GLIOMA IN THE FOURTH VENTRICLE, WITH INVOLVEMENT OF THE TRIANGULAR VESTIBULAR NUCLEUS.

REPORT OF A CASE WITH SYMPTOMS OF DYSTONIA, DYSMETRIA, TREMOR AND TONIC FITS.*

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INTRODUCTION.

THE great interest shown of late years by anatomists, physiologists and clinicians in the mechanism of the motor nervous system has included a considerable research on muscle tonus. Muscle hypertonicity, with which we are concerned in this case report, may be defined as such an increase and alteration in muscle tension as give a constant fixation unassociated with fatigue or increased metabolism. A typical physiological example is the action of a sphincter muscle. The most common pathological example is the spastic rigidity of the cerebral hemiplegic; and from the experimental physiological side the celebrated experiments of Sherrington producing decerebrate rigidity by midbrain section are classical. Such increase of tonus affects predominantly the extensor or antigravity muscles, and therefore has aptly been termed postural tonus. This tonus is stimulated and maintained by impulses which attain the central nervous system by the afferent spinal nerves of the extremities of the neck, and by the vestibular apparatus. Ewald was the first to study alterations in muscle tonus in consequence of experimental destruction of the labyrinth. The recent paper of J. Gordon Wilson on labyrinthine tonus is an excellent summary of this particular phase. Tonic neck and vestibular reflexes have been particularly studied separately by Magnus and de Kleijn in decerebrate animals—by labyrinthine destruction in one set of experiments, and by eliminating the neck reflexes in others. For example, if the neck reflexes are eliminated and the experimental animal is placed in different positions in space it will be found that the maximum extensor tonus affecting all four extremities is produced by a head position of 180° , which corresponds to the supine position with a somewhat raised snout. In the reversed position, normal head position with somewhat lowered snout,

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the extensor tonus is at a relative minimum, although the preparation is still in evident decerebrate rigidity. In other head positions the extensor tonus is intermediate in degree. Leon Meyers has recently summarized this research and also contributed to the subject of neck and vestibular reflexes in clinical cases by publication of two personal cases with necropsy reports.

In the Leyden lecture (1923) Magnus reported on further experimentation growing out of his various studies on tonic neck and vestibular reflexes. Cross-sections of the central neural axis were made from the medulla upwards instead of the usual sections from the cerebral cortex downwards. A cross-section anterior to the eighth nerve exit will produce complete decerebrate rigidity, which disappears on cross-section in the region of the calamus scriptorius. Proceeding cephalad, decerebrate rigidity is still existent at sections through the anterior part of the midbrain, but a section cephalad to this, leaving an intact midbrain ('midbrain animal'), shows a different picture. There is an even distribution between extensor and flexor tonus, and this animal, contrary to the decerebrate animal, will regain the upright position after falling. This act has been termed a 'righting' reflex (Stellreflex). Further, the animal will walk normally when stimulated. From an analysis of ingenious experiments (free suspension of the animal and observation of head positions in relation to body positions with and without intact labyrinths) the 'righting' reflexes, at least in part, are shown to depend on receptors in the labyrinths (labyrinth 'righting' reflexes). The labyrinth 'righting' reflexes on the body are elicited from the maculae of the otoliths of the sacculus.

If a 'thalamus' rabbit (intact thalamus), whose labyrinths have been destroyed, is suspended, the head will show complete disorientation. If the animal be then placed on its side on a table the head will assume a normal position. This position will again be changed if symmetrical pressure be made on the opposite or upper side of the animal by means of a weighted board. These reflexes, which depend on the body pressure sense, are designated as body 'righting' reflexes on the head. When the above reflexes are acting the changed position of the head brings a third set of reflexes into being; the neck reflexes act on the forepart of the body, and these in turn act on the posterior part, in the manner of chain reflexes. They may be designated as neck 'righting' reflexes on the body. Finally, body 'righting' reflexes on the body may be demonstrated through the body pressure sense, after it has been previously forced into a physiological position through the neck reflexes. These four different reflexes doubly assure head and body position, and may replace one another by defect.

In another set of experiments brain stem sections and brain punctures controlled by the state of the 'righting' reflexes were made to

determine localizations. The centre for the neck 'righting' reflexes lies in the anterior portion of the medulla and in the pons region. It is definitely established by these experiments that the intact red nucleus prevents decerebrate rigidity and determines normal tonus distribution to the extremities. It is certain, further, that the red nucleus is the centre for the labyrinth 'righting' reflexes, and extremely probable that the body 'righting' reflexes also pass through this nucleus. According to Magnus, the central and efferent paths for normal tonus distribution and for a part of the 'righting' reflexes are established. After complete anatomically controlled cerebellar extirpation the labyrinth 'righting' reflexes on the head and the body 'righting' reflexes on the head still persist. The body 'righting' reflexes on the body, however, are diminished, which makes it possible that at least in part the tracts concerned with these reflexes pass through the cerebellum.

S. A. K. Wilson has called attention in human cases to the extension-pronation phenomenon in the upper extremities, head retraction and opisthotonus, illustrating a mechanism similar to or identical with decerebrate rigidity. There are frequently associated tonic fits, which in themselves are compared to decerebrate rigidity. Incomplete decerebrate attitudes are postulated. The position of the upper extremity in decerebration is compared with the position of this extremity in the ordinary Wernicke-Mann type of hemiplegia, with the exception that in the latter there is flexion instead of extension of the forearm.

This same author, in a recent important paper, discusses the central nervous station for tonus: "In respect of the mesencephalon, noteworthy motor reactions are obtainable in the decerebrate animal. We owe largely to the work of Graham Brown our knowledge of this part of the subject. Unipolar stimulation of the cross-section of the midbrain obtained by decerebration at the level of the anterior colliculi (anterior corpora quadrigemina), at a point entirely dorsal to the corticospinal tract in the crus, constantly produces a definite, specific postural motor reaction on the part of the animal experimented on. The area from which this result is invariably obtained is dorsal in the tegmentum and includes the region of the red nucleus, the part of the superior cerebellar peduncle running to it (tractus cerebello-tegmentalis) and the posterior longitudinal fasciculus. The attitude is as follows: the head is tilted back and also twisted so that the face looks to the side stimulated; the homolateral arm is flexed and the opposite one extended; the leg of the same side, on the contrary, is extended and the opposite one flexed (as a rule); the tail erects and is bent to the stimulated side. The back is usually slightly convex to the opposite side. When stimulation has ceased, the posture may continue unchanged for many seconds, even minutes."

Oliver Strong has discussed the mechanism of decerebrate rigidity;

ablation of the cerebellum does not abolish it (Sherrington). Injury to the vermis and partial ablation of the cerebellum may produce it; and after cerebellar operation decerebrate rigidity often sets in, but not always. Weed states that removal of the anterior portion of the superior vermis causes extreme extensor rigidity. Strong divided the efferent somatic nervous system into four integrative levels, viz. (1) lower motor neurone, (2) ground bundle reticular formation of cord and brain, including Deiter's nucleus and the nucleus ruber, (3) suprasegmental mechanisms of Meyer comprising the colliculi, portions of the cerebellum and older portions of the forebrain, mainly the globus pallidus and related structures, and (4) the pallium.

The case to be presented is exceptional in that a central progressive lesion (glioma) involved the triangular nucleus in the floor of the fourth ventricle, and this was the only important nucleus in the medulla materially affected. In the conclusion following the case reported the rôle of the vestibular nucleus in the production of the hypertonia and tonic fits will be discussed, and the relative importance of the other concomitant lesions.

CLINICAL CASE.

Case History.—T. W. (Dispensary No. 34404 Leland Stanford, Jr., University Medical School), female, age three years, was admitted to the children's ward on October 25, 1915, in the service of Dr. Langley Porter, where she remained two days for observation and diagnosis. As a consultant, I am indebted to Dr. Porter for permission to publish the case history.

History.—October 25, 1915. The patient was the second child born of healthy parents. There was no history of significant family disease or abnormal heredity. The birth lasted eleven hours and the baby was born asphyxiated although no instruments were used. Notwithstanding this difficult birth she developed normally, sat up at usual age, walked at 12 to 16 months, talked at two years, and compared favourably in development, according to the mother, with other children of similar age. There were no serious illnesses in the second year, and she apparently enjoyed normal health up to the onset of the present illness. In May, 1915, there was a fall down stairs, without apparent serious results. Five weeks later it was noticed that she staggered when she walked, reeled to the right, and would throw the right foot to the right. Later, vomiting occurred with no relationship to food. In July 'spasms' commenced, and continued until September, when they ceased. Some days there were as many as seven. These spasms were described as 'stiffening,' the eyes would roll up, and such a spasm would last for a few seconds to a few minutes. In August the sight began to fail; she would ask who was talking to her. Occipital

headaches were complained of. Intelligence was apparently unimpaired. After onset of the spasms she was unable to stand, and soon afterwards was even unable to sit; the use of the extremities also became increasingly difficult. The sense of taste was involved, so that she was unable to recognize anything sweet. Finally, control of the urine was lost and the body weight increased rapidly. Hearing was not affected.

Examination.—October 25–26, 1915. The child was fat, well grown for its age, of apparent normal mentality, but peevish and difficult to examine. The fontanelles were closed. The posture was in dorsal decubitus. On account of hypertonus there was a constant tendency to fall backward when supported in the sitting position. The head was held rigidly on the neck, but not turned. A very striking part of the picture was the extreme hypertonicity of the muscles of both extended lower extremities, although this was greater in the distal as compared with the proximal parts of the members. The legs were extended on the thighs and the feet on the legs. The feet were arched. There was moderate adductor spasm. The upper extremities assumed a position of flexion of the forearms on the arms, with spontaneous tremor. In so far as it was possible to test coordination there was marked dysmetria, especially in the left upper extremity.

The tendon reflexes were lively and apparently equal on the two sides, although the responses varied because of the dystonia. There were no definite pathological reflexes affecting the great toe, although on one occasion the Babinski sign was suspiciously positive on the right side. Somatic sensation was not disturbed. McEwan's percussion sign (cracked-pot note) was well marked in the right frontal region. Roentgen-ray pictures of the skull showed absorption of the inner table. The pupils were dilated and reacted sluggishly to light. An inconstant vertical nystagmus was occasionally noted. Apparently vision was markedly defective. Both optic discs were pale, poorly outlined, and the neighbourhood vessels were small. No facial paresis was noted. The face and the arms occasionally twitched. There was no bulbar paralysis, and the speech was clear.

Functional vestibular tests were made by Dr. H. B. Graham. "Ears: Drums intact, hearing evidently good. Caloric: cold in either ear gives a drawing of the eyes to the same side, with a large slow nystagmus to the opposite side. Spontaneous nystagmus to either side on looking to that side. Eyes move unequally. There is a corneal anaesthesia on either side (not total). There seems to be some interference with the vestibular cerebellar tract, but more central than cortical."

The thoracic and abdominal viscera were normal. The routine laboratory tests, including a Wassermann reaction in the blood and the cerebrospinal fluid, were negative. The cerebrospinal fluid pressure

was recorded at 180 mm. of fluid ; it came in drops, and 10 c.c. were removed. Because of admixture of blood a complete analysis could not be made. There were no apparent harmful results from the puncture.

While in the hospital a typical convulsion was observed and described : " The child drew a long breath, the left upper extremity was extended above the head because of spasm of the deltoid and trapezius on that side ; this phase lasted for perhaps half a minute, when a slight clonus of the hand occurred. During this time the right upper extremity was held in tonic spasm parallel with the body, the increased tonus finally giving way to clonus, slight in the forearm,

marked in the hand. At the same time there were irregular and fugitive movements of the eyes."

The diagnosis of tumor cerebri was made, probably cerebellar, with complicating symptoms of an internal hydrocephalus. The patient was operated upon in another hospital on November 2, 1915, by Dr. Sol Hyman, of San Francisco, and died November 4, 1915. The record of the operative procedure, operative findings, and post-operative course, was unfortunately lost. At the necropsy the brain only was removed.

PATHOLOGICAL EXAMINATION.

After hardening in liquor formaldehydi, the brain was sectioned and a cerebellar tumour was found filling the fourth ventricle. No other lesions were found above the cephalic end of this ventricle. The cerebellum, medulla, midbrain and interbrain were included in celloidin without previously chromating and transverse serial sections were made of the entire block. Every other section cut was preserved. Mlle. Loyez*' method of staining myelin fibres was used with excellent results, the employment of this method permitting

FIG. 1.—Loyez stain. Posterior pole of cerebellum. Tumour in fourth ventricle.

FIG. 2.—Loyez stain. Tumour mass filling fourth ventricle, and compressing floor.

* Roussy-Lhermitte : *Les techniques anatomo-pathologiques du système nerveux*. Immersion in 4 per cent. solution of ferric ammonium sulphate for twenty-four hours as mordant ; stain in alkaline hæmatoxylin for twenty-four hours ; differentiate in above mordant and Weigert's decolorant.

alternate sections to be stained by Nissl, and by hæmatoxylin and Van Geison. In all 300 sections were stained.

The posterior part of the cerebellum (*Fig. 1*) was infiltrated by tumour, which involved the left hemisphere somewhat more than the right. The whole of the fourth ventricle was filled with a tumour mass (*Fig. 2*), causing great dilatation. The tumour originated from the cerebellum itself, possibly from the subependymal glia. The morphology alone was not sufficient to determine the direction of growth. The histology of the tumour was studied through serial sections and not by special stains. It consisted chiefly of round cells with large nuclei and small peripheral protoplasm. Between the cells hair-like glial fibres could be demonstrated. It was moderately vascular and showed no

FIG. 3.—Loyez stain. Upper part of fourth ventricle. Nodulus involved.

FIG. 4.—Loyez stain. Upper end of tumour. Left superior peduncle and left lateral lemniscus attenuated and fused.

FIG. 5.—Loyez stain. Level of the decussation of the superior peduncles.

areas of degeneration or hæmorrhage. Histologically it was classified as a rapidly growing cellular glioma.

The tumour cells infiltrated the triangular nucleus (vestibular nucleus) superficially and bilaterally and to a considerable portion of its extent. The cephalic end where the nucleus attains a more mesial position was most involved. In this locality the tumour was found to break through the striæ medullares and invade the substance of the nucleus (*Fig. 6*).

Deiter's nucleus was not involved. The dentate nuclei were markedly compressed and flattened bilaterally (*Fig. 2*), the left being more involved than the right. The roof nuclei were directly involved, the nucleus emboliformis and nucleus globosus only partially persisting on the right side. The inferior vermis was invaded, the nodulus (*Fig. 3*) being involved but distinctly defined.

The upper portion of the tumour corresponded to the cephalic end of the fourth ventricle. In this region (*Fig. 4*) the left superior peduncle

and lateral lemniscus were attenuated and fused ; the superior peduncle on the right was well marked. The decussation of the superior peduncles (*Fig. 5*) showed no apparent defect, thus indicating more function of the dentate nuclei than one would suspect from their marked compression. At and above this level no pathological condition was demonstrable.

The lower portion of the tumour invaded the vestibular nucleus and the medulla superficially in the region of the median raphe above the hypoglossal and the dorsal vagal nuclei (*Fig. 7*). These nuclei, however, were not involved.

DISCUSSION.

In the foregoing case report we have a fairly definite history of onset of illness in June, with a rapid development of symptoms, so that

FIG. 6.—Loyez stain. Tumour breaking through the striæ medullares and involving the triangular nucleus.

in October the clinical picture was well marked and advanced. The loss of vision and optic atrophy, with inner table absorption demonstrated by *x*-ray, would ordinarily indicate a longer period of intracranial pressure than the clinical history suggested. The tumour invaded the whole of the fourth ventricle but did not, however, interfere entirely with cerebrospinal fluid circulation, as 10 c.c. were removed by lumbar puncture without shock, and there was no evidence of papilloedema or engorged retinal vessels at the time of our examination.

With the exception of the greatly dilated fourth ventricle, the chief pathology due to the tumour growth was threefold : firstly, the

cerebellar hemisphere involvement ; secondly, the vermis involvement ; and thirdly, involvement of the triangular vestibular nucleus. To correlate the clinical symptoms with the pathology, the dysmetria occurred principally on the left side, or side of the greatest hemisphere and dentate nucleus involvement. The lack of maintaining any position

FIG. 7.—Nissl stain. Lower part of medulla. T = Tumour. C = Central canal.
D = Dorsal vagal nuclei. H = Hypoglossal nuclei.

of the trunk might be explained by implication of the vermis and the roof nuclei. The increased muscular tonus and maintained attitudes, as well as tonic fits, might well be explained by a central vestibular stimulus originating in the vestibular nucleus because of invasion and irritation by the involving tumour and its pressure on this nucleus, which lies superficially in the floor of the ventricle. This latter lesion can possibly explain also, or at least in part, the inability to change trunk posture by reason of the increased muscular tonus.

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