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FIVE HUNDRED AND THIRTY-FIVE DEARBORN AVENUE
CHICAGO
A CASE OF EXTENSIVE BRAIN DISEASE FROM ENDARтерITIS PROBABLY OF SYPHILITIC ORIGIN*

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The following case offers evidence of some interest in the occurrence of a diplopia explainable only on the functional activity of a small remaining portion of one visual field and of the retention of a very limited word-hearing capacity, in spite of the destruction of practically all of the left cortical auditory arrival-platform and the surrounding audiotypic skirt. From the history there can be little doubt that the condition was luetic and yet the microscopic examination failed to reveal evidence of the specific nature.

History.—The patient, a man of 32, with a negative family history as far as could be ascertained, was born and raised in Brookline, Mass. His childhood was uneventful. He proved to be a student of average ability, but left school at the third grade of the high school. After one year as an errand boy in a large department store he went into the leather business and rose rapidly to the position of assistant manager. At the outbreak of the Spanish War he enlisted with the Naval Reserve and was assigned to duty on the U. S. S. Prairie and served throughout the war. He was a moderate though not constant drinker. In April, 1903, he had a sore on his penis followed by a bad sore throat lasting two months and a rash lasting two weeks, and was under treatment for syphilis for one year.

On discharge from the navy he was in poor physical health and showed some irritability. In July, 1904, one year and three months after the primary lesion, he experienced some difficulty in using his right leg. This was transitory and

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was diagnosed as a rheumatic disturbance. Two weeks later he had a "shock" while at his place of business, which was followed by some loss of power in both legs. At this time it was noted that his memory and judgment lacked their former accuracy. The shock did not give rise to unconsciousness, but left a diplopia which necessitated the closing of one eye to cut out the second image. Later he lost the use of his right eye for a short period. Early in September he had a general epileptiform convolution and was sent to the Boston City Hospital where the diagnosis of cerebrospinal syphilis was made and where he remained six weeks under active iodid treatment. From the middle of October, 1904, to Jan. 1, 1905, he remained at home and during this time he had a general convolution, which was very severe, lasting three hours, and which was followed by aphasia. He deteriorated rapidly during this time, was often confused and disoriented, and finally became aphasic and filthy.

Jan. 1, 1905, he was committed to the Boston State Hospital where he remained until transferred to this hospital, March 28, 1905.

Examination.—On admission he was quiet and showed a dementia-like reaction. He showed partial word-deafness, paraphasia and jargon aphasia, would not obey requests, was unable to name objects, exhibited paralexia, paraphagia and continually closed his left eye in attempting to read or observe objects. A note at this time suggests that a hemianopsia was already present. On physical examination on the day after admission, the patient was quiet with expressionless features; talked freely without slur or tremor of words, but was markedly paraphasic and appeared demented. Many of the tests of special senses could not be applied on account of failure of cooperation. Reaction to pin-prick was prompt and equal on the two sides. The knee-jerks were exaggerated, especially on the right. Babinski was present on right; no ankle-clonus. The pupils reacted very sluggishly to both light and accommodation. He was unable to walk without support. The hand-grasp was weak, especially on right. The left angle of mouth was a little depressed. The right nasolabial fold was obliterated. There was no wrinkling of forehead on right. Ophthalmoscopic examination quoted as showing a probable subsiding neuritis with commencing consecutive atrophy.

Course of Disease.—During his residence in the hospital (a period of six years and four months) he remained for the most part quiet, either in bed or in a chair, though occasionally he was noisy, and on a few occasions he apparently reacted to hallucinations. He was filthy at all times. He fed himself spontaneously and used the spoon correctly and when attempting to write used the pencil correctly with
his right hand, showing no evidence of apraxia, though fur-
ther tests of this could not be carried out owing to the
dementia and lack of cooperation. His attention was easily
held and at times he seemed to recognize the physicians and
attendants. He was unable to give the alphabet and could
not name the days of the week or count spontaneously. At
times he was able to give his surname when asked and
would recognize his given name when spoken to and generally
would react to his name, and on one occasion produced a
coherent sentence in response to the imitation of cats’ mew-
ing. He was unable to name pictures with one exception
(dog) or geometrical figures and would obey only the simplest
commands. Spontaneous drawing produced symbols of no
likeness to the figure requested, while copied drawings were
crude. One note records that when asked to read from print
the word growler with the right eye closed it was read as
clapper, with the left eye closed he spelled out the letters
r-o-w-l.

Under emotion he would occasionally produce a coherent
sentence, and an attempt to stimulate the Babinski reaction
or a threatening movement toward the eyes from the sound
side would often provoke a coherent and fairly fluent flow of
profanity.

During the six years of his residence the records show a
total of over 300 epileptiform convulsions. Many of them
have been well recorded and included tonic spasms only, as
well as tonic spasms of one or more extremity (not con-
stant in origin), spreading to general tonic involvement
followed by clonic spasm, frothing of the mouth, uncon-
sciousness and involuntary evacuations.

Necropsy.—The autopsy was performed six hours after
death.

In the trunk organs there was noted marked sclerosis of
the coronary arteries, fibrous adhesions at the apex of the
left lung, moderate excess of fluid in dependent portions
of the lungs and slight mitral and aortic endocarditis. The
microscopic examination added evidence of a slight broncho-
pneumonia, an early interstitial nephritis and a marked
thickening of the vessel walls in the heart, spleen and
kidneys.

On removing the calvarium the dura proved thickened,
opaque and adherent. The pia was thin and fairly trans-
lucent, and contained a marked excess of clear serous fluid.
The cerebrum weighed 1,055 gm., the cerebellum, medulla and
pons 145 gm. In the right temporo-occipital region was a
large cyst of softening filled with clear fluid (illustration, solid
black). Externally the cyst was covered with pia while in
those parts overlying the ventricles its floor consisted of
ependyma only. It occupied all of the lateral surface of the
temporal pole except the first temporal gyrus and all of the lateral surface of the occipital below a line of prolongation of the first temporal sulcus. On the mesial surface the anterior or recurved portion of the first temporal gyrus and the pyriform lobule were the only parts of the temporal area not involved. In the occipital field on the mesial surface the upper boundary of the cyst was formed by the common calcarine trunk and the calcarine sulcus proper, thus involving all of the visuosensory or visual arrival-platform cortex of this hemisphere, except the cuneus.

On the left hemisphere the first temporal gyrus was very much narrowed and of a softer consistence than the adjacent gyri (illustration, cross-hatched). This alteration was most marked in the midportion, but involved the gyrus from a point beneath the anterior sylvian trunk to the posterior end of the main sylvian fissure, including a portion of the supramarginal gyrus. There was a suggestion of regression below the general cortical level with some pial thickening in two areas of about 2 cm. in diameter, one over the foot of the third left frontal convolution (Broca's area) and the other at the foot of the first left frontal (illustration, light shading).

In the floor of the left lateral ventricle was a wedge-shaped area of softening 1.5 cm. in size lying in the head of the caudate nucleus and extending down into close relation with the anterior limb of the internal capsule.

The basilar artery showed areas of opacity and had a tendency toward irregularity of course though with no definite varicosity. The anterior and middle cerebrials and the left posterior cerebral were similar. The right posterior cerebral was very much thickened and its lumen, which stood open, was very small. Both the anterior and posterior temporal branches were very narrow at their points of origin from the posterior cerebral. The occipital branch in its course along the pial roof of the cyst bore several small opaque yellowish nodules.

The microscopic examination of the cortex from a variety of areas not involved in the gross lesions showed no evidence of alteration of any type, except a moderate thickening of the walls of the arterioles, a slight satellitosis and a slight marginal gliosis. The vascular thickening here was in the main unassociated with perivascular exudate though an occasional vessel could be found which showed a few lymphocytes. Sections from Broca's area showed apparently normal structure.

The atrophic first left temporal convolution was practically a glial cast of the original gyrus with no nerve cells or fibers in the cortex and only a portion of the fibers in the white matter. These intact fibers of the white core were, I take it, the fibers of communication from other cortical areas whose cells were uninvolved.
The cyst of softening showed at its margin the usual close glia meshwork associated with long standing softened areas and a few adjacent vessels showed some perivascular exudate with pigmented phagocytes and a very few plasma cells.

Levaditi stains for the *Spirochaeta pallida* in the large cerebral vessels proved negative.

Sections from the right posterior cerebral artery showed an interesting condition. The much contorted elastic lamina of the original vessel had apparently withstood the degenerative changes which had been at work and remained as a loose, ill-fitting jacket around a very small vessel built up within the lumen of the original. In other sections a marked thickening of the subintimal connective tissue had occurred giving rise to thick masses of fibrous tissue rich in fibroblasts, markedly narrowing the lumen but giving nowhere microscopic evidence of a specific process.

Unfortunately no Wassermann test was done at any stage of the process. The occurrence, however, of lesions of this type in a man of 32 with a definite history of a recent active syphilitic infection leaves little room for doubt that the process was a syphilitic endarteritis.

The similarity of the gross findings and that part of the microscopic picture pertaining to the involved areas to the organic dementias opens a question of how great
the etiologic rôle of syphilis may be in this group. Certain it is that in this case in which syphilis may be fairly safely accepted as the cause, one could not attempt a diagnosis from the histologic picture, and it is entirely conceivable that many of the organic dementias which are similarly negative as regards microscopic evidence might likewise be luetic.

The reflex alterations and paresis of the right side are explained by the area of softening in the left lenticular nucleus and internal capsule, and the hemianopsia, on the basis of the loss of the greater part of the right calcarine cortex. The occurrence of diplopia, however, which necessitated the closing of the left eye, indicates the occurrence in consciousness of two unfused images. The cuneus was the only remaining part of the receptive cortical visual field on the right, and this must then have been the area of reception of the confusing image. The isolated observation recorded above suggests that the right eye, i. e., the left hemisphere, approached more nearly a true image of the object viewed than did the other. One is tempted to explain the occurrence of the confusing image by the comparatively intact group of association fibers leading from the cuneus to the adjacent posterior association field, i. e., parietal cortex, and the diplopia to interference with the interhemispheric association fibers crossing in the corpus callosum, many of which seem to have been lost.

The aphasic disturbance was of the sensory type and must be explained on the basis of the temporal lobe lesions. The method of serial total sections in these cases yields the only accurate localization of the lesions, but renders impossible careful histologic study of the nerve-cells, and in cases studied by this method one must discount the possible occurrence of wide-spread focal or stratigraphic cell losses in fields, which in the total sections are apparently normal, and yet which stand in close functional relation to the areas which have been destroyed. In choosing between these two methods one must take into consideration the clinical findings, and I feel inclined to reserve the total section procedure for those cases showing the least complication, while applying the histologic studies to those whose clinical expression has been complicated, or in which record is incomplete. In the present instance the concurrence of apha-
sia, hemianopsia, diplopia, alexia and agraphia, together with the difficulty of carrying out many tests because of the dementia, has seemed to warrant the use of the second method.

On the left hemisphere the temporal-lobe lesion occupied all of the cortex of the first temporal gyrus from a point below the anterior sylvian fissure around the end of the main sylvian and including a portion of the supramarginal gyrus. All the superior surface of the temporal lobe covered by cortex of the auditosensory type, i.e., transverse temporal gyri and adjacent cortex with the exception of a small portion of the second transverse temporal gyrus near its union with the convolutions of the insula, was also involved. Microscopically, this cortex belonged to the border-line between auditosensory and insular types.

This lesion had not resulted in complete softening but in an atrophy of the cortex with glial replacement. The white core of the gyri still contained a considerable number of fibers which took the stain well, while the losses in the more deeply lying white matter were apparently small in amount.

The lesion of the right temporal lobe was practically complementary to that on the left, i.e., it had obliterated all of the temporal lobe except the superior and transverse gyri.

The patient recognized his given name when addressed and at times would carry out very simple commands, so that if we accept the cortex of the transverse temporal gyri as the arrival-platform for auditory impulses we must either grant a functional capacity to the small practically isolated island of cortex of mixed auditosensory and insular type, or accept a very limited word-hearing capacity reaching the cortical centers of the left hemisphere by way of the right auditory arrival-platform and commissural tracts.

The dementia was of comparatively sudden development and not of the gradually advancing type usually following loss of the word-hearing capacity. This disturbance is better explained perhaps on the tremendous loss in the right hemisphere than on the smaller and probably more slowly progressive lesion of the left temporal lobe. Spitzka has put forward the suggestion that the right hemisphere is the predominant sensory
hemisphere as the left is the predominant motor. The left hemisphere in right-handed persons has been accorded the centers for origination of motor-speech impulses and such localization seems to be definitely proved, but this in no way reacts against the possibility that the right temporal field may be an important storehouse of the auditory memories, which probably form in most persons the basis for internal speech.