CENTRAL ATROPHY*

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Atrophy occurring in muscles the seat of paralysis due to central lesions has been repeatedly observed. Its prevalence is perhaps imperfectly recognized. It is much more frequently present than generally believed and is a symptom not habitually looked for or observed.

In a case reported before the Philadelphia Neurological Society on March 24, 1916, and already published, the atrophy was very marked and was of such unusual origin that I quote it here in some detail (1).

Charles Jordy, aged 24, applied to the Philadelphia Polyclinic Hospital, February 28, 1916.

On examination it was discovered that the leg and to a less degree the arm on the left side were smaller than on the right. This wasting, he stated, followed a gun-shot wound of the cranium in March, 1905.

He was admitted to the University Hospital, and on examination a wound was found over the parietal and motor regions on the right side. The face, arm and leg on the left side were paralyzed and astereognosis was present.

He was operated on a week after admission, the wound was explored, and two or three fragments of bullets and many hairs were removed. Craniotomy was done, and an electrode applied, but without response. The wound was reopened for drainage and two weeks after this it was opened more deeply to permit granulating from the bottom of the wound.

On May 25 he was discharged with some weakness in the left leg and exaggerated patellar reflex, the astereognosis having disappeared. He began to work again in six months, after having been in bed for four months.

On examination at the Philadelphia Polyclinic Hospital on February 28, 1916, it was found that the left arm and leg were distinctly smaller than the right arm and leg. He was able to move the foot at the ankle joint only slightly in flexion and extension. The knee jerks were equal and slightly exaggerated. A left-sided Babinski and Oppenheim were present. On testing his ankle clonus

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none was obtained, but there followed the test an irregular tremor of the thigh. There was an irregular coarse tremor of the left leg, when the muscles were put on tension. Sensation was normal.

There was observed some wasting of the left side of the face. The left arm was smaller by 1.5 cm. in circumference, and the left leg at the thigh was 6.5 cm. smaller in circumference than the right, and the calf 2.5 cm. smaller.

The gluteal muscles were distinctly wasted. No sensory change to pain, touch, heat or cold; nor astereognosis was present.

The X-ray plates located the bullet in the posterior portion of the middle fossa just a little above the base of the brain on the right side.

In view of the fact that the atrophy followed soon after the original injury, it is fair to assume that the present position of the bullet has nothing to do with the causation of the atrophy and that the atrophy was due to the cortical lesion.

My interest in the subject was aroused by this case and through the kindness of Dr. Spiller I was permitted to study the following cases with pathological findings from the material in the laboratory of the University of Pennsylvania.

CASE I. Emma G., age 58, presented left hemiplegia of unknown duration and a right hemiplegia of a few days' duration. It was impossible to obtain any further history on account of the clouded mental condition. The patient, an aged white woman, fairly well nourished and fairly well developed, was paralyzed on both sides. The left arm and leg presented a moderate degree of atrophy. The muscles of both arms and legs were somewhat rigid, especially on the left side. The knee jerks were increased and the Babinski phenomenon was present but there was no ankle clonus.

Each crossed pyramidal and one direct pyramidal tract were degenerated, especially the left crossed pyramidal tract. There was a small area of sclerosis found in each centrum ovale above the internal capsule involving the motor fibers. These areas showed masses of fatty granular cells but no round-cell infiltration.

In the left anterior horn there was considerable change in the cells. Some cells had totally disappeared and some of the remaining ones were swollen and atrophied and had lost their nuclei, and a few were normal.

CASE II. Winifred L., aged 61, admitted with left hemiplegia of unknown duration. The face was slightly turned to the right side and the naso-labial fold on the right side was more distinct than on the left. The tongue was protruded slightly to the right. The left arm was paralyzed and spastic and presented beginning flexure contractures. The left leg was held out straight and somewhat rigidly with marked contracture of the Achilles tendon with plantar flexion. The right leg seemed to be somewhat wasted but there was distinct atrophy of the left leg. The knee jerks were increased and there was positive Babinski on the left.

There was found a moderate degree of round-cell infiltration in the pia in the left paracentral region and in the cortex in this region

there were two more areas evidently of beginning necrosis, as there were compound granular cells present. There was a moderate degree of perivascular distension in the right cortex. In the right foot of the peduncle, corresponding to the second fourth from within outward, there was acute degeneration by the Marchi method. The cord was not studied. This was probably a case of syphilitic disease. No gross lesion was found.

CASE III. George S., aged 69, presented left-sided spastic paralysis of seven months' duration with exaggerated reflexes, double Babinski, and some tenderness over the nerve trunks. There was also weakness of the right arm and leg of three months' duration and the reflexes were exaggerated on this side also. Both legs were held in marked flexor contractures. The arm jerks were exaggerated but the leg reflexes could not be elicited on account of the extreme contracture. The arms were edematous from the elbow downward and over the surface of the elbow there were blisters evidently trophic in origin. There was an area of local gangrene on the heel of the right foot. There was some atrophy of the muscles of the hands including the interossei on the left side. He complained of sharp shooting pains in the limbs especially the lower ones.

There was slight infiltration of the paracentral lobule and thickening of the blood vessels and slight perivascular round-cell infiltration of the cortex here as well as in the perivascular and occipital regions. An area of softening on one side of the pons was found and the pia over the pons was the seat of round-cell infiltration and the vessels were thickened. The pyramidal fibers on one side of the medulla stained less deeply than on the other. In the cervical region of the cord the crossed pyramidal and direct pyramidal tracts showed degeneration on both sides by the Marchi and Weigert methods. The pia of the cord in this region also showed round-cell infiltration to a moderate degree. In the lumbar region the crossed pyramidal tracts were slightly degenerated, the pia infiltrated with round cells, and there was a mild degree of perivascular infiltration within the cord. No gross lesion.

CASE IV. Samuel M., aged 54, presented a left hemiplegia of three years' duration and a second attack of two months' duration on the same side. There was slight paralysis on the left side. The left arm was held in a flexed position across the chest and movements caused pain. The muscles of the arm were markedly atrophied. Temperature sense was impaired but touch sense was not implicated. There was lowered power in the leg muscles and muscles of the leg showed some wasting. The right side was normal. The reflexes were increased on both sides. Babinski phenomenon was present on the right and ankle clonus was absent on both sides.

There was an extensive area of softening in the right basal ganglia. The middle cerebellar artery was occluded. There was no degeneration of the pyramidal tracts by the Weigert method, but intense degeneration was observed by the Marchi method on one side. There was round-cell infiltration of the pia of the paracentral

region on the left side and some perivascular round-cell infiltration of the cortex on the right side.

The anterior horn cells in the cervical region showed a moderate degree of yellow pigment change but otherwise the cells were not diseased. In the thoracic region there was a distinct change in the cells. The nuclei had disappeared, many were displaced, the cells were swollen, though a number of them were not altered. A similar change was found in the lumbar region.

CASE V. Susan W., aged 66, had left hemiplegia of unknown duration. There was marked spastic paralysis of the left arm. When an effort was made to move the arm at the shoulder it appeared to be painful. There was considerable atrophy of the left arm muscles. There was lowered power in both legs and some atrophy present on both sides. The reflexes were increased on both sides but more on the left and there was a positive bilateral Babinski.

There was a linear scar in the left occipital lobe cutting the optic radiations. The right and left lenticular nuclei appeared to be softened. The right lenticular nucleus contained areas of rarefaction, with some fatty and granular cells present. The anterior pyramidal tracts stained well. The cord was not obtained.

CASE VI. Kate W., aged 63, presented a right-sided hemiplegia of five months' duration. There had been a similar attack a month previous on the left side. The tongue, pushed to the right, was not atrophied. There was flaccid paralysis of the right arm with contraction in the extension of the fingers except the little finger which was contracted in flexion. The muscles of the forearm were flattened out and there was wasting of the thenar and hypothenar muscles. The right leg was flaccid, the foot being held in a position of foot drop.

At the autopsy there was found a minute cavity sharply confined to near the center of the outer part of the left lenticular nucleus. There was a focus of hemorrhagic encephalitis of the left occipital lobe almost confined to the upper cortex just below the calcarine fissure. There was a minute old scar in the right optic thalamus which explains the weakness in the right upper extremity. There was an old cyst which cut the uppermost fibers as they form the left internal capsule. The cord was not studied.

CASE VII. Dora K., age unknown, had a right hemiplegia of unknown duration. There was wasting of the tongue around the tip and it could not be protruded beyond the border of the lips. There was some wasting of the right arm and there was marked fixation at the shoulder joint. The right arm was flexed at the elbow, the hand flexed upon the forearm and the whole arm tightly adducted. The right leg was the seat of contractures and presented no atrophies.

A sclerotic area had destroyed the left upper portion of the left optic thalamus and half of the posterior lobe of the left internal capsule. The lenticular nucleus was involved only in the extreme posterior portion and then very slightly. The carrefour sensitif was destroyed and the optic radiations cut. There was a very small

area of sclerosis in the right optic thalamus. There was intense degeneration of both crossed pyramidal tracts more on the right. This degeneration was pronounced in the cervical and slight in the lumbar regions.

The anterior horn cells in the cervical region and the lumbar region showed a moderate degree of change. While there were many healthy cells, many showed yellow pigment change in a pronounced degree in the thoracic region.

SUMMARY OF PATHOLOGICAL FINDINGS

In the first case there was a bilateral lesion of the centrum ovale, degeneration of the pyramidal tract, and change in the anterior horns in the cervical region.

In the second and third cases there were specific encephalitis and meningitis.

In the fourth case there was an area of softening in the basal ganglia and change in the anterior horn cells in the cervical region.

In the fifth case there was a focus in the left optic lobe and the lenticular nuclei were implicated.

In the sixth case the left lenticular nucleus was involved, and there were encephalitis of the left occipital lobe, a focus in the right optic thalamus and one in the internal capsule.

In the seventh case, there was a destructive lesion of both optic thalami, the internal capsule, the lenticular nuclei, and the carrefour sensitif. The anterior horns were degenerated.

If any conclusion may be justified from a small number of cases it would be to support the view that atrophy is caused by lesion of the basal ganglia, as well as of the cortical regions.

A series of thirty-eight cases was also studied in the wards of the Philadelphia General Hospital, for the privilege of which I am indebted to Drs. Potts, Weisenberg and McCarthy, who kindly placed their material at my disposal. The patients were hemiplegics, left and right about equally divided, of an age ranging from 28 to 81 years.

Observation was made for atrophy of the face, tongue, shoulders, arms, hands and legs. The duration of the paralysis was from 17 days to 9½ years. There were four cases of two months' duration, one of ten months' duration, and one of ten weeks' duration, but most of them were from one to several years' duration.

Of 38 cases, 7 showed some atrophy in the muscles of the face. The muscles implicated were those about the mouth and of the cheek adjoining the mouth. It was slight in most cases but pronounced in some and in all cases some palsy of the muscles show-

ing the atrophy persisted. The tongue showed no atrophy in any case.

The deltoid was more or less wasted in 32 cases. The posterior part of the deltoid was the favorite seat of the atrophy, where it was most pronounced, though the anterior and lateral portions were also affected.

The supra- and infra-spinati group were involved in 28 cases.

The biceps and triceps showed wasting in 19 cases, the circumference of the paralyzed side varying from 1.2 cm. to 6 cm., averaging 2 to 3 cm.

The forearm was the seat of wasting in 29 cases.

The small hand muscles were involved in 25 cases, that is the thenar and hypothenar eminences, some to a moderate degree, some extensively and the interossei muscles were wasted in 20 of these.

The leg showed atrophy in 18 instances, involving the gluteal and thigh muscles and in the calf more particularly the anterior muscle group.

There was no evidence of neuritis in any of these cases, notwithstanding the fact that in two cases there was general tenderness of the limbs, and in spite also of the fact that in eleven of the cases pain in the paralyzed limbs was present. This was independent of joint changes or contractures, in fact was spontaneous pain.

The joints showed no change except in the finger joints. While the movements at the shoulder joint and elbows were in all instances more or less restricted by reason of contractures of the muscles, no change in the bones of the joint could be recognized and no fixation was demonstrated except in two cases where the shoulder joint showed some slight change, and relaxation.

On the contrary in the finger joints and the wrist joint some fixation had occurred in seven cases. The fingers were tapering in these cases, the joint fixed, the skin shiny and the condition presented an appearance suggesting the trophic changes observed in neuritis.

While disuse may explain some of the wasting in these cases, it plays but a minor part.

There is a general disuse of both sides in those chair and bed patients who practically are motionless day in and day out, and yet there is atrophy to a pronounced degree on the palsied side. If the wasting were alone due to disuse one would expect little or no difference in the two sides.

Against this view also is the fact that in one case in which there was total paralysis with marked contractures it was impossible to

discover any atrophy in any of the muscles except the posterior deltoid and the forearm. Only in two of the cases was the atrophy so slight as to be negligible including this case.

Atrophy appearing in the paralyzed limbs in cases of hemiplegia of central origin has been attributed to cortical changes, lesions of the optic thalamus, lesions of the caudate nucleus, of the corona radiata, centrum ovale, corpus callosum, the internal capsule, the pons and to leptomeningitis. Lesions of the anterior horns have been assigned as the cause of the atrophy in a certain number of cases. It has been demonstrated also that peripheral neuritis is one of the causes and finally atrophic changes in the muscles due to joint disease, being of the nature of an arthritic atrophy, have been described.

Many instances of cortical lesion have been reported.

Gliky (2) described a glioma of the central convolution, the third frontal, the supermarginal and operculum. Buressi (3) described tubercles in the right anterior lobe, the pons, the medulla oblongata, and with atrophy of the anterior horns. Brissaud (4) later described an area of softening in the fissure of Rolando, in the anterior and posterior part of the marginal convolution and a second case with softening at the foot of the frontal convolution, the middle frontal and superior parietal convolution. There was involvement also of the nerves which showed atrophic change. Quinke (5) in 1888 cited two cases of glioma, one in the right central convolution and paracentral, and the other in the anterior central convolution. Eisenlohr's (6) case was one of abscess of the anterior left central convolution with negative spinal cord. Kramer (7) described a case of leptomeningitis involving the frontal lobe, operculum, gyrus marginalis, and angularis. In Borgherini's (8) case there was a sarcoma of the left central convolution. Senator's (9) case was one of abscess in the left frontal lobe. Petrini (10) cited a case of glioma of the right posterior central convolution. Muratow (11) described a case of atrophy of the frontal and anterior central convolutions, gyrus fornicatus, and callosa marginalis, also implicating the optic thalamus and corpus callosum. There was an involvement also of the cells in the anterior horns which showed marked atrophy. Roth and Muratow (12) cited a case of glio-sarcoma of the right central convolution.

The central ganglion is the seat of lesions in a number of cases, for example, Eisenlohr's case showed the posterior part involved. In Anton's (13) case the posterior part of the optic thalamus was involved. In Jakob's (14) case the optic thalamus and the corpora

quadrigemina. In Klippel's (15) case the optic thalamus and the lenticular nuclei were the seats of lesions. Muratow's case involved not only the optic thalamus but also the frontal and anterior central convolutions and the occipital convolutions. The caudate nucleus was the seat of the lesion in the cases reported by Kirchhoff (16), Monakow and Eisenlohr. The corona radiata was involved in Darkschewisch's (17) case. The centrum ovale was involved in the cases of Charcot (18) and Babinski (19). The pons was involved in Markelowitch's (20) case, the corpus callosum in Steinert's (21) case and the internal capsule in the case of Darkschewisch (22).

Anterior horn changes were described first by Charcot (23) in 1887. Later Brissaud, Babinski, Kirchhoff, Margulies (24), and Dejerine (25) described similar changes. Joffroy (26) and Archard described anterior horn changes and sclerosis of the nerves. They believed that in the majority of cases there was a dynamic irritation in the motor cells analogous to the changes found in peripheral lesion occurring after amputation, such as those described by Marinesco. Margulies found atrophy of the cells of the anterior horns which he attributed to the loss of dynamic action of the interrupted central neuron and the nutrition and function of the cells of the anterior horns.

Peripheral neuritis was first described by Dejerine (27) in 1889, who found in four cases in which the spinal cord was negatively altered nerves in the thenar region. Brissaud found atrophied nerves with fatty granulations of the transverse striations and multiplication of the embryo-plastic nuclei between the primary fibers.

Muscle changes were described by Löwenthal (28). The nuclei were round and moderately numerous and many fibers were thickened and polygonal and the nuclei were in rows.

Joint changes were described by Darkschewisch who found diseased joints in six of nine cases. Gilles de la Tourette (29) found arthritis change in ten of twenty cases. Steinert found in seventeen cases muscle changes consisting of thickening of the muscle fiber and increase in the nuclei. There was nothing found in the peripheral nerves. In a later case there was a moderate atrophy of the nerves of the affected side.

Experimental work adds some data. Lewy (30) experimented upon apes, causing atrophy in the paralyzed muscles of cerebral and spinal origin. In the atrophic muscles there was no change in the transverse striations and the muscles were really free from degenerative processes.

Caracciolo (31) experimented on eleven dogs, removing portions of the brain, and explained the atrophy which followed on the basis of the propagation of the cerebral lesions to the cells of the anterior horns by way of the descending fibers which degenerated.

The atrophy appears to be late usually according to Borgherini but in one case which he reported it occurred in seven days; in Gliky's case the paralysis was of sixty-three days' duration; in Burassi's case twenty-four days; in Patella's (32) case two months' duration; in Quinke's two cases four and five days' duration. Margulies says that the duration is usually two months while Leri stated that it may occur at the end of the first week but usually is an old manifestation. In Kirchhoff's case it was of four months' duration.

Central pain and atrophy may be associated as in the case of Brissaud and Kirchhoff, and in my own cases.

It may be concluded from this study that (1) cortical lesions give rise to atrophy in the paralyzed muscles, which is independent of spinal cord changes as in the cases described by Eisenlohr and others.

It may further be stated that in a number of cases there are associated changes in the anterior horn cells which may be looked upon as secondary to the cortical lesion. (3) Some cases of hemiplegia show little atrophic change even when the palsy is pronounced. (4) Neuritis plays some part in the causation of trophic muscle changes found in central lesions. (5) Disuse plays a minor rôle in the causation of the atrophy. (6) Arthritic atrophy does not often occur. (7) Atrophy in paralyzed muscles of central origin is common and its absence unusual.

The shoulder muscles, especially the posterior part of the deltoid, show the atrophy most usually and in greater degree than the other muscles. All the arm and forearm muscles are usually involved and the small hand muscles are almost always implicated.

The legs show the atrophy to a less degree than the arm muscles.

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