration seemed not only justified but imperative.

The ill effects of the surgical measures are to be attributed to exploratory punctures, with an aspirating needle, in search of a possible cyst.

In diseased and softened brain tissue, aspiration, no matter how delicately performed, is not without danger, and is liable to induce hemorrhagic extravasations, which break down and form favorable foci for infection.

Furthermore, there is no question that the dangers of infection, even with all modern aseptic precautions, are much more imminent when a softened, ill-nourished area of the brain is the seat of exploration. This danger should always be borne in mind when the central cortex is exposed and found normal and the question of further exploratory measures arises.

The unusual sequence of symptoms, so unlike that of softening in general, is to be referred to the parietal arteriosclerosis at the junction of the internal carotid, middle cerebral, and posterior communicating arteries, with occlusion of central perforating branches at their point of origin from the main trunks.

This lesion had induced a gradual and successive obliteration of these central perforating end arteries, producing the progressive and insidious development of hemiparesis and hemiparæsthesia with objective sensory disturbances. In brief, the picture of the so-called chronic progressive softening of the brain.

Case II.—While in this case the lesion had been progressive, involving first the right leg, then the right arm, and later the left leg, the development was by no means so constantly or uniformly progressive as in Case I.

The mode of onset and the acute exacerbations suggesting rather a vascular process than a neoplasm. As the symptoms in cases of vascular tumors not uncommonly suffer a sudden increase in the focal manifestations from hemorrhages, and as optic neuritis with beginning atrophy was present, the possibility of tumor in the region of the corpus callosum invading the Rolandic area on the two sides was for a time given serious consideration.

The marked inertia, extreme apathy, and mental enfeeblement

tended to favor this interpretation; but as the focal symptoms failed to progress materially during eight months' observation, a generalized arteriosclerosis and encephalomalacia came to be regarded as the probable underlying cause and the optic neuritis referred to the glycosuria which was present or possibly, as is rarely the case, dependent upon arteriosclerosis of the vessels of the optic nerves.

Pathologically, the case is of interest from the limitation of the obliterating thrombotic process to the central perforation arteries passing to the internal capsule and basal ganglia. The main trunks of the circle of Willis were singularly free from sclerosis, and the thrombi contained in the middle cerebrals were of quite recent origin.

More difficult of interpretation is the extensive breaking down of the walls of the lateral ventricles. This process cannot be regarded as an encephalomalacia in the ordinary acceptation of the term, as there is total absence of the usual histological changes accompanying such softening, viz., myelin degeneration and granule cells.

Only minute hemorrhages and red blood cells were present in the ragged margin surrounding the ventricles, which would seem to indicate that we are not dealing with a postmortem change or artefact. The periventricular localization of the breaking-down process is also unusual, and suggests an etiological relation to the contents of the lateral ventricle. The influence of diabetes and the tendency which it induces to tissue gangrene also arises as a possible predisposing or etiological factor.

No lesion of a similar character has come under my personal observation nor do I find it described in the literature.

I am inclined to interpret it as a periventricular softening, occurring very shortly before death, from an irritation or erosive action of the cerebrospinal fluid, in a brain the nutrition of which had been undermined by chronic softening and still further reduced by sclerosis of the cortical and central perforating end arteries.

In general the clinical picture presented by my cases, that of a progressive and gradually developing hemiplegia, and the character of the general cerebral symptoms resemble those described by Wernicke as chronic progressive softening, without evident vascular lesions. The main trunks of the vessels of Willis in my case were likewise free from occlusion, but histological study demonstrates the obliteration of the central end arteries in the ganglionic and capsular regions.

Such histological studies were not carried out in those cases in which Wernicke's description was based. It is readily conceivable how such central arteriosclerosis, with occlusion, would not only produce degeneration in the distribution of the terminal end arteries obliterated, but might also determine more extensive areas of softening in that portion of the white substance in the centrum ovale,

where the cortical and central circulation meet, and which represents an area of diminished nutritional resistance. It therefore seems to me that the existence of a progressive softening not dependent upon vascular disease of the type described by Wernicke may be fairly questioned, certainly until demonstrated by a case subjected to modern laboratory methods.

In addition to central arteriosclerosis as a cause of chronic progressive softening must be added constriction and obliteration of the carotid arteries in the neck.

The clinical importance of the subject lies in the possible resemblance to abscess, encephalitis, and tumor of the brain. Only recently, Williamson, in a paper entitled "The Importance of Progressive Hemiplegia in the Diagnosis of Brain Tumor," says: "When hemiplegia of gradual onset does occur there is no single sign of greater value in the diagnosis of cerebral tumor," and, again, "hemiplegia of gradual onset just described does not occur in central hemorrhage, embolism, or thrombosis." His report includes three cases of tumor of the brain with gradually developing hemiplegia, slight general cerebral symptoms, all without optic neuritis.

Ziehen, in a case with progressive hemiplegia, paræsthesiæ, and headaches, operated for a supposed abscess and found thrombosis of the vessels with softening. No doubt errors of a similar nature occur much more frequently than might be inferred from a perusal of the literature.

It should be emphasized that the diagnostic difficulties may be further enhanced by the presence of optic neuritis, as shown by one of my cases, and a case recorded by Zacher with extensive softening of both frontal lobes and double optic neuritis.

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